Post-Liposuction Paraspinal Desmoid Fibromatosis: First Case Report and Literature Review

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ABSTRACT

Desmoid fibromatosis is a rare, slowly growing, locally aggressive mesenchymal tumor. It primarily affects young adult females with a female-to-male ratio of 2:1. It rarely affects children and elderly patients. Most desmoid fibromatoses arise in extra-abdominal locations, mainly, the extremities. We report the case of a 39-year-old female patient who presented with a lower back post-liposuction paraspinal desmoid fibromatosis. Lumbar spine magnetic resonance imaging (MRI) revealed a mass in the right paramedian area opposite to the level of L2, not infiltrating the underlying structures, with normal overlying subcutaneous tissue. Wide local excision with free margins was performed. Literature review revealed 97 cases of paraspinal desmoid fibromatosis. We extensively analyzed the clinical, radiological, and pathological features of these cases. Fibromatosis is a non-metastasizing, but locally aggressive slowly growing tumor currently classified by the World Health Organization (WHO) as a mesenchymal tumor of intermediate (borderline) malignancy. The exact cause of desmoid fibromatosis is not clear. It has been narratively linked to genetic abnormalities, trauma, steroid sex hormones, and bone malformations. The diagnosis of fibromatosis is challenging due to its ambiguous and pathological features. To the best of our knowledge, this is the first report of a post-liposuction paraspinal desmoid fibromatosis and is the first report of a paraspinal desmoid fibromatosis in the Arabian Gulf region. Universal guidelines for the management of desmoid fibromatosis are still lacking.

Keywords: Desmoid tumor, fibromatosis, mesenchymal tumor, liposuction, trauma.

INTRODUCTION

Fibromatosis is a non-metastasizing, but locally aggressive slowly growing tumor, currently classified by the World Health Organization (WHO) as a mesenchymal tumor of intermediate (borderline) malignancy [1, 2]. It is considered a rare tumor accounting for approximately 0.03% of all neoplasms, and less than 3% of all soft tissue tumors [3]. The incidence of desmoid fibromatosis is 3-4 cases per million per year, mainly between the age group of 15 to 60 years, with a female predominance [3, 4]. The clinical presentation depends on the location of the tumor. The most common anatomic location is within the extremities (30 - 40 %), followed by the abdominal wall (15 %), chest wall (20%), abdominal cavity (15%), head and neck (10%) and rarely involves paraspinal region, or flanks [5-8]. The etiology remains unknown. However, genetic factors, trauma, steroid sex hormones, and bone malformations seem to contribute to the development of the disease. Microscopically, the majority of desmoid tumors are characterized by the proliferation of fibroblastic spindle cells within a collagenous stroma. The differential diagnosis of desmoid fibromatosis is challenging. It should be differentiated from bony lesions, including giant cell tumors, chordomas, and fibrous dysplasias of the spine. At the histological level, primary low-grade fibrosarcoma of the bone is the most important differential diagnosis [9]. Herein, we report the first case of a post liposuction paraspinal desmoid fibromatosis.

CASE REPORT

A 39-year-old female patient underwent an uneventful abdominoplasty with lower back liposuction. Six months postoperatively, the patient felt a painless swelling in her lower back. A soft, small lump was detected in the right lower paraspinal area (lumbar) on physical examination. Lumbar spine Magnetic resonance imaging (MRI) revealed an abnormal signal intensity along the right superficial perimuscular fascia in the right paramedian area opposite to the level

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of L2. The size of the lesion was 28x11x30 mm. It was a well-defined fusiform hypointense lesion in T1W with a subtle hyperintense signal in T2W sequences at the fascia of the subcutaneous tissue. The right paravertebral muscles showed an enhancement pattern which mimics fibrous tumor nodular fasciitis (Figure 1). There was no evidence of infiltration of the underlying or the overlying subcutaneous tissues.

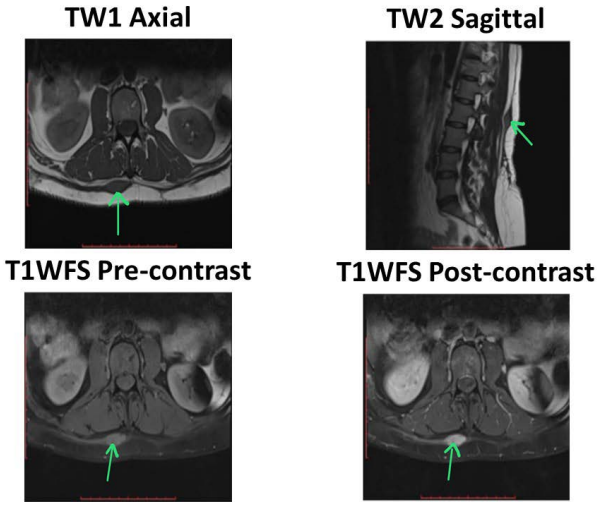

Figure 1. Magnetic resonance imaging revealing oval hypointense lesion at posterior fascia in TW1 axial, subtle hyperintense in TW2 sagittal, pre-contrast subtle hyperintense and post-contrast homogenous enhancement in T1WFS.

The patient refused biopsy at this stage. Two years later, the lump became painful and increased in size. Upon palpation, it was tender and hard in consistency with no skin changes. Ultrasound showed the same lesion in the right paraspinal area. It measured 3.4x5.3 cm in size, with no evidence of muscle infiltration. The clinical impression was either a lipoma or a soft tissue tumor. Tru-cut biopsy of the paraspinal mass suggested a benign fibrous tissue lesion, either fibromatosis or fibroma (Figure 2).

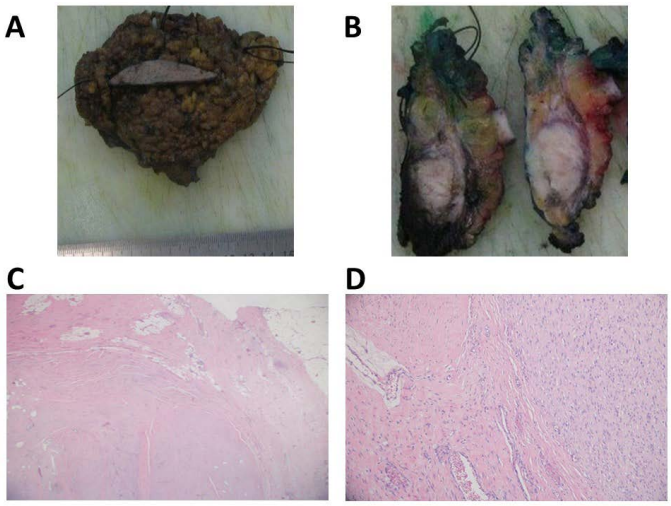

Figure 2. Tru-cut biopsy histomorphological findings revealing fibro-collagenous tissue interdigitating through the hosting adipose tissue [HandE stained MPF]

Surgical intervention was decided, and a wide local excision with a free margin was performed. The excised mass was sent for histopathological assessment. Microscopically, the tumor was formed by a bland uniform spindle cell proliferation with variable fibroblastic/myofibroblastic appearances. The neoplastic spindle cells run in sweeping fascicles with apparent elongated, spindly cytoplasm within a background of stromal fibro-collagenous component, including occasional peripheral lymphoid cell aggregates. Scattered stromal thin-walled curvilinear capillary-size blood vessels were present. No significant pleomorphism, large cellular forms, or significant hyperchromasia were seen. There were no perineural or lymphovascular invasions. No hemorrhage, necrosis, or calcification was noted, and the overall tumor mitotic activity was exceedingly low (Figure 3).

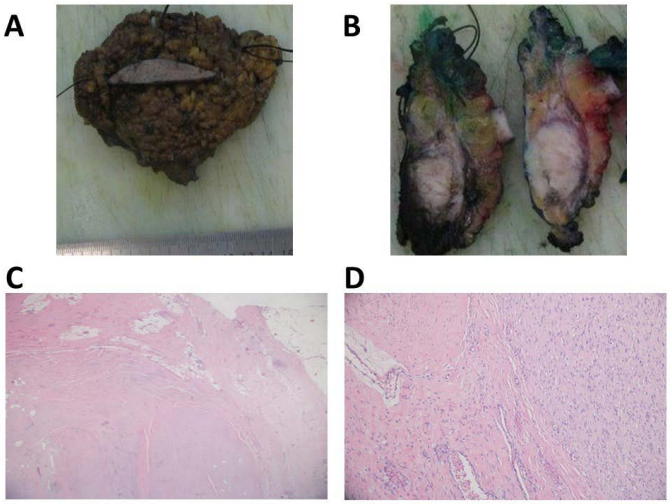

Figure 3. (A) and **(B)** demonstrate the macroscopic appearance of the wide local excisional specimen of the paraspinal mass. **(C)** Photomicrograph showing a nodular, relatively well-delineated, and formed by a bland uniform spindle cell proliferation with variable fibroblastic/myofibroblastic appearances (HandE stained LPF). **(D)** Photomicrograph showing neoplastic spindle cells run in sweeping fascicles with apparent elongated, spindly cytoplasm within a background of stromal fibrocollagenous component and scattered stromal thin-walled curvilinear capillary size blood vessels. No significant pleomorphism, large cellular forms, or significant hyperchromasia is seen (HandE stained MPF).

The neoplastic spindle cells werw diffusely positive for smooth muscle actin (SMA) (diffuse 3+), while Beta-Catenin and Cyclin D1 were multifocal immunoreactivities (1+ - 2+). Estrogen receptor (ER), progesterone receptor (PR), CD34, CK AE1/AE3, Desmin, CD117, SMM, Muc4 and S-100 protein were all diffusely negative. Tumor proliferative index (by Ki-67) <2%. No molecular studies for CTNNB1 and adenomatous polyposis coli (APC) mutation were performed. Based on the above histomorphological and immunohistochemical findings, the final diagnosis of Paraspinal Fibromatosis, Extra-Abdominal Desmoid Type, was concluded.

The blocks and slides of the case were sent abroad to a pathology lab specialized in soft tissue tumor for a second opinion. The diagnosis of Paraspinal Fibromatosis, Extra-Abdominal Desmoid Type was confirmed. The case was discussed at Tumor board meeting of Bahrain National Oncology Centre, and the plan was for a six-monthly follow-up lumbar spine MRI.

DISCUSSION

Our experience with a 39-year-old female patient who presented with the previously unreported lower back post-liposuction paraspinal desmoid fibromatosis prompted us to extensively review the related literature.

We encountered 97 cases of paraspinal desmoid fibromatoses. We analyzed the clinical, radiological, and pathological features of these cases (Tables 1 and 2).

Desmoid fibromatosis is a rare mesenchymal tumor that is locally aggressive. It primarily affects females with a previously reported female-to-male ratio of 2:1. Our review of literature showed a female predominance of 73 % [4, 10]. The reported ages at the presentation ranged between 1– 84 years. Few cases have been reported among the pediatric age group. The most reported age was 14 years. Based on our literature, the mean age at presentation was 33.8 years. The size of the tumor varied from one patient to another. The exact cause of desmoid fibromatosis is unknown. In our literature review, 32 of the cases occurred postoperatively, 7 cases gave a history of a preceding trauma, and five cases had a genetic predisposition.

While reviewing the genetic causes, Schlag et al. reported an association with CTNNB1 mutation [11]. A mutation in the APC gene results in a syndrome known as familial adenomatous polyposis (FAP), which accounts for 10% of desmoid tumors affecting 5 to 30% of FAP patients, particularly those with the Gardner subtype [12]. Kenning at el. reported an association with Gardner's syndrome, R Luo et al. found a link to b-catenin mutation, Neffa et al. also found a link to APC gene mutation, and Vitfell-Rasmussen et al. case was associated with familial adenomatous polyposis (FAP) [8, 11, 13].

Other genes have been involved in the development of desmoid tumors, including a mutation in the beta-catenin gene that has been implicated in most sporadic desmoid tumors. This mutation dysregulates the Wbt/B-catenin signaling pathway and, in turn, alternates the normal immune response to neoplasia. The degradation of b-catenin is regulated by the APC gene, which is also implicated [14].

Trauma has been hypothesized that initiates inflammation which in turn upregulates beta-catenin. If beta-catenin degradation is impaired, transcription factors can pile up and upregulate WISP1, SOX1, ADAM12, and Fap-1, leading to the abnormal proliferation of monoclonal fibroblasts and extracellular matrix proteins. Spinal surgeries, as a cause of trauma, have been listed as a risk factor for the formation of desmoid tumors [15]. We suggest that the surgical trauma (liposuction) in our case was the initiating cause for the formation of the desmoid tumor.

In regard to endocrine causes, it has been suggested that women of childbearing age have a higher risk of developing aggressive fibromatosis [16].

It has been established that patients with extra-abdominal desmoid tumors have pre-existing bone malformations in the appendicular and axial skeleton. Hara et al. documented that about 80% of the affected patients had minor bone abnormalities demonstrable on radiographs compared with 5% in a control population. Abnormalities include cortical thickening, exostoses, cysts, deformities in long bones, osteomas, anomalous teeth in mandibles, Scheuermann's disease, scoliosis, and anomalous vertebral bodies in the spine might be present [17].

The symptoms of desmoid tumors differ depending on the location and size of the tumor. In some cases, the tumor was discovered incidentally on imaging for another complaint [18]. Most patients present early with painless swelling similar to our case. Later patients tend to experience pain, possibly because of deeper tumor invasion. When the tumor expands further, it might compress or invade local nerves resulting in neurological symptoms such as radiculopathy, paresthesia, or motor weakness [19].

In our literature review, pain was the most associated symptom. Sixty-one patients presented with a painful mass, compared to 36 patients who presented with a painless mass. The mass was soft in 1 case, firm in 26 cases and hard in 11 cases. In the majority (54 cases), the mass was tender [5-13, 15-18, 20-67].

Fibromatosis involves a spectrum of fibroblastic proliferations that can be divided into superficial and deep. Superficial fibromatosis is characterized by slowly growing tumors that develop insidiously in superficial structures as in our case. In contrast, deep fibromatosis is often large and rapidly growing tumors that involve deep anatomic structures and are known for their high rate of recurrence [6]. Deep fibromatosis is usually classified as intra-abdominal, abdominal, or extra-abdominal. The intra-abdominal desmoid tumors emerge from the mesentery and the pelvis. They are challenging to treat and are often associated with FAP in Gardner's syndrome. Those with APC gene mutations are usually reported as rare cases of multiple desmoid tumors [12].

Abdominal desmoid tumors usually arise from the rectus or internal oblique muscles and fascia in parous women, young pregnant women, or oral contraceptive users. The intra-abdominal and abdominal desmoid tumors account for 57% of the total cases [18]. The extra-abdominal tumors constitute 43% [18, 19, 22, 68, 69].

A plain X-ray of the involved area with a desmoid tumor is usually normal. Sometimes, calcifications or cortical erosions may be seen due to the extension of the growing tumor into the adjacent bone. The medullary extension is not usually seen. Ultrasonography shows nonspecific findings such as a poorly defined, hypoechoic soft tissue mass or an occasional posterior acoustic shadow associated with more extensive lesions. In our case, ultrasonography showed nonspecific findings. Computed tomography (CT) is of little diagnostic yield. The intravenous contrast might increase enhancement due to the tumor's high angiogenic activity. MRI is considered the investigation of choice to assess location, size, and type. Desmoid fibromatosis is homogeneously isointense on T1-weighted images and highly heterogeneous on T2-weighted images. The signal is significantly enhanced with intravenous contrast administration and is decreased in cases where the collagen component of the tumor is high secondary to hypocellularity [19].

In our literature review, MRI was the most widely used modality in the diagnosis of desmoid fibromatosis (n=70). Other used modalities included: CT (n=40), x-rays (n=27), ultrasound (n=11), bone scan (n=2), positron emission tomography (PET) scan (n=2), electromyography (n=1), thallium scintigraphy scan (n=1), and nuclear magnetic resonance (n=1). Although the signal patterns on MRI can change throughout the disease [11], MRI is still superior to other radiological modalities because it shows if the tumor is invading the neighboring neurovascular structures or not, as we demonstrated in our case [19].

A preoperative core biopsy is usually required due to these nonspecific imaging findings [6]. In literature review preoperative diagnosis of desmoid fibromatosis was confirmed in 30 cases only. Provisionally the diagnosis after biopsy was fibromatosis (n=28), soft tissue mass (n=14), malignancy (n=5), hematoma (n=3), infection (n=2), and autoimmune (n=1) [5-13, 15-18, 20-67, 70-91]. In our case, the true-cut biopsy was not conclusive, and the final diagnosis was reached only after wide local excision of the tumor.

The present literature review revealed that tissue diagnosis was mostly obtained by open surgical biopsies (n=33), either incisional or excisional. Fine needle aspiration cytology (n=10), true-cut biopsy

Table 1. Clinical Presentation of the Reported Cases in Literature

Reference	Clinical presentation and Physical examination findings
Our case	-Painless swelling in the lower back -Two years later, it became painful, a small soft lump in the right para midline area was felt. -Two years later, it became hard in consistency
Lee et al. [7]	Pain in the lower back and buttock with palpable non-tender mass at the level of the left L5-S3
Neffa et al. [8]	Age of 21: Severe pain with neuropathic component
Lacayo et al. [6]	Painful, palpable firm neck mass (all four cases)
Alherabi et al. [5]	-Left neck mass for a year which was gradually increasing in size -On examination, hard, non-tender, fixed neck swelling involving the left postero-lateral aspect of the upper neck
Daneyemez et al. [9]	-Complaints of headache and neck pain radiating to the left arm with numbness for 2 months -The pain was slowly increasing and was unresponsive to analgesics -On physical examination, local tenderness was observed in the C5 area, no change in motor function
Remedios et al. [10]	Mild backache
Schlag et al. [11]	Painful right sided neck swelling
Kenning et al. [12]	-A 15-month history of headaches, and numbness progressing from the right side of her face and extending into the right side of her neck, arm, and first three digits. -2 years after: The growth of the mass caused local discomfort in the neck, hyperreflexia in the right upper and lower extremities, a right Babinski sign and poor heel-to-shin coordination. -2 years after the initial surgery: There was an enlarging mass in the right neck and occipital neuralgia.
Luo et al. [20]	-1 year history of recurrent, dull left neck, shoulder, and upper arm pain, occurring at night and was not accompanied by numbness or pain of the limbs, dizziness, or headache and not relieved with non-steroidal anti-inflammatory drugs or analgesics -> 3 months before admission: neck and shoulder pain became worse, with radiation to the left deltoid and lateral upper arm -A poorly mobile, tender, deep mass was palpable on the left side of the neck, not warm, red, or ulcerated -Paracanthosis and percussion pain in the C4-C5, acupuncture revealed hypoesthesia in the skin of the lateral left upper arm -Level 4 muscle strength of the left deltoid and biceps brachii, positive spurling test was and hyporeflexia of left biceps brachii -Visual analogue scale was 7 points
Vitfell-Rasmussen et al. [13]	Severe pain in the right shoulder
McCabe et al. [15]	Painful swelling in the upper right back, tense and tender mass when palpated
Rispoli et al. [16]	-Initially: growing mass at the rostral end of the incision done at the upper thoracic intramedullary neurinoma. -After: increased local pain and pressure, no neurological deficits -A growing mass superior to the surgical lesion (upper thoracic intramedullary neurinoma)
Satsuma et al. [21]	-6-month history of a gradually enlarging mass in the posterior aspect of the right shoulder -On physical examination, an 18-cm posterior longitudinal scar from C4 to T9 was noted over the cervicothoracic spine -The patient had mild scoliosis in the thoracolumbar spine -The right scapula was 3 cm higher than the left -Range of movement of the right shoulder was limited in forward elevation and abduction -A subcutaneous mass measuring 6x5x3 cm was palpated beneath the scar, medial to the medial border of the right scapula -The mass was protuberant, firm, nontender, smooth surfaced and well defined with normal overlying skin
Gupta et al. [18]	-Initially: asymptomatic -9 months later: local pain and restriction of neck movements with palpable lump
Pallarés et al. [22]	A huge and firm mass with associated restriction of movement
Shanmugasundaram et al. [23]	15x10cm swelling in the right scapular region of 4 years duration with recent onset of pain and limitation of shoulder movements
	Pain and swelling in the left axilla of 6 months duration
Cohen et al. [28]	Large subcutaneous firm mass in the upper back, measuring 15 × 10 cm, adherent to its surroundings, with no apparent pathological vasculature or satellite lesions.
Ataizi et al. [25]	Painful left sided immobile mass adhered to the surrounding tissue
Goldstein et al. [31]	Acute neck and bilateral shoulder pain, with normal neurological exam, including motor, sensory and reflex testing
Mubeen et al. [33]	Firm subcutaneous mass in the upper back adherent to its surroundings.
Bekci et al. [27]	Lower back pain, nonmobile, palpable mass in the middle of posterior low thoracic region.
Furlan et al. [30]	Low back pain, palpable, soft, nonmobile mass in the right paraspinal lumbar region
Nikolaou et al. [35]	- Firm, round mass, tender in palpation, in the right parasacral area, fixed to the deep tissues, with no overlying skin changes. - Neurologic examination revealed normal motor strength, sensation and reflexes in her lower extremities.
Pawluś et al. [36]	- A fixed, firm mass palpable, below the right scapula and on the neck and in the right lumbar region. - Gradually enlarging, non-tender and firm in consistency - The range of movement in the patient's right arm was limited.

Barnouti et al. [26]	Left neck mass for one year, which was gradually increasing in size, involving the left postero-lateral aspect of the upper neck, which was hard, non-tender and fixed to the skin and the deep neck structures.
Almarri et al. [24]	-Initially: asymptomatic that later had numbness in the right arm and firm, immobile and non-tender supraclavicular mass
Eksi et al. [29]	- Low back pain and neurological claudication, radiating from the lateral aspects of the legs to dorso-lateral aspects of her feet. - Physical examination no motor weakness and bilateral hypoesthesia in lateral aspects of her thighs
Savy et al. [39]	25-year history of recurrent sciatica in the right leg with absent right ankle jerk
Sahn et al. [38]	Multinodular, firm, non tender and not fixed to underlying structures mass that developed and enlarged for two years
Kim et al. [32]	Back pain and mild numbness from the left buttock to the ankle, no abnormality in motor or neurological examination
Nam et al. [34]	Lower back pain, both buttock pain, and radiating pain in the left leg that were present for 1 month.
Puvanesarajah et al. [37]	-Lower extremity clumsiness, balance, and ambulation difficulty. -Ten months postoperatively (after first surgery), the patient reported moderate pain at the rostral end of the surgical incision. -Initially: NR -Ten months postoperatively after 1st surgery, a mass reported by the patient to have enlarged was found on palpation, just superior to the surgical incision. As the mass continued to grow, the patient suffered from increasing local pain and pressure, but there were no neurological deficits.
Hood et al. [45]	-Painful, slowly enlarging posterior neck mass, which was round, firm, and fixed to the underlying tissue - Neurologically intact with no radicular pain or weakness.
Sevak et al. [49]	-Initially: intractable neck pain and right upper extremity radicular pain. -Two years after: She developed an enlarging left upper back mass beneath the site of the previous surgical incision. -A neurological review of systems was negative for any recent change. -Examination revealed a sizable subcutaneous bulging firm nontender nonerythematous mass beneath cervicothoracic incision.
E Silva et al. [43]	-Painful hard cervical mass in the left neck for 2 years, well adhered to the deep layers, painful on palpation -Restricted lateral rotation movement and left inclination without compromising the left cingulate movement -No evidence of significant restrictions of the left shoulder -During passive mobility, cervical pain irradiated to the left forearm and arm
Mujtaba et al. [47]	Intractable occipital headaches and lower extremity weakness
Hendriks et al. [44]	-Progressive trismus, a swelling in the retromandibular area, loss of sensibility of the maxillary and mandibular branch of the trigeminal nerve had developed. - Relapsing otitis media and weight loss 8.5 kg of body weight within 3 months no fever / night sweats. -Several weeks later, she developed diplopia and torticollis, because diminished vision of the left eye occurred, she was started on prednisone 1 mg/kg/day, her vision deteriorated. -Swelling retromandibular on the left side and a limited mouth opening of only 2 to 3 mm was seen. -Some atrophy of the left masseter muscle was found, suggesting a sustained dysfunction of the trigeminal nerve. -Inspection with the patient under general anesthesia showed a blue lucent swelling of the soft palate and tonsillar. -During muscle relaxation, the mouth opening was 35 mm instead of 2 to 3 mm (involving pterygoid muscles) -Neurological investigation after 4 months of prednisone treatment showed severe loss of function of the left optic nerve with torticollis of the head.
Oweis et al. [48]	18-month history of insidious onset pain in her right shoulder and neck and intermittent tingling in the first three digits of the right hand, full range of motion with intact motor and sensory function of the limb.
Coelho et al. [42]	8 months after surgery, a non-pulsatile and painless paravertebral swelling was observed.
Sonmez et al. [51]	Painful, firm, tender thoracic paraspinous mass for 6 months, palpated just cranial to the upper skin incision.
Zhang et al. [53]	-Progressive neck pain and limitation of activity that had persisted for 3 months and was unresponsive to analgesics. -Physical examination: local tenderness was palpated in the area of C4 and C5 with mild paraspinal muscle spasm. -The range of motion of the neck was limited. -No apparent muscle weakness or sensory loss was detected in the extremities or in the trunk. -Deep tendon reflexes were normal in the upper and lower extremities, and no abnormality in pathologic reflexes
Chung et al. [41]	-Age of 9: 5-week history of headache, vomiting, and ataxia. -Two years later: New-onset intermittent vertical diplopia - Examination revealed right trochlear nerve palsy and bilateral papilledema secondary, lesion was found to be firm and rubbery. -Two years further later: an asymptomatic C-7 vertebral body lesion was discovered. -One year further: Asymptomatic lesion.

Skhiri et al. [50]	-Left cervical mass that had been developing for several months. -On examination, A left latero-cervical mass, posterior, measuring 10 cm in height, axis, of hard consistency -Fixed in relation to superficial and deep planes, without cutaneous inflammation or satellite adenopathy. -A rhinoscopy: anterior, an otoscopy and an endoscopy of the cavum, were normal.
Yogesh et al. [52]	-The second year from the excision of aggressive fibromatosis: 1 year duration of upper back pain, progressive focal thoracic kyphosis, with spastic paraparesis in the lower limbs and wheelchair bound -Neurological examination demonstrated grade 2-3 strength in both lower limbs and hypoesthesia was noted below T5
Bouatay et al. [40]	Right neck mass for 9 months that was increasing in size, hard swelling fixed to the skin and deep neck structures
Lechien et al. [46]	-12-month history of decubitus dyspnea in the context of painless mass of the right side of the neck -The ear, nose and throat examination revealed a very painful mass located in the right side of the posterior cervical area next to the cervical scar of a previous biopsy -The mobilization of the shoulder was very limited with regard to the pain and the mass effect
Bohl et al. [98]	Non tender, fullness in the right upper back and neck
Avinçsal et al. [54]	A 6-month, large, non-tender, hard neck mass that had gradually increased in size and attached to deep neck structures
Klonaris et al. [59]	-Slowly growing, hard, tender, palpable, painful mass at the right lateral neck over the upper half of the right sternocleidomastoid -Painful head rotation and right upper extremity extension with no other sensory deficits or motion restrictions
Eichberg et al. [55]	Neck mass regrowth
Pyon et al. [64]	Stabbing pain in her donor site just under the linear scar with palpable mass
Pan et al. [63]	-Rapidly progressive mass for 8 months with waist pain over the next 1 month -Normal limb activity and promontory on the skin of the sacrococcygeal region was seen -Slightly tender, soft, and rounded mass measuring approximately 5x3x1cm
Fuji et al. [56]	-Clumsiness of the hand and gait disturbance including hand paresthesia for 1 year -Decreased finger dexterity and feet paresthesia for 8 months -Reduced finger dexterity, deficiency of finger extension, a slow repetition of grip, and a slow release of the fingers in association with an exaggerated wrist flexion. -Able to walk without a cane but has spastic gait, exaggerated deep tendon reflexes in upper and lower extremities. -Positive Hoffmann and Babinski reflexes
Oberthaler et al. [62]	Back Pain and fatigability with hemi lateral lumbar bulging
Wyler et al. [66]	Subcutaneous firm tender, non-pulsatile mass, decreased sensory perception of the 4th and 5th fingers of the left hand.
Lau et al. [99]	-Gradual progressive neck pain radiating into the right trapezius muscle / numbness and paresthesia's in her right arm extending into her hands and fingers/ twitching in her right fourth and fifth fingers / no deterioration in writing or fine motor skills. -She had full range of motion of the cervical spine, but cervical flexion and extension elicited pain. -Motor examination was normal.
	-4 months of neck pain and paresthesia in the left arm. -On physical examination, she was found to have left-sided Horner syndrome and a large, palpable, firm fixed mass in the left side of her neck, extending into the thoracic inlet. -Neurological examination showed left hand weakness and numbness. -She also presented with increased venous engorgement in the left precordium and diminished left radial pulses. -Her breath sounds were significantly diminished throughout the lung field.
Shakur et al. [65]	-Initially in 1990: her only symptom at that time was mild neck pain. She did not have any weakness or numbness. -In 2007: progression of her neck pain with a radiating pain into her arms. She had associated paresthesia but no weakness. -The patient had a progressive kyphotic deformity at the cervicothoracic junction with associated cord compression. -In 2007: On examination, she had full strength in all extremities, her reflexes were within normal limits and her gait was intact.
	-Mid-thoracic back pain with intermittent numbness and tingling in both legs but no weakness. -On examination, she demonstrated full strength, intact sensation, normal reflexes, and normal gait.
Juergens et al. [58]	-Acute history of back pain while horseback riding in December 1999. -History of dorsalgia for 1 year and had been treated conservatively with physiotherapy and oral analgesia -The pain has been increasing during the previous 3 weeks prior to presentation -During physical examination, the patient could only lie on her side or in prone position due to excessive pain -No abnormalities during examination of the vascular status, motor function, and sensitivity of the lower extremities
Maurer et al. [60]	A growing swelling around the operation scar and another lump further proximally in the region of the left scapula
Gerek et al. [57]	-Neck mass in the site of a previous trauma, aesthetically unappealing -Examination revealed a mobile, nontender, firm mass on the left side of the neck occupying the left supraclavicular triangle approximately 12x15cm in diameter with no other otolaryngologic findings or any neurologic abnormalities
Hara et al. [17]	Pain in left shoulder for the past 2 years
McCall et al. [61]	Disequilibrium
Yan et al. [67]	Mild recurrent back pain

Table 2. Radiological and Pathological Features of the Reported Cases in the Literature

Reference	Radiological and Pathological Feature
Our case	Postoperative Back liposuction was done 6 months prior to the clinical presentation Mass in the right superficial perimuscular fascia in the right paramedian area True-cut biopsy showed benign fibrous tissue with a preoperative diagnosis of Paraspinal desmoid fibromatosis Treated with wide local mass excision
Lee et al. [7]	Mass in the left multifidus muscle Incisional biopsy showed intramuscular schwannoma and Excisional biopsy showed desmoid fibromatosis Treated with surgical excision
Neffa et al. [8]	Unknown genetic cause Presented with mass in the left scapular joint, diagnosed preoperatively with desmoid tumor At 20 years treated with surgical excision followed by polychemotherapy. From 21 to 25 years, she underwent surgery for tumor mass-reduction and started on multiple different chemotherapy regimens which were unsuccessful. At 26 years, treatment shifted to a palliative and radiotherapy and at 27 years, she died due to tumor progression
Lacayo et al. [6]	Case 1: Postoperative cervical spine surgery 5 years prior to the clinical presentation Mass located in right cervical region Preoperative tissue sampling for diagnosis of desmoid tumor Radiation therapy as definitive treatment, was lost to follow-up
	Case 2: Postoperative cervical spine surgery one year prior to the clinical presentation Mass located in left cervical region Preoperative tissue sampling for diagnosis of desmoid tumor Treated with wide surgical excision, slow enlarging recurrence 16 months post operatively treated by external beam radiotherapy. At 16-month postoperative follow up, small residual was discovered locally above the carotid artery in CT scan. Close follow up for 2 years in our clinic with radiographic studies, which revealed significant regression of the tumor
	Case 3: Postoperative cervical spine surgery 4 years prior to the clinical presentation Mass located in right cervical region Preoperative tissue sampling for diagnosis and all biopsies showed desmoid tumor Treated with excision of the mass, no improvement in the pain. So, 8 months after the last operation date, it was decided to explore the donor site with tumor resection. Follow up MRI at 12 months post-surgery revealed no sign of recurrence
	Case 4: Postoperative cervical spine surgery 4 years prior to the clinical presentation Mass located in left cervical region Preoperative tissue sampling for diagnosis and all biopsies showed desmoid tumor Treated with mass surgical excision, uncomplicated recovery and was free of disease at 1 year postoperative follow up
Alherabi et al. [5]	Mass located in left postero-lateral aspect of the upper neck 2 non-conclusive fine needle aspirates (FNA), a biopsy was taken and was consistent with fibromatosis Treated with surgical excision and no recurrence
Daneyemez et al. [9]	Mass located in 4th cervical spine Biopsy was not performed, the differential diagnosis was non-ossifying fibroma, giant cell tumor and fibrous dysplasia before treatment Treated surgically, treatment with etodolac and exemestane in March 2013 In august 2013, the MRI showed stable disease, but due to side effects from exemestane, the started on letrozole daily. In June 2014, a Whipple procedure performed. December 2014 began treatment with sorafenib. MRI in April 2015 showed stable disease, but increasing pain, so higher dose of sorafenib. In February 2016, she received electrochemotherapy under general anesthesia with bleomycin according to ESOPE protocol. The patient received a second treatment with electrochemotherapy in June 2016. The patient is now experiencing a good quality of life, is no longer on pain medications and there has been no sign of progression after treatment
Remedios et al. [10]	Caused due to instrumentation to the back, post lumbar epidural analgesia for bilateral lower limb surgery 2 weeks prior to presentation Mass located in right paraspinal area at the level of L4-L5 Muscle biopsy showed low- grade sclerosing myofibroblastic proliferation without myonecrosis or myositis Ultrasound showed resolving hematoma or collection MR showed epidural related infection or local anesthetic induced myositis Muscle biopsy showed right paraspinal desmoid type fibromatosis Treated with oral pazopanib
Schlag et al. [11]	Postoperative, C5-C7 laminectomy/instrumental fusion 2 years prior to the clinical presentation Located at posterolateral right neck between the trapezius muscle and splenius cap Diagnosed with desmoid fibromatosis Treated with surgical excision, no recurrence within 12 months postoperatively

Kenning et al. [12]	Initially: A total colectomy and ileostomy for adenomatous polyposis coli 4 years prior to her initial presentation An isolated right sided foramen magnum mass, diagnosed as Meningioma, treated with surgery and recurred 2 years after: A history of meningioma resection at right posterior triangle of the neck at her previous surgical site Biopsy was done, diagnosed as Neurofibroma. Treated with surgical excision with no recurrence of the second mass.
Luo et al. [20]	Mass located in the left side of the head and neck, invading the left paravertebral region of C2-6 and the left intervertebral foramen of C4-5 CT guided biopsy showed no neoplasm Dumbbell tumor, with a high probability of being a neurogenic tumor Treated with surgical resection, local radiotherapy and chemotherapy. No recurrence one year after the operation and no nerve injury
Vitfell-Rasmussen et al. [13]	Colectomy Mass located in right shoulder in close relation to the scapula Biopsy showed aggressive fibromatosis Treated with surgical without recurrence 6 months postoperatively
McCabe et al. [15]	Postoperative, instrumental correction of scoliosis was done three years prior to the clinical presentation Mass located in right paraspinal area at the level of C6-T3 Biopsy showed Benign fibroblastic/ myofibroblastic spindle cells with mostly a fascicular architecture with diagnosis of extra Abdominal desmoid tumor Treated with surgical resection
Rispoli et al. [16]	Postoperative, neurosurgical procedure of D2-D3 laminectomy with removal of intramedullary neurinoma at the upper thoracic Mass located at the right subcutaneous tissues and paraspinal muscles at the T1-T3 levels Treated with surgical excision, with no recurrence at two years post-operatively.
Satsuma et al. [21]	Postoperative, Woodward's procedure (detachment and caudal relocation of the insertions of the trapezius and rhomboid) due to Sprengel's deformity of the right shoulder at 1 year of age. Mass located at posterior aspect of the right shoulder Treated with chemotherapy between September 1980 and February 1981, followed by surgical resection and continued chemotherapy. The patient was free of recurrence or metastasis 4 months after completion of chemotherapy and 19 months after surgery, but had local recurrence at 21 months
Gupta et al. [18]	Postoperative, resection of a fibrous meningioma Mass located in right paravertebral from C1 to C3 CT-guided biopsy showed typical conventional pattern of fibromatosis Initially active surveillance, after 9 months of surgical removal Recurrence of a paraspinal lesion of a similar morphology after a year at a different location from the level of C3 to D1
Pallarés et al. [22]	Traumatic, with a background of a self- inflicted injury with a knife 4 years prior Mass located in posterior right lateral area of the neck Fine-needle aspiration showed spindle- shaped cell tumor without evidence of atypia diagnosed as fibromatosis Treated with surgical resection and postoperative radiotherapy with no recurrence at 6 months postoperatively on MRI
Shanmugasundaram et al. [23]	Case 1: Located at right periscapular muscles True-cut biopsy showed fibromatosis Treated with surgical excision and chemotherapy with no recurrence
	Case 2: Located at left periscapular muscles Open biopsy done showed fibromatosis Treated with surgical excision The patient developed a recurrence after 9 months of surgery in the axilla over the axillary vessels without infiltration and underwent re-excision of the lesion, adjuvant radiotherapy and tamoxifen for 5 years
Cohen et al. [28]	Post traumatic injury to the back, with unremarkable past medical history. Motor vehicle accident 3 years back sustaining a brain concussion, a fracture of the right lamina of the C-6 vertebra and comminuted fractures of the left radius, ulna and femur. Mass located in the left paraspinal region beneath the trapezium muscle, compressing the paraspinal muscles medially. Core needle biopsy showed desmoid tumor. Initially treated with chemotherapy and surgery, no recurrence.
Ataizi et al. [25]	Postoperative, compression of the 11th thoracic vertebral body was experienced 2.5 years prior to the clinical presentation Located at left dorsolumbar at the level of T11-L2 Soft tissue mass Treated with extended surgical resection

Goldstein et al. [31]	Postoperative, history of C5-C7 anterior cervical decompression and fusions Paraspinal mass in the posterior elements from C2 to C4 CT guided needle biopsy showed rare spindle cells, suggestive of a spindle cell neoplasm Initially, conservatively using NSAIDs followed by surgical resection.
Mubeen et al. [33]	Trauma to the back, one year ago. Mass involves the subcutaneous tissue planes of back at the level of T5-T8 with possible infiltration into erector spinae muscle. Fine needle aspiration cytology showed cellular smear of poorly cohesive cells with bland spindle cell nuclei. Features were compatible with the diagnosis of benign fibroblastic lesion possibly fibromatosis. Treated with surgical excision
Bekci et al. [27]	Well defined mass between T9-L1 vertebrae levels originating from trapezius muscle in the neighborhood of thoracal spinous processes. Fine-needle aspiration biopsy showed desmoid type fibromatosis. Treated with surgical excision
Furlan et al. [30]	No prior surgery Mass at the right paraspinal region Fine-needle aspiration revealed a hypocellular spindle-cell neoplasm with a vague fascicular arrangement, set in a collagenous stroma, diagnosed as desmoid type Fibromatosis
Nikolaou et al. [35]	No history of trauma. Subcutaneous fat of the right paravertebral region Treated with wide excision of the lesion, no signs of recurrence after clinical examination and MRI.
Pawluś et al. [36]	Between muscles in the posterolateral aspect of the right part of the body, extending from the neck through the thoracic wall to the right lumbar region. Incisional biopsy from a visible mass showed thoracic wall tumor, highly suggestive of malignancy. Treated with chemotherapy
Barnouti et al. [26]	The left postero-lateral aspect of the upper neck. After 2 non-conclusive FNA, a biopsy was taken and was consistent with fibromatosis. Treated with surgical excision, above the carotid artery 16 months postoperative.
Almarri et al. [24]	No history of trauma Mass at right supraclavicular region Core needle biopsy showed desmoid fibromatosis Treated with surgical excision, no signs of recurrent or residual disease 2 years postoperatively
Eksi et al. [29]	Spinal canal stenosis between L2-L3 and L3-L4 disc levels and intervertebral disc herniations at L2-L5 levels. Ill-defined mass in the left longissimus muscle An open biopsy was done to achieve the diagnosis. The mass was hard and whitish in color Frozen section biopsy result was indecisive about malignancy. Treated with surgical resection of the entire mass, and radiotherapy, postoperatively. 6 months postoperatively (first surgery) the tumor recurred. The mass never reoccurred but fluid collections occurred after 2nd surgery was resected turned out to be soft tissue mass. 6 months after the removal of the mass, no recurrence.
Savy et al. [39]	History of sciatica Mass located at right part of S1 extending up to L5 2 trephine biopsies and 1 surgical biopsy showed desmoplastic fibroma with an aggressive pattern
Sahn et al. [38]	History of osteopenia Mass on the posterior neck Deep incisional biopsy diagnosed with fibromatosis Treated with surgical excision, no recurrence
Kim et al. [32]	Traumatic, previous motor vehicle accident, 1 year and 5 months ago. Mass at left L3-4 facet joint. The patient underwent bone biopsy, and pathologically, the tumor was diagnosed as a desmoid tumor (soft tissue tumor) Treated with surgical excision
Nam et al. [34]	Traumatic, traffic accident as a passenger 1 year ago, but he had no history of spine surgery. He had undergone nerve blocks several times for the treatment of pain. Mass at left L3-4 paraspinal area Treated with surgical excision, no recurrence 10 months postoperatively.
Puvanesarajah et al. [37]	Ten months postoperatively (1st surgery) 2nd mass postoperatively treated with embolization and resection of the back. -2nd mass within the right subcutaneous tissues and paraspinal muscles at the T1–T3 levels. The mass was located superficial to a pedicle screw at the rostral end of the fusion construct. -Initially: the patient underwent embolization followed by resection. -2nd mass: En bloc resection. No recurrence after 2 years.

Hood et al. [45]	Paraspinal soft tissue mass extending from the suboccipital region to slightly below, the seventh cervical vertebra. A core needle biopsy was performed that revealed nondiagnostic spindle cells. Incisional biopsy. A longitudinal midline incision was made and carried laterally to circumscribe the tumor. Initially: subtotal resection was performed. Then chemotherapy with Gleevec Two years after: Using a combination of electrocautery and the Cavitron Ultrasonic Aspirator, the tumor was resected from the occiput and posterior arch of C1. Complete tumor resection was performed. Two years postoperatively (after first surgery), tumor recurred.
Sevak et al. [49]	-Initially: C7-T1 right sided nerve sheet tumor. Treated with surgical resection, recurred two years after -Two years after: postoperatively, resection of the mass with laminectomies of C6-T1 and instrumentation of C6-T3 with depuy Mountaineer. Mass located left to the midline of the cervicothoracic spine without evidence of bony involvement extending through the surgical level Preoperative core-needle biopsy of the lesion was consistent with spindle-cell proliferation, diagnosed as fibromatosis (Sarcoma) Treated with surgical resection, no recurrence.
E Silva et al. [43]	Use of estrogen and progesterone-based contraceptives for 10 years, no history of radiation exposure, traumas, or previous treatments for any malignant disease Mass located in left cervical region, laterally close to the left paravertebral neck space and supraclavicular region Complete tumor surgical resection with tumor free margins, no recurrence in the one year follow up
Mujtaba et al. [47]	Presumed to be caused by iatrogenic trauma/ posttraumatic, resection of an intradural spinal meningioma 6 years prior to the clinical presentation Located at sorsal aspect of the upper thoracic spine from C7-T1 to the inferior portion of T2 Recurrent meningioma Treated with surgical resection
Hendriks et al. [44]	The masticator, parapharyngeal, and prevertebral and paravertebral space on the left with intracranial extension through the orbital fissure by perineural and perivascular spread. Bone marrow changes were present in the mandible and clivus. Wide biopsies were taken from a lucen swelling of the soft palate and tonsillar areas, Malignancy was still strongly suspected New biopsies were performed, no diagnostic material was obtained, no signs of infection, no signs of actinomyces or yeast were found. New tissues for biopsy were taken even deeper into the retromaxillary area and a left tonsillectomy was performed. 15 months after initial presentation, a biopsy was taken and showed malignant or fibrous tumor, Infection, Autoimmune disease (rheumatology), and desmoid type fibromatosis (15 months after). Treated with radiotherapy, Corticosteroid treatment was tapered off after cessation of radiotherapy. Follow up showed intermittent progressive increase in the size of the tumor.
Oweis et al. [48]	Along the posterior cortex of the right humerus proximal metaphysis. Ultrasound-guided core biopsy of the lesion revealing scar tissue and skeletal muscle, non-diagnostic. Treated with surgical excision of the mass, no recurrence
Coelho et al. [42]	Neurosurgery treatment for right T12-root neuroma after progressive paraparesis. Mass at paravertebral Treated with surgical excision, no recurrence after 3 years.
Sonmez et al. [51]	Postoperative, history of lumbar disc surgery four years ago. Three years after disc surgery, she underwent a T10-L1 laminectomy for the resection of a spinal schwannoma Mass is 3 cm above the scar of a previous skin incision on the back Treated with surgical excision
Zhang et al. [53]	Unremarkable history Mass at vertebral bodies of C4 C5 Frozen sections intraoperatively diagnosed as desmoid fibromatosis Treated with tumor resection, no recurrence in 3 years postoperatively.
Chung et al. [41]	-Age of 9: A right posterior Fossa lesion extending from 4th ventricle to the right cerebellopontine angle. Treated with surgical excision, following surgery, the child underwent radiotherapy and then chemotherapy according to the St. Jude protocol. -Two years later: surgical excision of the previous mass. Extra axial lesion compressing the parietal superior sagittal sinus Treated with completely excised via a midline craniotomy, recurred -Two years further later: excision of the previous two masses C-7 vertebral body lesion Tumor excision, with recurrence of asymptomatic lesion -One year further: excision of the previous 3 masses. right frontal convexity extradural lesion.

Skhiri et al. [50]	Chronic dialysis, internal jugular catheter placement 7 months earlier. (Two occasions- six months gap) Mass at the laterocervical level of C1 At C7. A biopsy of this mass was then removed, showing a desmoid tumor. Treated with surgical excision.
Yogesh et al. [52]	Recurrence, thoracic laminectomy of T2-T5 and excision of aggressive fibromatosis involving the spinous process 1 year prior to the recurrence of the tumor Mass at posterior elements of T2-T5 and paravertebral soft tissues Recurrence of aggressive fibromatosis Treated with surgical excision, external radiotherapy, and physiotherapy. At 2 years follow up, MRI showed no evidence of tumor recurrence
Bouatay et al. [40]	Mass at posterolateral region of the neck, extending from the retro auricular region to the suprasternal notch Biopsy showed demonstrated fibromatosis Treated with surgical excision, no local recurrence on physical examination and CT
Lechien et al. [46]	Lack of previous surgical trauma Mass at posterior cervical area Fine needle aspiration biopsy results were noncontributory Treated with complementary radiation for the residual tissue, the 4 years follow up was unremarkable
Bohl et al. [98]	Postoperatively, surgical deformity correction of adult idiopathic and degenerative scoliosis with progression secondary to multiple sclerosis two years prior to the clinical presentation Mass at right upper back and neck Core needle biopsy showed a spindle cell proliferation consistent with fibromatosis or desmoid tumor Treated with surgical resection
Avinçsal et al. [54]	Mass at posterolateral side of the lower neck Fine needle aspiration was inconclusive Preoperative incisional biopsy showed fibromatosis Treated with wide excision of the tumor. The recurrent lesion 3months postoperatively was treated by radiotherapy. At his 3-month postoperative MRI follow up exam, a small residual tumor was detected in the scalenus medius muscle. For the last 7 years, radiographic studies have shown no evidence of tumor recurrence
Klonaris et al. [59]	Past medical history included a resected desmoid tumor with free surgical margins from the same region six months ago in another center Mass at right lateral neck at the cephalic part of the sternocleidomastoid Local tumor recurrence following surgery Treated with surgical excision. On the 3-month postoperative follow up, no clinical signs of tumor recurrence were noted. On the 6 month follow up, local pain and swelling indicated new tumor recurrence which was confirmed by MRI and treated with 54 Gy of external beam radiotherapy over 6 weeks. No evidence of after 12 months of follow up
Eichberg et al. [55]	Postoperatively, left cervical neurofibroma resection two years prior to the clinical presentation Treated with surgical excision At eight years postoperative follow up, cervical spine MRI showed no tumor recurrence
Pyon et al. [64]	Postoperatively, breast reconstruction with an extended latissimus dorsi flap immediately following a total mastectomy for left breast cancer 2 years prior to the clinical presentation Mass at sonor site of an extended latissimus dorsi flap Excisional biopsy showed chronic inflammation with dense fibrosis, diagnosed with desmoid tumor Treated with resection and local radiation, no recurrence
Pan et al. [63]	No history of trauma, pain or discharge related to the swelling Mass at sacrococcygeal region Postoperative pathology diagnosed the resected specimen as aggressive fibromatosis Treated with surgical resection, recurrence of cervical mass in the same location.
Fuji et al. [56]	Located in lamina, facets, and spinous process of the axis invading the spinal canal at C2 level Biopsy was carried out posteriorly showing benign fibroblastic tumor -Initially: 2-staged operation: was proposed it was believed that chemotherapy would be indicated before anterior surgery. The anterior stage was postponed, but the patient refused chemotherapy. Instead, she chose to try herbal medicines. -Six months postoperatively, there was a massive growth of the lesion, a spinal column resection was precluded. Subsequently underwent a surgical resection. Six months postoperatively after first surgery. After second surgery, the residual mass was causing discomfort.
Oberthaler et al. [62]	Soft tissue tumor Surgical treatment, no recurrence
Wyler et al. [66]	Postoperative, elbow dislocation with ulnar nerve transposition, Neurolysis, selective C7, C8 and T1 blocks Mass at left cervical hemilaminectomy, left side of the midline scar and extending into the paraspinous musculature laterally beyond the laminectomy site. In 2007: surgical treatment with no recurrence

Lau et al. [99]	Case 1: C6–T2 paraspinous mass with invasion into rib heads and vertebral bodies. 6 months postoperatively: mass extended without any dissectible planes between the tumor and the nerves. After 2nd surgery: residual mass was in the craniocaudal extent and at the level of the aortic arch. Two separate biopsy samples were obtained, showing a spindle-cell neoplasm not otherwise specified, malignancy could not be ruled out. Treatment was surgical, with no recurrence.
	Case 2: Mass extending through the thoracic inlet; the tumor originated from the C-4/5 vertebral bodies. During surgery: tumor extended into the thoracic inlet and occupied the entire left hemithorax, and displaced vital structures and extended completely across the midline. Biopsy of the mass confirmed that the mass was a desmoid fibromatosis Surgical treatment, follow up cervical CT was consistent with the lack of recurrence at the end of 4 years after surgery
Shakur et al. [65]	Case 1: History of chronic obstructive pulmonary disease was found to have a cervical lesion during a workup of a whiplash injury in 1990. -In 1990: cervical lesion, the patient underwent a biopsy of this lesion, which was reportedly normal. -In 2007: a large enhancing mass extending from C5 to T1 with destruction of the posterior elements. The lesion effaced the cord dorsally at the cervicothoracic junction. Intraoperative frozen sections were concerning for a spindle cell tumor or osteochondroma / sarcoma. Surgical treatment done
	Case 2: One year previously, she had a similar pain and underwent an excisional biopsy of a spinal mass at another institution. Paraspinal soft tissue mass at T9–T10, extending through and widening the neural foramen. Biopsy of spinal mass one year prior to presentation, reported as normal. Initial frozen specimen was consistent with neurofibroma. Surgical treatment, after non-oncological resection of both tumors, a subsequent MRI showed recurrence only in the scar; positron emission tomography failed to elucidate the local situation
Juergens et al. [58]	Mass at T9-T11 of the thoracic spine CT guided core biopsy was performed using a left paravertebral dorsal access, diagnosed with desmoplastic fibroma Surgical treatment, 6 months after the operation, there is no sign of local recurrence
Maurer et al. [60]	Postoperatively, surgical stabilization of a spine injury 1 year prior to the clinical presentation Mass at first lumbar vertebra and at the region of the left scapula The first mass in the scar appeared to be a hematoma sonographically and the second one a solid tumor Conservative treatment, followed by radiation therapy
Gerek et al. [57]	Post Traumatically, external trauma to the left side of the neck 5 years prior to the clinical presentation Mass at left neck/supraclavicular triangle Surgical treatment
Hara et al. [17]	2 years prior had abnormality on chest x-ray Mass at left lung apex CT- guided percutaneous needle showed desmoid tumor Radiotherapy, no evidence of local recurrence 32 months following the conclusion of radiotherapy
McCall et al. [61]	Radiation therapy postoperatively, resection of cerebellar astrocytoma at age 13 Mass at the back of the head at the site of a previous suboccipital craniectomy Desmoid tumor Surgical treatment
Yan et al. [67]	Postoperative, L3-L5 decompression and posterior spinal fusion 2 years prior to the clinical presentation Mass located in right paraspinous soft tissue in the level of L1-L2 Lumbar spine MRI showed chronic calcified hematoma CT- abdomen pelvis showed soft tissue mass CT- guided biopsy showed desmoid fibromatosis Conservative management
Goepfert et al. [75]	Recurrence, history of surgical resections at 2,3 and 7 months of age for a rapidly growing F-1-DT Mass at lower right quadrant of the neck fixed to the strap and sternocleidomastoid muscles, the trachea, and the manubrium of the sternum Desmoid tumor Tumor resection was done, postoperative treatment consisted of tamoxifen and diclofenac.
Chu et al. [73]	Mass at paraspinal muscles bilaterally extending from the posterior cervical neck to the upper back Surgical treatment with symptoms persisted, and the swelling recurred 6 months later. The tumor recurred multiple times in the following years 1968-1971 and was treated by surgical excision. Surgical amputation was done, no recurrence of tumor since amputation.

Scheer et al. [89]	There was no previous history of back pain or injury Mass at left side of L3 Biopsy of the lamina and body of L3 was carried out on September 1960, through a posterior approach Surgical treatment with no recurrence
Lackner et al. [80]	Patient 4: Subscapular mass Surgical and Radiation therapy with no recurrence
Barber et al. [72]	Case 1: mass at humerus at deltoid insertion. After reoccurrence the probability of a "desmoid tumor" was suggested Surgical and radiation therapy done
Hardes et al. [76]	Mass at 10th thoracic vertebrae and into the left lamina CT-guided core biopsy showed desmoplastic fibroma Surgical treatment
Shindle et al. [90]	No recent epidemic and only ashes of polio Mass at left posterior thoracic cavity extending from the carina to the level of the left kidney and eroding the vertebral bodies Intraoperative specimens showed neurofibroma Surgical treatment
Niv et al. [85]	No history except ectopic pregnancy Mass at mid anterior border of left trapezius muscle Aggressive fibromatosis Surgical treatment with no recurrence
Lynch et al. [83]	Postoperative, T9-11 laminectomy for resection for intraspinal meningioma Mass at T11 Fibromatosis Surgical treatment, no evidence of local recurrence
Garant at al. [74]	Minor neck injury Mass at right prevertebral space of the infrahyoid portion of the neck Fine needle aspiration showed fibromatosis Surgical treatment
Aliyu et al [70]	The left posterior-lateral aspect of the neck, extending from below the occiput to just above the scapular vertically; and from the cervical spinous processes to the posterior border of the left sternocleidomastoid muscle, and attached to the underlying structures, but not the skin Fine-needle aspiration showed a few foam cells, fibrous stromal fragments, adipocytes, and hemorrhage only. Surgical treatment with adjuvant radiation therapy and no recurrence 8 months postoperatively
Nishio et al. [84]	There was no history of antecedent trauma Mass at left axillary region An open biopsy was performed, differentials showed neurofibroma, desmoid-type fibromatosis, fibrosarcoma, and low-grade fibromyxoid sarcoma. Surgical treatment, no recurrence for 17 months postoperatively
Rubio et al. [88]	Mass at back lumbar spine Biopsy was done Steroids and chemotherapy
Pereyo et al. [87]	Overlying acromioclavicular joint 2 punch biopsies and 1 incisional biopsy showed extra abdominal desmoid tumor Surgical treatment with no recurrence
Kriss et al. [79]	Mass at mid posterior cervical region Frozen section biopsy showed musculoaponeurotic fibromatosis Surgical, radiotherapy, and chemotherapy was done, with recurrence 22 years post the traumatic amputation
Lidov et al. [82]	Mass at left side of the neck Open biopsy showed Fibromatosis Surgical and palliative radiation with recurrence and patient died
Kamby et al. [78]	Case 1: Postoperative, history of traumatic amputation Mass at left deltoid region Mediastinoscopic biopsy showed aggressive fibromatosis No recurrence
	Case 2: Postoperative, history of recurrent aggressive fibromatosis Mass at right scapula Aggressive fibromatosis Chemotherapy and recurrance

Humar et al. [77]	Case 1: Mass at mid posterior neck Extra abdominal fibromatosis Surgical treatment, recurred 6 months postoperatively
	Case 2: History of breech with forceps delivery Mass at sternocleidomastoid muscles Excisional biopsy showed Fibromatosis colli Surgical treatment, and postoperative radiation was received.
	Case 3: Postoperative, history of neck mass resection Mass at anterior neck extending posteriorly to left scapula Infantile fibromatosis The tumor recurred twice; all 3 masses were surgically managed
	Case 4: Postoperative, history of extra abdominal fibromatosis Posterior neck mass Extra abdominal fibromatosis Surgical treatment
Sobani et al. [91]	Initially: right side of the neck 2nd mass: right posterior neck Fine needle aspiration cytology showed spindle shaped cells with elongated vesicular nuclei and pink cytoplasm admixed with pink fibro collagenous material suggestive of fibromatosis 3rdmass: right posterior neck
Palacios al. [86]	Right side of the neck
Lessow al. [81]	Mass at left posterior neck Incisional biopsy showed benign-appearing fibrous neoplasm without diagnosis
Atahan et al. [71]	Recurrence, history of two biopsies in February and April 1982, which were both reported as fibromatosis, followed in May 1982 by partial scapulectomy and total excision of the mass. Mass at left shoulder Aggressive fibromatosis Surgical treatment

(n=14), CT-guided needle biopsy (n=7), ultrasound guided needle biopsy (n=1), trephine (bone) biopsy (n=2), and muscle biopsy (n=1) were the other used modalities to obtain biopsies. No data was available for the rest of the cases.

Histologically, the conventional pattern of desmoid tumors is the commonest. It is characterized by the proliferation of benign, elongated, and slender fibroblastic spindle cells in a collagenous stroma with parallelly arranged thin-walled blood vessels [6, 18]. Other patterns include hypocellular/ hyalinized pattern, staghorn vessel pattern, keloidal pattern, myxoid pattern, hypercellular pattern, and nodular fasciitis-like pattern. All patterns share the feature of minimal cellular atypia without hyperchromasia or appreciable mitotic activity [18]. The immunohistochemistry profile of desmoid tumors demonstrates positivity for b-catenin, vimentin, ER-B, c-kit, and cathepsin but stains negatively for CD-24, ER alpha, PR, and HER2 with variable staining for smooth-muscle actin [6, 19].

In this literature review, we analyzed the histopathology of all reported paraspinal desmoid tumors. The conventional pattern was found in 84 cases, which were characterized by the proliferation of spindle cells within a collagenous matrix. Additional features included low mitotic rate in 22 cases, absence of tumor necrosis in 7 cases, the lack of significant pleomorphism in 5 cases, and a non-hemorrhagic lesion in one case. Exceptionally, one case reported mild pleomorphism and another case encountered a hemorrhagic lesion. The histopathology data were not recorded in 13 cases. The cells stained invariably positive for SMA, beta-catenin, S100, calponin, Desmin, CD34, vimentin, ki-67, collagen factor XIIIA, elastic tissue, collagen stains, cytokeratin, epithelial membrane antigen, CD-68, CD3 and CD20. On the contrary, they stained invariably negative for ER, PR, CD34, CK AE1/AE3, Desmin, CD117, SMM, S-100, SMA, b-catenin, factor XIIIA, vimentin, EMA, MUC4, CD68, MIB-1, c-kit, CD30, KP1, muscle-specific actin, epithelial membrane antigen, and e-cadherin.

The proper treatment of desmoid tumors requires a multidisciplinary approach. Due to the tumor's rarity, there are no consensus management guidelines to follow [5]. However, the international consensus from the Desmoid Tumor Working Group promotes active surveillance as the primary management plan [5-13, 15-18, 20-67, 70-91]. In that sense, newly diagnosed asymptomatic patients should be initially managed with active surveillance or the "wait and see" approach because studies have proved that a significant percentage of desmoid tumors show long-term stability without treatment. In symptomatic patients, active treatment is required to limit tumor invasion. In such cases, the Desmoid Tumor Working Group promotes surgery as the mainstay of treatment. The aim of surgery is to establish a wide, microscopically negative resection margin (R0). However, if an extensive resection possesses a risk of compromising function or cosmesis, then positive microscopic margins (R1) are acceptable.

Evidence showed that macroscopically positive margins increase the risk of recurrence, whereas there is no difference in the recurrence rate between microscopically positive resection and microscopically negative margins [15]. Adjuvant radiotherapy has been found to reduce the chances of recurrence with variable rates, especially in cases where wide surgical margins are not possible [23]. Radiotherapy may also help to get rid of any residual tumor cells [5]. A recent meta-analysis demonstrated that surgery with adjuvant radiotherapy reduces the rate of failure compared to surgery alone [92]. Some sources contradict the

previous studies by stating that radiation therapy is not recommended for positive microscopic margins as no proven benefits exist [15]. Meanwhile, primary radiation or medical treatment is reserved for cases with rapidly progressive desmoid tumors and nonsurgical candidates. Medications such as anti-estrogenic drugs (tamoxifen and toremifene) may result in the regression of tumor cells due to the association of estrogen levels with the growth of desmoid tumors, especially in intra-abdominal types [5]. Other medications include tyrosine kinase inhibitors (imatinib, sunitinib, pazopanib, sorafenib, and sirolimus), non-steroidal anti-inflammatory drugs (NSAIDs) (meloxicam, indomethacin, sulindac, and celecoxib) and low-dose chemotherapy (doxorubicin, methotrexate, and vinblastine) proved to be effective [23, 92]. Electrochemotherapy is also considered an option. It works by combining bleomycin with local electric pulses to enhance the permeability of cells to bleomycin allowing it to reach its intracellular target more readily. Studies have demonstrated effectiveness of electrochemotherapy in treating subcutaneous/cutaneous tumors, including those large or metastasized tumors [13].

In the present literature review, it was concluded that surgical resection of the tumor is considered the mainstay of treatment, including wide local excision with free margin (n=66), and partial excision (n=21). In some instances, surgery was followed or preceded by adjuvant therapy, such as radiotherapy (n=20), chemotherapy (n=13), physiotherapy (n=1), and tumor embolization (n=1). In other instances, medical management was implemented with some conservative management of the tumor (n=3), corticosteroid use (n=3), and herbal medicine (n=1). Radiotherapy was utilized as a first-line treatment in 5 cases and as palliative treatment in one case. Palliative treatment was opted in 3 cases; one case received only radiotherapy, another one had combined intensity-modified radiotherapy with corticosteroids, and the last case had combined electro-cautery with cavitron-ultrasonic aspiration. Electrochemotherapy produced effective results in one case [5-13, 15-18, 20-67, 70-91].

Local recurrence is common. Approximately 50% of the cases tend to recur within 5 years [93, 94]. Wider surgical resection of desmoid tumors seems to lower the recurrence rates.

Many theories have been postulated to explain the high recurrence rates, one of which suggests that it is difficult to establish clear margins of the tumor due to its extension within muscle fibers [19]. Furthermore, desmoid tumors larger than 5cm are associated with a higher recurrence rate and poorer prognosis [95].

Fibromatosis may cause significant morbidity related to its locally aggressive and infiltrative growth once the tumor invades vital structures, as may happen in intra-abdominal fibromatosis that involves the retroperitoneum or extra-abdominal fibromatosis of the head and neck region [96, 97]. Death from the disease is rare. It has been established that the five-year survival rate is more than 90% [25]. In this literature review, the patients were followed up with MRI (n=26), plain Xray (n=9), CT (n=5), and fluorodeoxyglucose PET (n=1). In our case, we opted to choose MRI follow up every six months.

CONCLUSION

To the best of our knowledge, this is the first report of a post-liposuction paraspinal desmoid fibromatosis worldwide. In addition, it is the first report of a paraspinal desmoid fibromatosis in the Arabian Gulf region. This is the first extensive literature review involving all the reported cases of paraspinal fibromatosis (97 cases). Universal guidelines for the treatment of desmoid fibromatosis are still lacking.

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Abbreviations: APC: adenomatous polyposis coli, FAP: Familial adenomatous polyposis, NSAIDs: non-steroidal anti-inflammatory drugs, MRI: magnetic resonance imaging, SMA: smooth muscle actin, CT: computed tomography, PET: positron emission tomography, ER: estrogen receptor, PR: progesterone receptor.

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