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Solitary Fibrous Tumor of the Abdominal Wall

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Abstract

Solitary fibrous tumor is a rare neoplasm of mesenchymal origin that usually arise within the pleura. Its origin in abdominal wall is extremely rare, with only 15 cases described in the English literature.

We report the case of a 55-year-old woman presented with a mass located at the left lower quadrant of the abdominal wall. Microscopic studies revealed histologic and immunohistochemical features consistent with a SFT. Eighteen months after surgical excision of the mass followed by adjuvant radiotherapy, the patient is alive and disease-free. The authors discuss the clinicopathological features of SFTs, differential diagnosis and treatment options.

Keywords: Solitary fibrous tumor; Abdominal wall; Soft tissue; Hemangiopericytoma

Introduction

Solitary Fibrous Tumors (SFT), represent a group of rare tumors of mesenchymal origin with unique morphological and immunohistochemical features, usually identified in the pleura, although it can also be found in other locations including soft tissues [1-4]. Extrapleural SFTs represent 0.6% of all soft tissue tumors. SFTs of the abdominal wall are extremely rare, with only 15 cases reported in the anglosaxonic literature.

SFT usually presents as a well-defined, solid hypervascular mass [4]. Histologically it is composed of spindle-shaped, bland and randomly organized, fibroblast-like cells, presenting a patternless arrangement. It is also characterized by a combination of hypo-and hypercellular areas separated by thick bands of hyalinized collagen and branching vessels. The cellularity and higher or lower expression of the hyaline component characterize cellular and fibrous types. Most tumors cells mark positive for CD34, CD99 and pBcl2 (*B-cell lymphoma* 2) [4]. The ubiquitous and indolent nature of TFSs obscures and delays the diagnosis.

The authors describe a new case of solitary fibrous tumor of the abdominal wall, and discuss the clinicopathological features and differential diagnosis.

Case Report

A 55-year-old female presented with malaise, pain in both wrists and nonspecific abdominal pain. The patient had no concomitant diseases. On objective examination, she revealed well-defined and painful mass at the left lower quadrant of the abdominal wall.

The complete blood count and biochemical analysis were within the reference values. Tumor markers CEA, AFP, CA 19.9, CA 125 and CA 15-3 were within range. Abdominal ultrasound was performed, identifying a heterogeneous structure with two hyporeflective areas, being highly vascularized therein. Abdominal computed tomography (CT) located the mass at the left flank/iliac fossa, adjacent to the anterior wall of the abdomen, but apparently extraperitoneal, measuring 6.7×11.8×3.8 cm. The mass was heterogeneously hypodense, well-defined and lobulated, suffering marked enhancement effect after contrast administration.

The patient underwent exploratory laparotomy, having been identified an extraperitoneal encapsulated mass, which was excised. The macroscopic surgical margin was tumor-free, and the excision site was marked with metal clips. The patient was discharged on the 4th postoperative day, uneventfully.

Upon gross pathological examination, the mass presented a pinkish yellow, smooth and polished surface, with partially bosselated contour, showing in section a white pinkish elastic tissue. Histologically, the entire lesion consisted of cell proliferation with oval nucleus and mild atypia, distributed in different directions, in a hemangiopericitical pattern. In the periphery, the tumor presented expansive growth, bordered by a thin fibrous capsule.

The immunohistochemical analysis showed strong reactivity for vimentin, as well as for CD34, less intense for CD99 and was negative for pS100, alpha actin, desmin and C-kit. The cell proliferation marker Ki67 was immunoreactive in about 5% of the tumor cells.

The patient was submitted to adjuvant radiotherapy, having developed mild local radiodermatitis, being completely asymptomatic and without clinical or imaging evidence of disease recurrence 18 months after surgery. Follow up appointments have been performed, every 6 months, with a clinical and imaging evaluation (ultrasound or toracoabdominal CT).

Discussion

SFTs were initially thought to originate from mesothelial cells, but immunohistochemical and ultrastructural studies have identified a mesenchymal origin [2]. Hemangiopericytoma, first described by Stout and Murray in 1942, identified a group of vascular tumors with probable origin in the pericytes of Zimmerman [5]. However, all of these tumors have a common mesenchymal origin, with similar histological and immunohistochemical characteristics [1,6].

The nomenclature of this group of neoplasms has therefore been altered over the years, having some authors added the two designations-solitary fibrous tumor and hemangiopericytoma-as part

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of the same group, although the discussion remains. Therefore, solitary fibrous tumors can be divided into two main groups: the cellular forms, classically referred to as Haemangiopericytomas and hyalinized/fibrous forms, conventionally referred to as SFT. This set of lesions represent a subtype of soft tissue sarcoma that can be found anywhere on the body and in any organ containing mesenchymal cells [1,2,7].

SFTs present similar incidence in both sexes, being more frequent in patients between 20 and 70 years of age, and are located almost exclusively in the deep soft tissues, particularly in the lower extremities, retroperitoneum, pelvis, head and neck. The tumors range in size from 1 to 36 cm (mean, 6 cm) [2,4,8]. The literature has identified a female predominance in SFT of the abdominal wall. Several authors suggest a possible role of sex hormones in the pathogenesis of SFT in this location, although the issue remains controversial [2]. To our knowledge, sixteen cases, including the present one, have been reported in the English literature. Previously reported cases, including the present case, are summarized in Table 1 [1,2,9-17].

SFT behaves as an asymptomatic mass which can become painful as it grows and compresses surrounding structures. In the majority of cases identified on the abdominal wall, the patients were asymptomatic, but in 3 cases the mass causes abdominal pain or tenderness [2,5].

Two paraneoplastic manifestations may be associated, especially in the pleural forms. The most common type, hypertrophic pulmonary osteoarthropathy, is present in about 20% of cases and it presents as pain in long bones, finger clubbing, joint stiffness and swelling. Our patient presented initially with bilateral wrist pain that disappeared gradually in the first weeks after surgical excision, most likely associated with this condition. The Doege-Potter syndrome occurs in less than 5% of cases, most often associated with lesions located in the pelvis and retroperitoneum and it presents as hypoglycemia. Many authors claim that this presentation is caused by the production of IGF-II [1,8]. Hypoglycemia is more often associated with large tumors or with the presence of malignancy, present in 40% of tumors associated with this symptom [1,8].

Several imaging tests can aid the diagnosis of solitary fibrous tumor. Ultrasound can observe a well-defined, highly vascular, lobulated mass with heterogeneous echogenicity, which tends to displace adjacent structures. In most cases a nutritive vascular pedicle is identified. CT with intravenous contrast also shows a well-defined hypervascular soft tissue mass, while angiography shows a centripetal opacification from early arterial phase. Magnetic resonance imaging (MRI) exhibits an iso/hyperintense mass in T1 sequences and a hyperintense mass in T2 sequences, when compared to muscle structures. It also identifies imaging features that relate to the probability of malignancy, such as tumor size and a heterogenic signal intensity or contrast enhancement [3,4]. Positron emission tomography (PET) has shown potential to assess the degree of malignancy of these tumors, since an increase in the tracer uptake reflects an increase in intracellular glucose metabolism. More active tumors will tend to be more aggressive [2].

Macroscopically, SFT presents as a lobulated mass, with regular borders, usually encapsulated. By sectioning the tumor, we can feel a hard-elastic consistency, with white-grayish color [1]. However, only microscopic characteristics confirm of the diagnosis. Histologically, STFs are composed of fibroblastic, fusiform, spindle cells, scattered and disorganized, with alternating hypo-and hypercellular areas, and presenting varying amounts of hyalinized collagen. The cells are usually separated by enlarged and angulated branching vessels, with thickened hyalinized walls, resembling the form of "staghorns" [1,2].

As the histologic variability of solitary fibrous tumors may contribute to diagnostic difficulties, immunohistochemistry plays an important role in the differential diagnosis. CD34, a hematopoietic progenitor cell antigen, also present in endothelial cells and in some fibroblasts, is stained in 78 to 100% of SFT. CD99, expressed in leukocytes, thymocytes and also in spindle-cell tumors is also frequently encountered, as well as vimentin. Bcl-2, an apoptosis regulatory protein with oncogenic properties, is expressed in 96% of these tumors [1,2]. SFT usually show negative staining for cytokeratin, smooth muscle actin, desmin, S100 protein, early membrane antigen (EMA) and c-kit [2,7]. In the present case, the diagnosis of SFT was based on the association of typical histological and immunohistochemical features.

Case no.	Author	Age (y)/Sex	Symptoms	Size (cm)	Treatment	Follow up (months)
1	Mentzel et al. [9]	51/M	Abdominal mass	4.8	Excision	NI
2	Nielsen et al. [10]	NA/NI	NI	NI	Wide excision	WED
3	Vallat-Decouvelaere et al. [11]	50/F	Painless mass	1.9	Excision	WED, 13
4	De Saint Aubain et al. [12]	45/F	Rapidly growing mass	14	Wide excision	NI
5	De Saint Aubain Somerhausen et al. [12]	35/F	Abdominal pain	11.5	Excision	Recent case
6	Brunnenmann et al. [13]	NA/F	Painless mass	NI	Excision	NI
7	Brunnenmann et al. [13]	70/F	Painless mass	16	Excision, margin positive, preoperative CT, postoperative RT	WED, 40
8	Hasegawa et al. [14]	60/F	Painless mass	5.5	Excision	WED, 156
9	Hasegawa et al. [14]	50/F	Painless mass	3	Excision	WED, 38
10	WED, 38	50/F	Tender mass	4	Excision	WED, 14
11	Huang et al. [15]	38/F	Painless mass	7.5	Excision	WED, 12
12	Sawada et al. [16]	45/F	NI	3	NI	NI
13	Migita et al. [2]	74/F	Painless mass	12	Excision	WED, 10
14	Pencavel et al. [17]	NI	NI	NI	Excision	NI
15	Yuri et al. [1]	57/F	Painless mass	3	Excision	NI
16	Present case	55/F	Abdominal pain	11.8	Excision, postoperative RT	WED, 18

Y: Years; CT: Chemotherapy; RT: Radiotherapy; NI: No Information; WED: Without Evidence of Disease

Table 1: Clinicopathological features of solitary fibrous tumors of the abdominal wall.

	Similar to SFT	Different from SFT
Haemangiopericytoma	Mesenchymal origin - Staghorn vascular spaces - Spindle-shaped tumor cells	- No combination of varying growth patterns - No collagenous stroma - Reticulin staining pattern - Weakly positive for CD34; negative for CD99
Fibromatosis/ Desmoid tumors	- Well defined margin or infiltrative growth on CT scanning - Tendency for local recurrence	- Infiltrative border - Lack of staghorn vasculature - Bundles of collagen fibers - Negative for CD34
Deep fibrous histiocytoma	Storiform pattern - Occasional presence of bizarre mesenchymal giant cells	More circumscribed - More uniform growth pattern - Negative for CD34 - Positive for factor XIIIa
Low-grade monophasic synovial sarcoma	- Branching staghorn vessels - Fascicular growth of spindle cells	- Positive for cytokeratin and EMA - Negative for CD34
Dermatofibrosarcoma protuberans	- Positive for CD34	- Superficial involvement of reticular dermis and/or subcutis - Ill-defined borders with honeycomb-like infiltrative patterns - Negative for bcl-2
Extraorbital giant cell angiofibroma	- Positive for CD34 and vimentin - Variable positivity for bcl-2 and CD99	- Pseudovascular angiectasic spaces - Pleomorphic or multinucleated giant cells
Pleomorphic hyalinizing angiectatic tumor of soft parts	Positive for CD34 and vimentin - Variable positivity for bcl-2 and CD99	- Multinucleated stromal cells - Pleomorphic tumor cells with clear pseudoinclusions - Presence of vascular trombi, subendothelial fibrin deposits and hemosiderin deposits - Propensity for local recurrence
Spindle-cell lipoma	Positive for CD34 - Mesenchymal origin - Rare cases of SFTs with fatty islands	- Predominant in subcutaneous tissue - No collagenous background or vascular pattern

EMA: Epithelial Membranous Antigen

Table 2: Differential diagnosis of solitary fibrous tumors of the abdominal wall.

The differential diagnosis of solitary fibrous tumors include various benign and malignant neoplasms, summarized in Table 2 [2,9,15].

Most SFTs have a benign course, although 10 to 15% show malignant behavior, both by means of local recurrence and metastatic disease [1,7]. The risk of distant metastases in soft tissue SFT is estimated to be about 25% [8], Histological criteria of malignancy remain controversial because the clinical behavior cannot be simply predicted by the histological features. However, the possibility of malignancy should be considered if the following characteristics are present: tumor larger than 5 cm, infiltrative growt h, high cellularity, nuclear pleomorphism, necrotic foci and the presence of more than 4 mitoses/10 high power fields [1,2,7]. A weak expression of CD34 has also been associated with increased incidence of malignant tumors. Resectability is believed to be the most reliable prognostic factor in SFTs [2].

Retrospective studies suggest that, regardless of the clinic and whenever possible, radical surgical resection with adequate margins is the treatment of choice for SFTs [1,7]. In general, these tumors exhibit a favorable clinical behavior, with an overall survival rate at 10 years of 54-89% after primary surgical resection [7]. When hypoglycemia is associated, corticosteroid therapy may improve symptoms [1]. Metastatic lesions, most frequently located at the lung, liver or bone, must be surgically removed whenever possible [4]. Radiotherapy may be used as adjuvant therapy in patients at risk for recurrence or as a primary therapy for inoperable masses. There are reports on the use of neoadjuvant radiotherapy in retroperitoneal and pelvic SFT. Systemic chemotherapy has not shown significant response [7]. Is has also been studied the use of angiogenesis inhibitors (Temozolomide and Bevacizumab) in the treatment of unresectable tumors or in cases of local recurrence and metastatic disease [7]. Literature has presented overall survival rates of 80% at 5 years and 70% at 10 years [4].

The lack of reliable prognostic factors implies long-term follow up, since there are reported cases of recurrence more than 10 years after surgery. To the authors knowledge, none of the sixteen cases showed evidences of recurrence or metastatic disease [2,7,9,17].

Conclusion

Solitary fibrous tumors represent a group of mesenchymal tumors, most frequently presented as asymptomatic masses, difficult to recognize until causing compression of neighboring structures. The correct diagnostic can only achieved with the aid of histology and immunohistochemistry. Treatment is primarily surgical and may be supplemented by adjuvant radiotherapy.

These tumors present an ambiguous biological behavior, although clinical imaging and histological characteristics can predict a higher likelihood of malignancy. Cases of late recurrence or metastatic disease have been identified in extrapleural SFTs, which may imply a long-term follow up.

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