Asian Journal of Case Reports in Surgery

3(1): 6-11, 2020; Article no.AJCRS.53678

The Solitary Fibrous Tumor with Unusual Retroperitoneal Localization

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Authors' contributions

All authors contributed to the conduct of this work. They also declare that they have read and approved the final version of the manuscript.

Article Information

Editor(s):
(1) Dr. Georgios Tsoulfas, Associate Professor, Aristoteleion University of Thessaloniki, Thessaloniki, Greece.
Reviewers:
(1) Michael Bordonaro, USA.
(2) Manas Bajpai, NIMS Dental College, India.
(3) Antonio Augusto Ornellas, Instituto Nacional de Câncer, Brazil.
Complete Peer review History: http://www.sdiarticle4.com/review-history/53678

Case Report

Received 20 October 2019
Accepted 23 December 2019
Published 03 January 2020

ABSTRACT

Solitary fibrous tumors (SFTs) are rare mesenchymal tumors, usually described at the pleura level, but since they can develop in different extra pleural locations. Retroperitoneal location is rarely described.

The anatomopathological study remains the key to the diagnosis of SFT, the curative treatment is based on surgical excision. Most extra-thoracic SFT are benign, but may recur or metastasize after complete removal, even in the absence of histopathological evidence of malignancy. Careful long-term monitoring is therefore recommended.

We report the case of retroperitoneal SFT in a 70-year-old male who was consulting for generalized abdominal pain. The aspects of imaging, anatomopathology and immunohistochemistry are detailed.

Keywords: Solitary fibrous tumor; retroperitoneum; immunohistochemistry; surgery.

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1. INTRODUCTION

Solitary fibrous tumors (SFTs) are mesenchymal tumors first described by Klemprer and Rabin in 1931 [1], they are generally developed at the expense of serosa and mainly at the pleura level. However, in recent years, several extra pleural locations have been reported, their low clinical aggressiveness, ubiquitous nature and ability to simulate a multitude of soft tissue neoplasias make the diagnosis of these mesenchymal tumors difficult. We report a case of a large retroperitoneal solitary fibrous tumor.

2. OBSERVATION

A 70 year old patient, operated on 23 years ago for a left inguino-scrotal hernia, had been consulting for generalized abdominal pain for one year with recurrence of his left hernia for 6 months, associated with without transit disorder, no externalized digestive bleeding, no urinary signs, all evolving in a context of apyrexia and preservation of the general state. The clinical examination found a voluminous hypogastric mass of 20cm of major axis, firm, fixed to the deep plane and mobile to the superficial plane with abdominal collateral circulation and a reducible left inguino-scrotal hernia, the digital rectal examination was normal. The rest of the examination is without particularities (Fig. 1).

The biological check-up was normal and in particular the blood sugar level.

The abdominal ultrasound showed a very large tissue mass of 20 cm of major axis, well limited, containing multiple fluid areas. This mass pushed back the bladder and digestive structures and compressed both ureters with bilateral hydronephrosis.

The contrast injection CT scan objectively showed a voluminous abdominal mass of 215/ 125 mm, with regular contours, spontaneously heterogeneous density, increasing intensely and heterogeneously after injection, with areas of hypodensity, and compressing both ureters without locoregional invasion, with large left inguino-scrotal hernia (Fig. 2).

A guided echo biopsy had isolated 2 cores, the morphological aspect correlated to the immunohistochemical profile was in favour of a solitary fibrous tumor.

The patient was operated on by a median laparotomy, the tumor was retroperitoneal of 24/20 cm, highly vascularized, pushing back digestive and vascular structures, contracting intimate adhesions, with the bladder, aorta, lower vena cava and iliac vessels. There was no extension to the surrounding organs, the release of the mass was laborious, a complete resection of the mass was performed with cure of the left inguino-scrotal hernia (Fig. 3).

The immediate postoperative consequences were simple for the first five days, complicated on the sixth day postoperatively by respiratory distress secondary to pulmonary embolism with death of the patient on the same day, despite a preventive anticoagulant treatment.

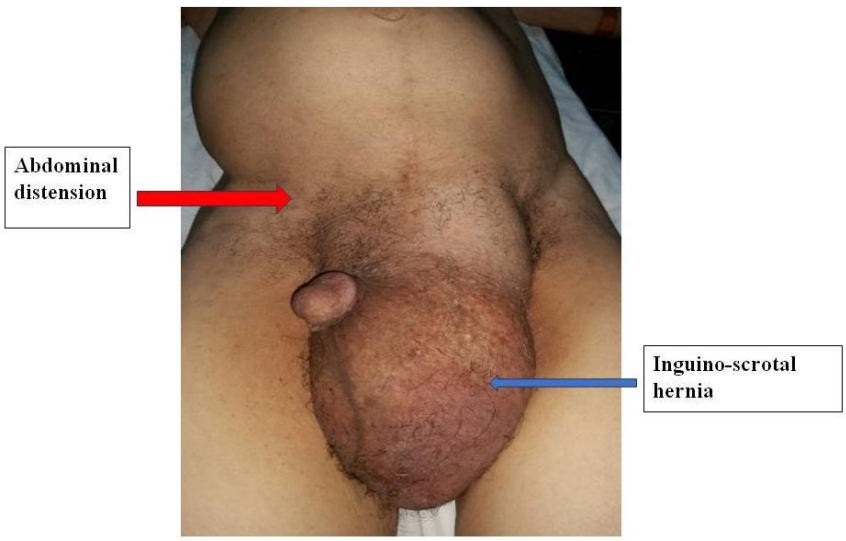

Fig. 1. Abdominal pelvic mass with left inguino-scrotal hernia

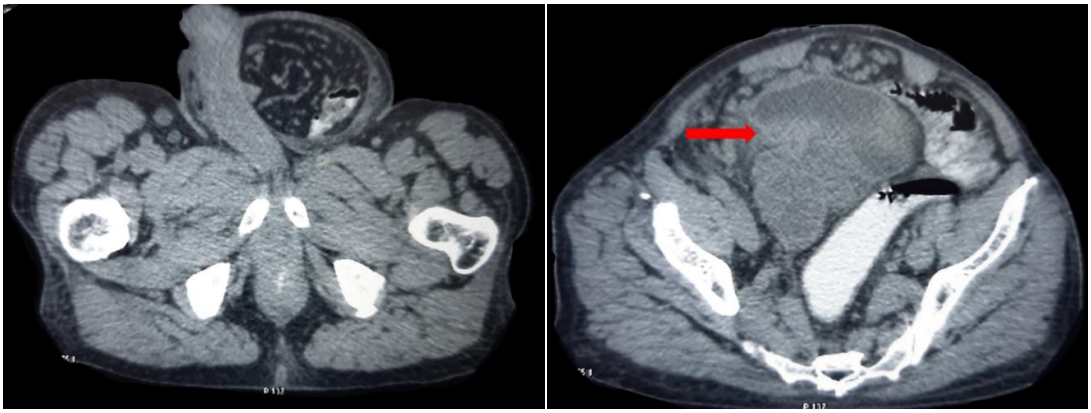

Fig. 2. Abdominal scan showing: A- the abdominal-pelvic mass (arrow). B- the left inguino-scrotal hernia

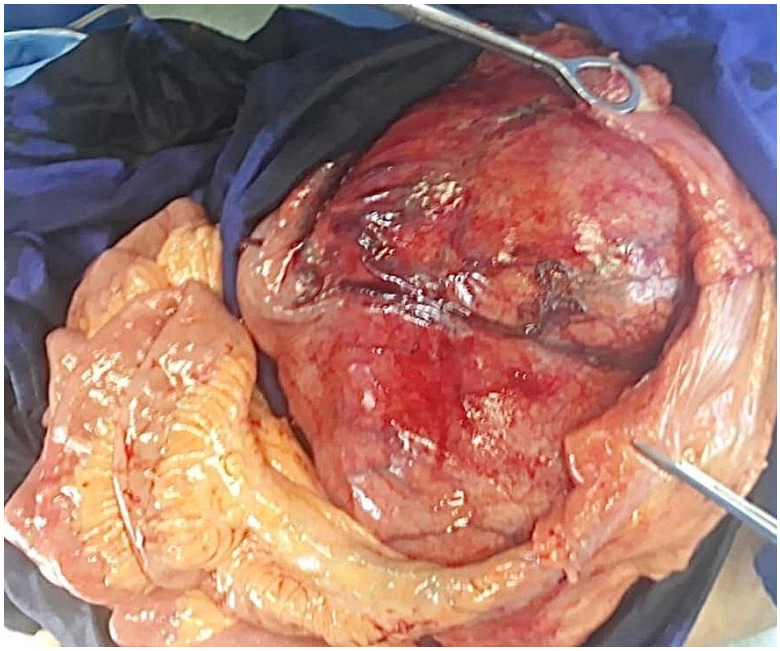

Fig. 3. Peroperative aspect of the tumor

The anatomopathological study of the part shows, a very limited and encapsulated voluminous tumor that weighed 4 Kg measuring 24/21/14 cm (Fig. 4), with on histological examination a heterogeneous mesenchymal tumor proliferation made of diffuse dense zone and loose zones with low cell count, the tumor cells are ovoid or fusiform with fibroblastic appearance, and the nuclei show no atypia, mitotic activity is very low estimated at less than 2 mitoses per 10 fields, the stroma has a hemorrhagic vascular ectasia, moreover it is found fibrous remodelling hyaline oedematous and necrosis outbreak.

The immunohistochemical study shows that tumor cells strongly express vimentin CD34, STAT6 and do not express PS100, AML or CD117. The diagnosis chosen is that of malignant SFT.

3. DISCUSSION

Retroperitoneal solitary fibrous tumors (SFT) are mesenchymal tumors rarely reported in the literature, many names have been given to them; benign mesothelioma, submesotheloma, localized fibrous tumor, hemangiopericytoma [2].

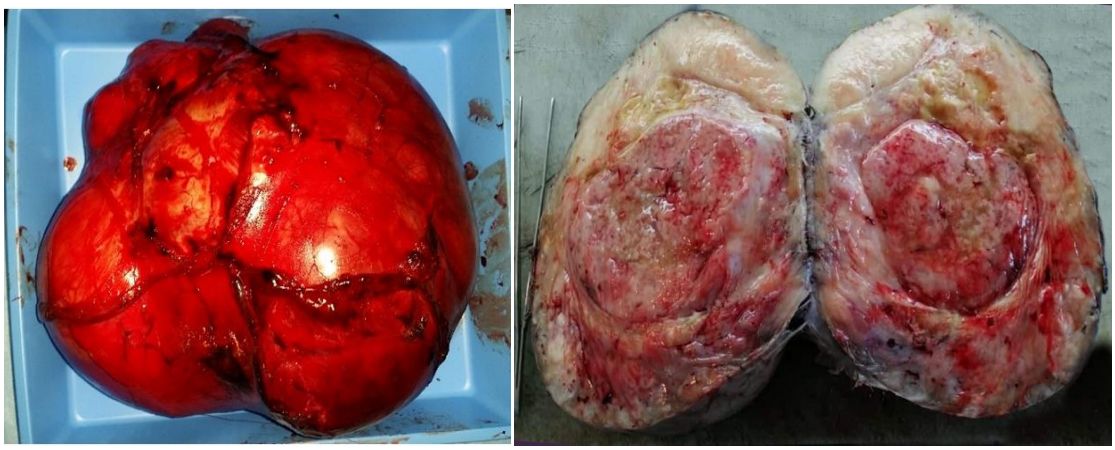

Fig. 4. Macroscopic aspect of the tumor mass

The discovery of a recurrent genetic alteration led to the grouping of these originally independent entities under the generic term SFT, it is an intrachromosomal fusion of genes (NAB2-STAT6) located on the long arm of chromosome 12q13 with a mutation responsible for tumor growth [3], they represent less than 2% of all soft tissue masses, 80% of SFT are located in the chest cavity, the abdomen represents the second site [4].

The extra pleural locations reported in the literature are very varied; ovaries, prostate, kidney, mammary gland, liver, mesocolon, vulva, orbit, central nervous system and thyroid [5,6,7,8,9]. SFT can be observed at any age, most often with the 5th and 6th decade, without predominance of gender [10].

The ubiquitous and indolent nature of SFT makes their diagnosis difficult and late. Clinically, the majority of extra-thoracic SFT are asymptomatic, their clinical manifestation depends essentially on their topography, after a long evolution, the appearance of a painful mass remains the main sign [11]. Hypoglycemic events have also been described associated with SFT (Doege-Potter syndrome) due to tumor production of high molecular weight insulin-like growth factor (IGF), specifically IGF-II [11].

On ultrasound, SFT is presented as a very limited, rounded or oval mass, hypoechoic, often heterogeneous in relation to the presence of foci of hemorrhage or necrosis [12].

CT shows a well limited and intensely enhanced tissue density formation after contrast injection due to a large vascular contingent [8]. Necrosis foci in large SFT can be seen giving it a heterogeneous appearance, calcifications are possible [12,13]. Nuclear magnetic resonance imaging (MRI) specifies the intense iso character in T1 sequence, with a heterogeneous aspect alternating hyper- and hypo-intensive ranges in T2 sequence, and an equally heterogeneous enhancement after gadolinium injection [8,14].

The anatomopathological study remains the key to the diagnosis of SFT, macroscopically, it is a solid tumor, with a smooth surface, often voluminous, well circumscribed, polylobulated, does not infiltrate the healthy parenchyma with foci of central necrosis, cystic areas or more rarely calcifications [10]. When the tumor sits on a serous surface, a vascular pedicle is often noted [8].

SFT typically produces a tumor proliferation made of single-morphous spindle cells arranged in intertwined bundles arranged in an anarchic manner within a fibrocollagen stroma, focally hyalinised and containing a rich vascular system, often of hemangiopericitaire type [10].

Immunohistochemically, CD34, O-13 (CD99) and the protein bcl-2 appear to be the most characteristic markers of these lesions. The high level of KI-67 protein and tumor cell labelling for the p53 oncogene confirm the malignant nature of SFT [4,15], more recently, the discovery of NAB2- The STAT6 fusion gene has led to a more accurate diagnosis of SFT [4,7].

According to the WHO classification, FSTs are classified as having intermediate malignant potential with a low risk of metastasis [12,13]. Demicco et al. Propose a risk stratification

model, which includes age (>55 years), high mitotic activity, tumor size and tumor necrosis as factors and defines three categories "low", "intermediate" and "high risk" [16,17]. The most recognized malignancy criteria are cytonuclear atypia, high mitotic activity, high cellularity, pleomorphism, importance of necrosis and bleeding [16,18]. While hypoglycemia, central nervous system localization and dedifferentiation have already been proposed as factors of poor prognosis. The preferred sites of secondary lesions are the liver, bone and lungs [7].

Therapeutically, management usually consists of complete surgical removal of the lesion.

Experience shows that embolization of SFT feeder arteries can allow safe resection or biopsy, but should not be used primary treatment [19].

Few articles in the literature have focused on adjuvant therapy, whether chemotherapy, radiotherapy, or more recently targeted therapies. Given the relatively good and indolent results and the nature of this tumor, radiotherapy is not currently recommended after complete resection with healthy margins [20,21], however chemotherapy may be of interest in malignant forms that are not immediately operable. This chemotherapy could be justified as a neoadjuvant, in large tumors or postoperatively, in the event of incomplete resection [22]. Because of the risk of recurrences and metastases noted in 10 to 15% of cases, close monitoring is necessary [23,24].

4. CONCLUSION

Lonely retroperitoneal fibrous tumors are uncommon in the literature. The nature and starting point of these tumors appears difficult to determine on simple imaging data, particularly in the case of large tumors. Biopsy of these tumors can guide the diagnosis. The most complete surgical excision possible is the treatment of choice. No studies have shown the efficacy of complementary treatment with chemotherapy or radiotherapy. Finally, it is very important to follow up in the long term these patients who are at risk of relapse.

CONSENT

As per international standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
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