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Case Report

Solitary Fibrous Tumor of the Peritoneum: An Unusual Location of A Rare Tumor

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Abstract

Introduction: SFT is an uncommon tumor representing 3, 7% of all soft tissue sarcomas and mesenchymal tumor. The most common location is pleura followed by the abdomen.

Case Report: A 68 years old male patient was admitted to our department with abdominal pain and a mobile masse in his abdominal hypogastric area. CT-scan showed a well-defined mass with vividly homogenous enhancing features in both the arterial and venous phases. A complete resection of the tumor was performed thought a median sub-ombilical laparotomy. The histological examination showed a fibroblastic mesenchymal tumor with expression of CD34, CD99 and Bcl2 in the immunohistochemical study that is specific of the solitary fibrous tumor.

Discussion: SFT are anatomically ubiquitous mesenchymal tumors developed from fibroblasts. It is an uncommon tumor represents 3, 7% of all soft tissue sarcomas and mesenchymal tumors. The most common location is the pleura followed by the abdomen. Clinically, abdominal SFT is usually manifest as abdominal fullness, gastrointestinal obstruction, weight loss, jaundice, fever, or hypoglycemia. Immunohistochemistry, the cell of the SFT typically express the following markers: CD34, CD99 and Bcl2. Complete surgical removal of the tumor is the gold standard treatment which can be completed by adjuvant radiotherapy.

Conclusion: Primary SFTs in the peritoneum are extremely uncommon. Clinical symptoms and imaging manifestations are nonspecific whish make the diagnosis difficult. Treatment includes surgical resection, embolization therapy, radiation therapy, chemotherapy and anti-angiogenic agents.

Keywords: Solitary fibrous tumor; Peritoneum; Resection; Immunohistochemistry

Introduction

SFT is an uncommon tumor representing 3, 7% of all soft tissue sarcomas and mesenchymal tumor [1]. This neoplasm can present anywhere throughout the human body and has high risk of malignant transformation, the most common location is pleura followed by the abdomen [2,3].

SFT is usually presents as slow growing mass, its clinical symptoms are caused by local compression or metastasis [4-6] and its typically appears as a hypervascular tumor on CT-scan [7,8].

The commonest treatment is complete surgical excision which can be completed by radiotherapy [9].

Case Report

A 68 years old male patient was referred to our surgical department with abdominal pain without any transit disorder or digestive hemorrhage. Physical examination found a patient in good general condition, presenting a 20 diameter mobile masse in his abdominal hypogastric area.

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The patient underwent an abdominal ultrasound, which revealed a well-defined and hyperechoic masse in the pelvic. CT-scan showed a well-defined mass measuring 27 x 19 cm, after contrast injection the mass presented with vividly homogenous enhancing features in both the arterial and venous phases, the appearance of this peritoneal tumor indicated benign behavior with no invasive sings or invasion of neighborhood organs (Figure 1).

Surgical treatment was planned under general anesthesia, a complete resection of the tumor was performed thought a median sub-ombilical laparotomy (Figures 2). Outcome were uneventful and the patient was discharged 2 days after. Histological examination showed a fibroblastic mesenchymal tumor with expression of CD34, CD99 and Bcl2 in the immunohistochemical study that are specific of the solitary fibrous tumor. 18 months of surgery, the patient remained free of occurrence.

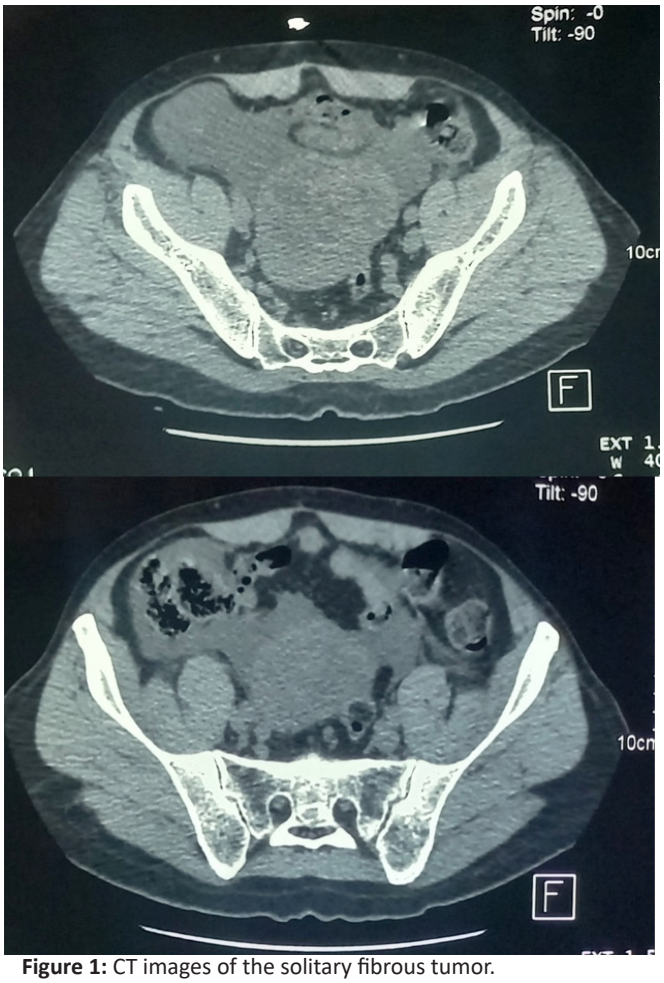

Figure 1: CT images of the solitary fibrous tumor.

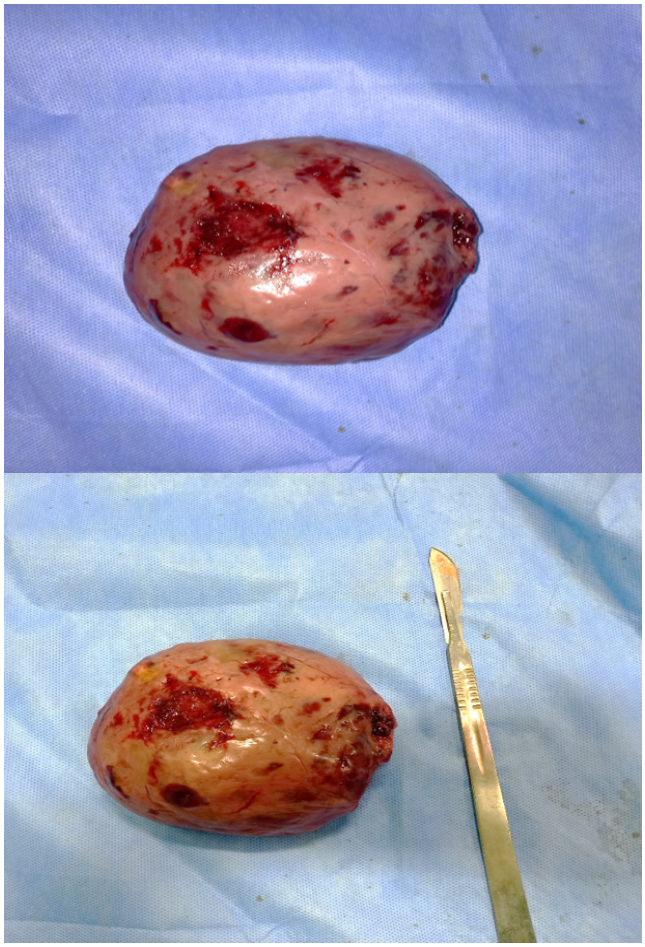

Figure 2: Postoperative images of the solitary fibrous tumor.

Discussion

First described by Klemperer and Rabin in 1931, SFT are anatomically ubiquitous mesenchymal tumors developed from fibroblasts. Subdivided by the fifth edition of the WHO classification of fibroblastic/myelofibroblastic tumors into benign, intermediate and malignant, they have the same clinical, histological, immunohistochemical and radiological characteristics regardless of their location. It is an uncommon tumor represent 3, 7% of all soft tissue sarcomas and mesenchymal tumors [1,10].

SFT occurs in patients during the 5th and 6th decades with no gender predilection. The most common location is the pleura followed by the abdomen. Pleural involvement is usually seen in older patients (56-60 years old) but abdominal location is seen in younger patients (fourth decade) [2,3].

Immunohistochemistry, the cell of the SFT typically express the following markers: CD34, CD99 and Bcl2 [11]. However, anti-STAT6s is a new marker more specific for the diagnosis and rarely positive in other types of tumors [12].

Clinically, abdominal SFT is usually manifest as abdominal fullness, gastrointestinal obstruction, weight loss, jaundice, fever, or hypoglycemia [4-6]. These symptoms are the result of adjacent organs compression by the tumor or the overproduction of insulin-like growth factor-2 (Doege-Potter syndrome) [13-15].

Computed Tomography (CT) is the first line imaging exam. It can show SFT as a well-circumscribed, hypervascular masses containing necrosis zone or calcification, with compression of nearby organs [7,8]. On Magnetic Resonance Imaging (MRI) SFT has intermediate signal intensity on T1-weighted images and heterogeneous low signal intensity with flow voids on T2-sequence. However, central focus of heterogeneity and variable contrast enhancement on CT or MRI are signs of malignancy [16].

Complete surgical removal of the tumor is the gold standard treatment. In case of positive margins (R1/R2), a second resection should be discussed. Otherwise, adjuvant radiotherapy is a reasonable option. Conservative attitude may be adopted in selected cases to preserve organ function when postoperative RT can be delivered, given a favorable long-term outcome.

Neoadjuvant RT may be an option in specific cases to improve tumor respectability or when wound complications are a concern [9].

No prospective study is available on the activity of chemotherapy activity in SFT [17].

Angiogenesis is an important feature involved in tumor growth and metastatic spread of SFT. Therefore, inhibition of angiogenesis pathways has been suspected to be a key therapeutic target to inhibit tumor cell proliferation. Given the angiogenic properties of FSTs, several case reports and clinical trials have investigated various angiogenesis inhibitors with promising activity in FSTs [9].

After complete resection, these tumors show a good prognosis, and the 10-year overall survival is in the order of 54% to 89%. However, recurrence may occur in 10% to 25% of the cases at 10 years [18,19] and it is more frequent in cases of incomplete resection (R1/R2) [20]. The risk of 5-year metastatic recurrence in lung, liver and bone is up to 40% in high-risk patients with criteria for malignancy as large size, dissemination at presentation, pleomorphism, necrosis, and a mitosis rate ≥ 4 per 10 high-power fields [20-23].

Conclusion

In conclusion, primary SFTs in the peritoneum are extremely uncommon. Clinical symptoms and imaging manifestations are nonspecific. Therefore, the diagnosis of SFTs can be challenging and relies on histological and immunohistochemical study. Treatment includes surgical resection, embolization therapy, radiation therapy, chemotherapy and anti-angiogenic agents.

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