

Desmoplastic Fibroma of the Mandible: A 5 Year Follow-Up

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Desmoplastic Fibroma is a rare benign intraosseous tumor with locally aggressive behavior. The mandible is the most frequent site of involvement. It causes bone destruction with tendency to invade the soft tissues if untreated. It has a high propensity for local recurrence if treated inadequately. We report a case of Desmoplastic Fibroma of the mandible treated with wide margin of excision and immediate rib graft reconstruction, five years follow up revealed no recurrence.

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Desmoplastic Fibroma of the bone is a rare locally aggressive myofibroblastic tumor which is considered the osseous counterpart of the soft tissue fibromatosis¹. This tumor constitutes less than 1% of bone tumors and about 0.3% of all benign osseous tumors, and it usually involves the tibia, scapula, and femur². The mandible represents the fourth most commonly affected site³. It has a predilection to affect the posterior part of the mandible, most often in the ramus-angle region.

Desmoplastic Fibroma of the jaws is most commonly discovered in the second and third decades with the mean age of 15.7 years and with slight female predilection⁴.

We report a case of Desmoplastic Fibroma of the mandible with a 5 year follow-up without any signs of recurrence and discuss the utilization of resorbable hardware in the surgical treatment.

Case Report

A three-years-old Bahraini boy was referred from a health center to the Department of Oral and Maxillofacial Surgery, Salmaniya Medical Complex, Bahrain with a 6 month history of painless rapidly progressive swelling on the left side of the angle of the mandible. There was loosening of teeth and bleeding from the mouth. The child was given a course of antibiotic therapy but remained symptomatic. The father denied any history of trauma to the jaw or abnormal oral habits. The child's medical history was insignificant and no related family history was reported.

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Clinical examination showed a non-tender, firm swelling on the left body-angle region of the mandible. There was an obvious expansion with soft tissue fullness on the left body of the mandible and multiple palpable rubbery non-tender lymph nodes (Figure 1). Intra-oral examination revealed loose lower left first and second deciduous molar teeth (mobility Grade II); there was fullness of the buccal vestibule and bleeding on gentle pressure on the gingiva. Panoramic radiography and Computed Tomography (CT) scans showed 2.5 cm mass on the left side body of the mandible with complete destruction of the buccal and lingual compact bone plates. There was encroachment on the first permanent molar tooth and breach on the lower border of the mandible (Figure 2). The lymph nodes were of non-significant size, less than one cm according to the radiologist.



Figure 1: There Is an Obvious Left Side Mandibular Swelling with Soft Tissue Neck Extension

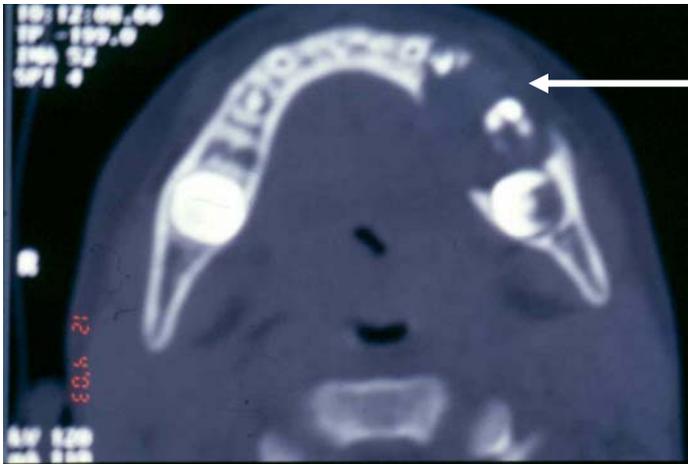


Figure 2 (a)

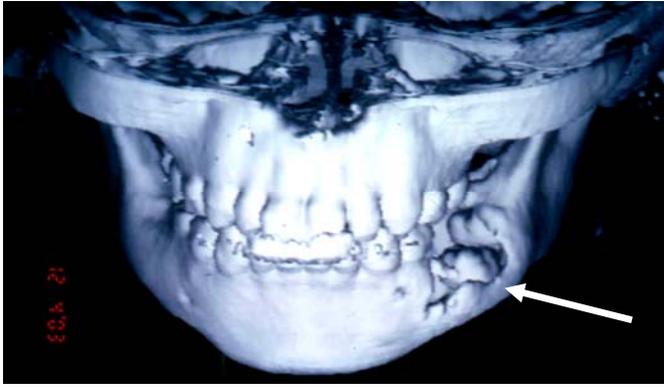


Figure 2 (b)

Figure 2 (a, b): CT scan demonstrating 2.5cm radiolucent mass on the left side of the mandible with resorption of the buccal and lingual plates.

Under general anesthesia, an incisional biopsy of the lesion was obtained and submitted for histology.

Microscopic examination showed spindle cells with fusiform plump nucleus arranged in solid sheets with varying degrees of myxoid changes. Immunohistochemistry showed the following: The tumor cells showed focal reactivity for smooth muscle actin, and strong and diffuse immunoreactivity for vimentin. Tumor cells were nonreactive for S-100 protein and muscle specific actin. Based on clinical, radiographic and histological changes, a diagnosis of Desmoplastic Fibroma was achieved (Figure 3, 4).

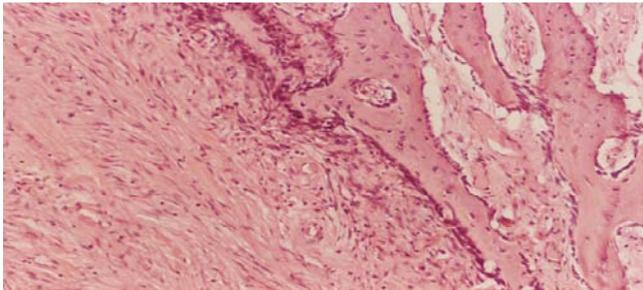


Figure 3 (a): The Spindle Shaped Cells Are Seen Touching the Bony Trabeculae of the Mandible Lined by Osteoblasts

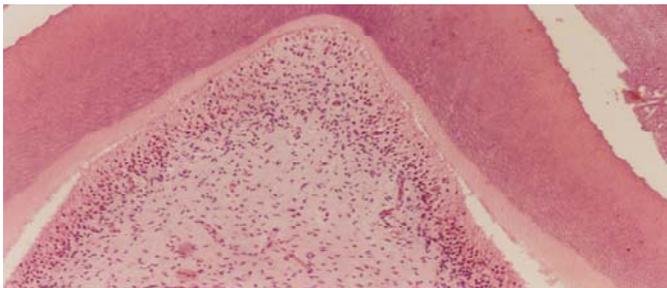


Figure 3 (b): Shows the Lesion Protruding into the Substance of the Tooth

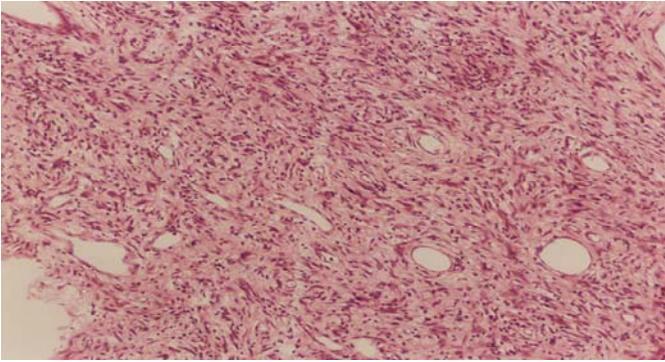


Figure 3 (c): Shows the Peritheliomatous Pattern of the Cells

Figure 3 (a,b,c): Shows a spindle-celled lesion arranged in sheets, perivascular and storiform pattern. A sprinkling of chronic inflammatory cells is noted amidst the neoplastic cells. Mitotic figures are absent (hematoxylin and eosin $\times 100$).

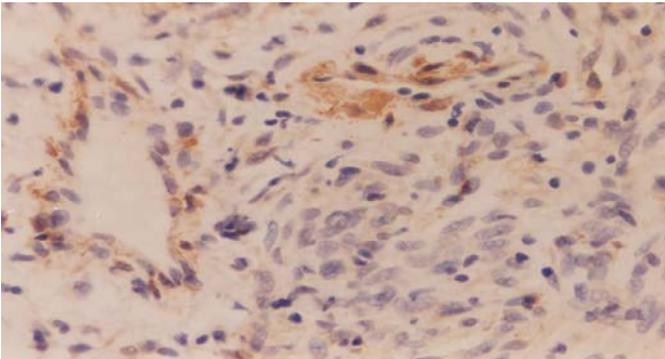


Figure 4: Shows smooth muscle actin (brown color) of the spindle-shaped cells. Staining is stronger around the blood vessels in the pericytes and indicates a myofibroblastic differentiation (immunohistochemistry $\times 100$).

The patient was admitted to the hospital. Under GA and via an extended submandibular neck crease incision the left body as well as the ascending ramus of the mandible was exposed (Figure 5). The tumor was outlined from CT scan and intra-operatively, an en block resection with 1 cm free margin off-tumor was performed (Figure 6). The resected part was reconstructed temporarily to maintain the size of the gap using titanium miniplate osteosynthesis (Figures 7, 8). Through a submammary skin incision, two rib grafts were harvested for reconstruction (Figure 9). The rib graft was split longitudinally, perforated in order to improve its vascular perfusion and fixed using resorbable plates osteosynthesis. This was fixed to both sides of the resected mandible using 4 holes resorbable plates osteosynthesis (Figure 10). The wound was closed in layers and the patient was transferred to the Intensive Care Unit. The patient was hemodynamically stable and was discharged from the hospital after one week.

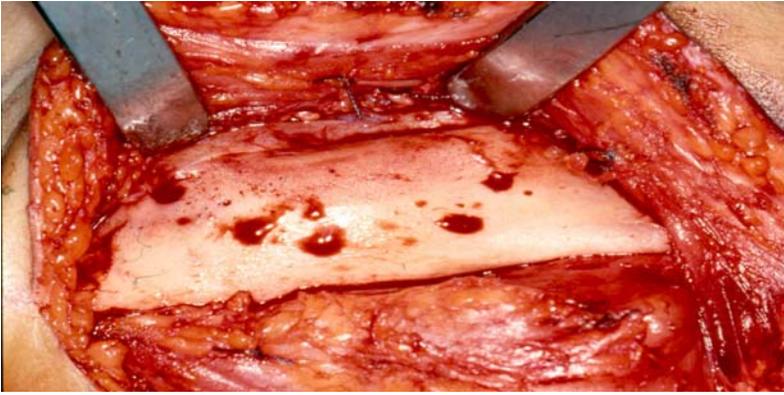


Figure 5: Exposure of the Right Side Mandible via Submandibular Incision

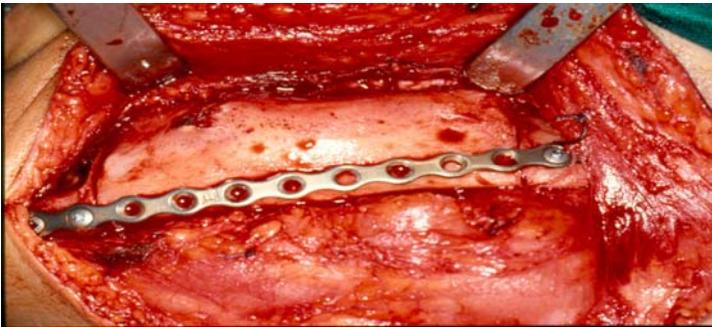


Figure 6: Temporary Fixation of the Mandible Prior to Resection



Figure 7: Temporary Fixation of the Resected Mandible



Figure 8: Gross View of the Resected Surgical Specimen

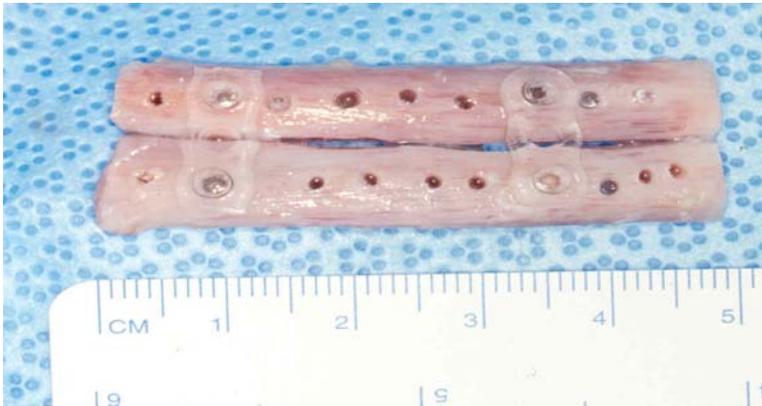


Figure 9: Two Split Rib Graft Perforated and Fixed Using Resorbable Plate Osteosynthesis

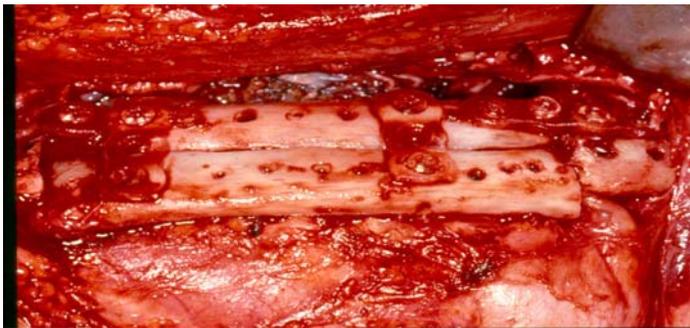


Figure 10: Reconstruction of the Resected Mandible Using Rib Grafts Fixed with Resorbable Plate Osteosynthesis

The child was on regular follow-up in the out-patient clinic for 5 years with yearly radiographic evaluation and showed no clinical or radiographic features of recurrence (Figure 11).



Figure 11 (a)



Figure 11 (b)



Figure 11 (c)

Figure 11 (a,b,c): 5 Years Follow-up Showing Acceptable Facial Symmetry and Absence of Recurrence

Histopathology

Gross features: Resected specimen consisted of part of the body and ramus of the mandible measuring 4.5×2.5×1.5 cm containing canine and deciduous molar teeth. The lateral surface is showing protuberance and unerupted teeth. Cut surface of the mandible showed 2 teeth within its surrounding gelatinous tissues. Further sectioning showed firm white soft tissue of the tumor inside the bone measuring 2×2 cm.

Microscopic Features: The Left side of the mandible showed an ill defined soft tissue neoplasm inside the bony cavity composed of spindle cells with fusiform plump nuclei arranged in solid sheets with storiform pattern and sprinkling of chronic inflammatory cells. In some areas, there was pericytomatous pattern with many compressed blood vessels. Some foci with hyalinization around blood vessels were noted. Foci of stromal hyalinization and myxoid changes were also seen.

Mitotic figures were absent. The periphery of the lesion shows infiltration into the surrounding bone. The anterior as well as the posterior resection margins were tumor free. The resected left side of the mandible shows a neoplasm with characteristic features of an infantile myofibromatosis (Desmoplastic Fibroma).

DISCUSSION

Infantile myofibromatosis (Desmoplastic Fibroma) was formerly known as congenital generalized fibromatosis and classically presents before the age of 2 years. It can occur in a wide age range but the majority in the 3rd decade. Approximately 25% of the affected individuals may have multiple lesions. It has a female predominance and the majority of lesions arise in the skin and superficial soft tissues, especially the head and neck region and trunk. Intraosseous lesions are quite common. In the maxillofacial region, the mandible is the most commonly affected site, but maxillary involvement has been described in the literature⁵. The lesion has more propensity to occur in the posterior part of the mandible namely the ramus-angle region⁶. A small proportion of cases are inherited but whether by an autosomal dominant or recessive mechanism remains unclear⁷. Most of the lesions measure 3-6 cm when detected, the majority of which are well-circumscribed but not encapsulated^{3,4,6}. Patients usually present with a firm painless swelling which is slowly growing without teeth mobility and root resorption^{7,8}. The case we are presenting had a mobile second deciduous molar tooth.

Radiographic features of Desmoplastic Fibroma are often non-specific and may range from well-outlined radiolucent lesion to multilocular radiolucencies with varying similarities to benign cystic lesions of the jaws, benign intraosseous tumors or jaw sarcomas. The margins of the lesion may be well defined to ill-defined border but bone expansion and cortical thinning is often present as a cortical reaction^{8,9}. The bone lesion can invade the cortical bone and erodes the buccal or lingual plates in order to penetrate into the soft tissues. When this

occurs, the accompanying soft tissue mass will pose dilemmas whether it is a true soft tissue extension of the lesion or another isolated soft tissue fibromatosis¹⁰.

At microscope level, Desmoplastic Fibroma shows two cell components, spindle shaped cells with areas of hyalinization, myofibroblastic features and more primitive areas composed of smaller round to spindle cells with round nuclei and eosinophilic cytoplasm. The less differentiated cells are arranged around blood vessels. Both cell types are known to exhibit actin in keeping with their myofibroblastic nature.

Various medical and surgical treatment options were suggested for Desmoplastic Fibroma. These include simple curettage, segmental resection, en block resection, radiotherapy and chemotherapy with or without additional surgical procedure¹⁰⁻¹². The type of surgical approach plays a vital role in the recurrence rate.

The most acceptable and widely used surgical treatment option remains wide resection with a safety margin¹²⁻¹⁴. This usually reduces the recurrence rates significantly. On the other hand, simple curettage or enucleation of the lesion is the least favorable because it is associated with 20%-70% recurrence rate¹⁵⁻¹⁷.

In this case, the treatment employed was wide segmental resection and immediate reconstruction using a rib graft. This played a vital role in the overall recovery and absence of recurrence in the 5 year follow-up period. Furthermore, we have employed resorbable materials in the reconstruction process, which partly reduced the infection rate and the need for second surgery for the removal of the hardware, to our knowledge; this is the first reported case in the literature to employ this treatment method¹⁸.

CONCLUSION

In this case report, we presented a case of Desmoplastic Fibroma in a child three years old who has been treated by wide resection and immediate reconstruction with a rib graft.

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