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Retroperitoneal Solitary Fibrous Tumor

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Solitary fibrous tumor of retroperitoneum is very rare with unpredictable behavior. We present a case of a 67-year-old male with large pelvic, abdominal mass measuring 20x13x7 cm and weighing 2.8 kg. Needle biopsy showed malignant spindle cell tumor. The mass was resected. The hypoglycemia and lower limb edema were resolved completely after surgery.

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Solitary fibrous tumor (SFT) is a rare type of mesenchymal tumor. Previously, it was thought to be a pleural based lesion; however, it has been reported to occur in many extra pleural sites with no mesothelial lining[1]. SFTs are usually benign and slow-growing, but malignant SFTs have been found to have a greater risk of recurrence and metastasis.

The aim of this presentation is to report a case of a 67-year-old male patient with huge pelvic, abdominal mass measuring 20x13x7 cm and weighing 2.8 kg.

THE CASE

A sixty-seven-year-old Bahraini male was assessed in India in January 2012 for progressive bilateral lower limb edema; CT scan of the abdomen showed a large pelvic, abdominal mass, see figure 1. The patient's condition progressed with repeated attacks of hypoglycemia.

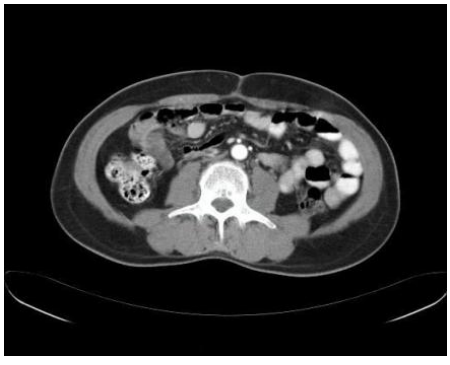

Figure 1: CT Scan of the Abdomen and Pelvis Showed Huge Retroperitoneal Pelvic-Abdominal Mass

Needle biopsy showed malignant spindle cell tumor. The patient was seen in Salmaniya Medical Complex; CT scan of the chest, abdomen and pelvis showed a huge retroperitoneal

pelvic-abdominal mass, with no distant metastases. In 2013, a mass 20x13x7 cm and weighing 2.8 kg was resected. The mass was multinodular, relatively encapsulated with myxoid changes and necrosis. Hypoglycemia and lower limb edema were resolved completely after surgery; follow-up CT scan showed no evidence of recurrence or residual tumor.

Microscopically, the tumor is "patternless" with a haphazard arrangement of bland appearing spindle cells, hypercellular and hypocellular sclerotic foci, stromal hyalinization, and a prominent branching vasculature, see figure 2. SFTs commonly express CD34, CD99 and BCL-2, see figure 3.

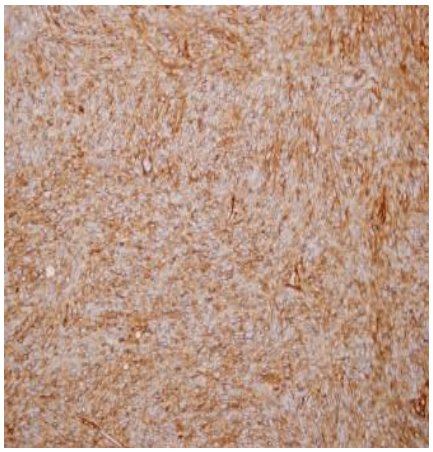

Figure 2: Tumor Exhibit High Cellularity Composed of Spindle Cells and Typical Staghorn Vessels

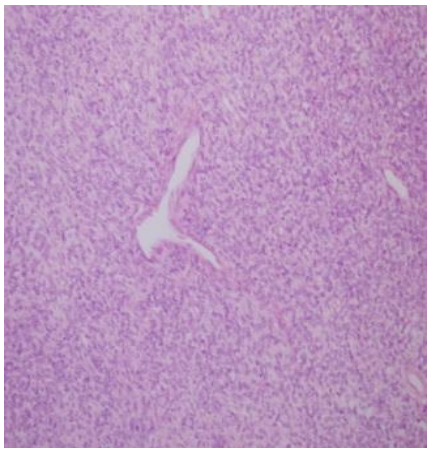

Figure 3: Diffuse Strong Positivity for CD34

DISCUSSION

SFTs are rare tumors[2]. The most common site of SFT is the pleura, but it has been described in a large variety of sites including head and neck area, sinonasal tract and oral cavity. Retroperitoneal SFT is extremely rare; only few cases were reported in the literatures[3]. Macroscopically, SFTs are well circumscribed, tan, rubbery masses often tethered by a pedicle. Microscopically, these neoplasms are described as "patternless" with a haphazard

arrangement of bland appearing spindle cells, hypercellular and hypocellular sclerotic foci, stromal hyalinization and a prominent branching vasculature.

Immunohistochemically, SFTs commonly express CD34, CD99 and BCL-2; epithelial membrane antigen and smooth muscle actin may also be expressed. They are usually negative for S-100 protein, desmin, and cytokeratins. Pathologic criteria of malignancy include a large tumor size more than 50 mm, infiltrative margins, high cellularity, nuclear pleomorphism, an area of tissue necrosis, and an increased mitotic index (more than 4 mitoses in 10 high power fields. Rare cases show abrupt transition from conventional benign appearing SFT to high-grade sarcoma[4]. Differential diagnoses of SFTs consist of epithelioid sarcoma, schwannoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, hemangiopericytoma, and synovial sarcoma, among others[5].

The treatment of choice for SFT is surgical resection. Surgical excision is curative for most benign lesions; however, malignant lesions would often recur and progress despite adjuvant radiation or chemotherapy. The most important indicator of the clinical outcome is whether or not the tumor could be completely excised in the initial operation or not. Few cases using adjuvant radiotherapy or chemotherapy in malignant SFT have been reported, and the effectiveness of radiation and chemotherapy has not been proven[6].

A long-term clinical follow-up is recommended for all patients with SFT because of its potential adverse biologic behavior which could lead to repeated recurrences and/or malignant transformation. The behavior of SFT is unpredictable, and the relationship between morphology and outcome is poor.

Approximately half of patients with malignant SFT could be cured; although, the rest often develop recurrences and metastases. A follow-up plan after resection of a malignant SFT should include semi-annual CT in the first two years, and annually after. A long-term follow-up is mandatory in the case of malignant lesions because of possible late recurrences; these could be locally aggressive and may lead to death through local invasion and compression[7,8,9].

CONCLUSION

A sixty-seven-year-old Bahraini male was diagnosed with progressive bilateral lower limb edema; CT scan of the abdomen showed large pelvic, abdominal mass.

Needle biopsy showed malignant spindle cell tumor. A mass 20x13x7 cm and weighing 2.8 kg was resected. Hypoglycemia and lower limb edema were resolved completely after surgery.

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