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Case Report

CT of retroperitoneal solitary fibrous tumor☆,☆☆

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ABSTRACT

Solitary fibrous tumors are rare tumors of pluripotent fibroblastic or myofibroblastic origin that generally arise among older individuals, with a mean age of onset ranging from 55 to 65 years. Though typically associated with pleural involvement, solitary fibrous tumors can emerge in virtually every anatomic location within the body. Although most solitary fibrous tumors are benign, approximately 20% may exhibit malignant features such as local invasion, recurrence, and metastases. In this article, we report the case of a 58-year-old male with a diagnosis of a retroperitoneal solitary fibrous tumor. We analyze computed tomography imaging findings and additionally correlate imaging features with the patient's unique pathological and genotypic findings to optimize diagnosis.

Introduction

Solitary fibrous tumors (SFTs), formerly classified as hemangiopericytomas, comprise a histologic spectrum of fibrotic mesenchymal neoplasms with rare metastatic potential. SFTs are rare, accounting for only 3.7% of all visceral and soft tissue sarcomas and tumors of intermediate malignancy [1]. The mean age of presentation of SFTs is between 55 and 65 years with no sex predilection [2]. Though typically regarded as tumors that arise within the thoracic region, SFTs can emerge anywhere in the body, with the abdomen being the most common extrapleural site of involvement [3]. Intra-abdominal SFTs pose a unique diagnostic challenge given that they typically remain asymptomatic and grow to large sizes (>20 cm) before they begin to exert mass effect onto other organs and are subsequently detected [4]. Cross-sectional imaging plays a central role in detection, localization, and characterization of these tumors and serves as a roadmap for operating surgeons. In this report, we highlight the case of a 58-year-old male who presented with new onset scrotal swelling in addition to worsening abdominal and lower extremity swelling that was eventually diagnosed as an abdominal SFT.

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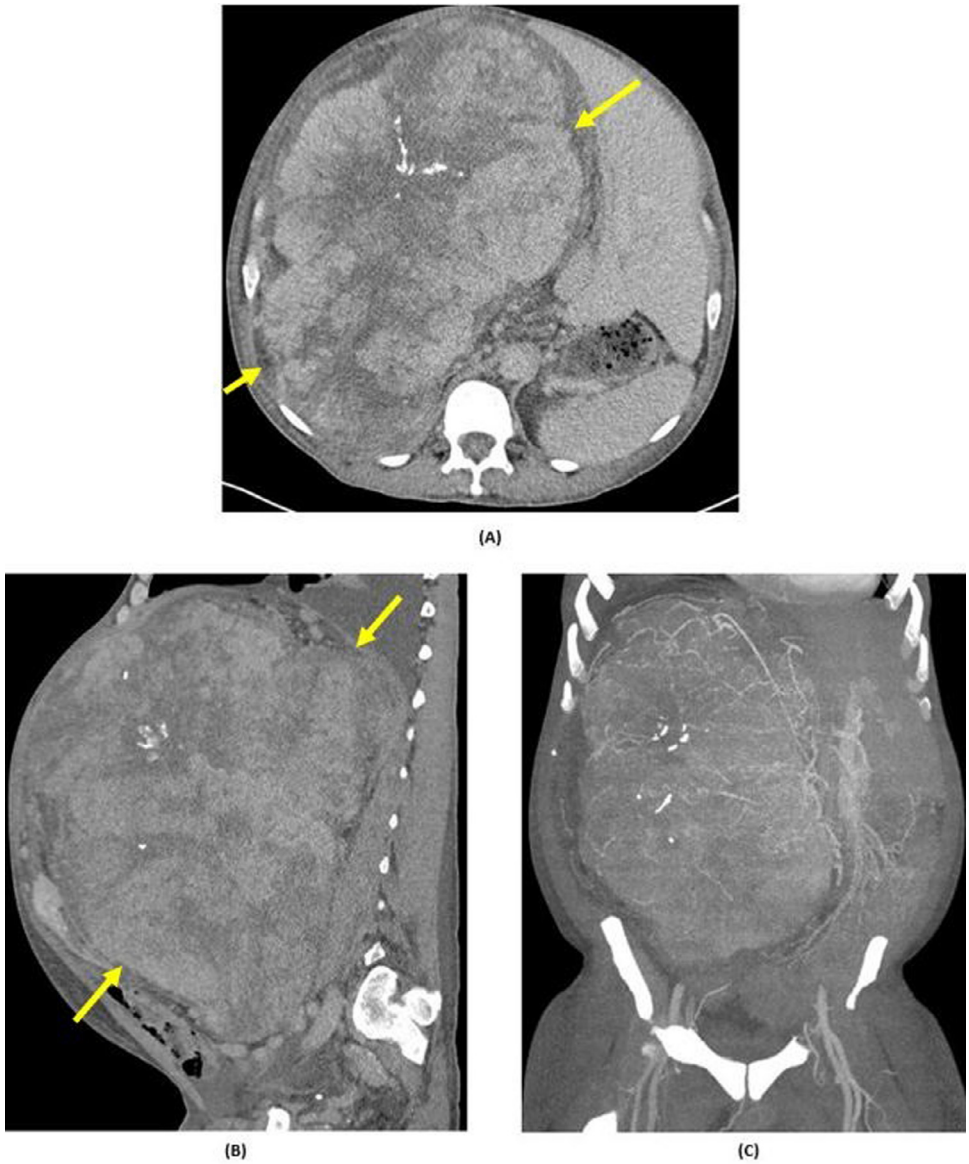

Fig. 1 – A 58-year-old male presented to his local emergency department for new onset scrotal swelling in addition to worsening abdominal and lower extremity swelling over the past year. A computed tomography (CT) scan was performed for further evaluation. (A) Axial CT demonstrates a large retroperitoneal mass measuring 28 x 22 x 33 cm with an irregularly lobulated contour (arrows). Faint calcifications can be seen within the mass. (B) Redemonstration of mass on sagittal CT (arrows). Note faint calcifications and irregular enhancement. (C) MIP imaging in coronal plane defines extensive vascularity of the tumor and its lack of defined margins.

Case report

A 58-year-old male with a past medical history significant for alcohol use disorder presented to his local emergency department for new onset scrotal swelling in addition to worsening abdominal and lower extremity swelling over the previous year. An ensuing computed tomography (CT) scan revealed a large heterogeneous right-sided retroperitoneal mass with a stellate-type appearance occupying the abdomen and pelvis (Fig. 1). The mass measured 28 × 22 × 33 cm, demonstrated an irregularly lobulated contour, and had areas of bright enhancement, septations, as well as faint calcifications. A significant mass effect was noted on the surrounding structures with elevation of the right hemidiaphragm, anteroinferior displacement of the right kidney, and left anterolateral displacement of the hepatic parenchyma. In addition, numerous surrounding enlarged venous collaterals were noted, alongside moderate ascites and right-sided pleural effusion. No biopsy was performed given concern for adrenocortical carcinoma and anticipation that surgical resection would be necessary irrespective of histology. The patient subsequently underwent successful radical resection of the retroperitoneal mass, right nephrectomy, right adrenalectomy, and partial hepatectomy.

Ensuing results from surgical pathology ultimately revealed a diagnosis of a SFT on the basis of histologic features as well as immunophenotype.

Discussion

First described as a unique pathological entity in 1931, SFTs were long thought to be of mesothelial origin exclusively involving the pleura, pericardium, and peritoneum [5]. Recent advances in pathology, however, have led to better understanding of the histogenesis, broad anatomical distribution, and histologic diversity of these tumors. It is now established that SFTs may arise from anywhere within the body, with extrapleural SFTs more common than pleural SFTs. However, with an annual incidence rate of 0.35 per 100,000, SFTs remain uncommon, and SFTs of the retroperitoneum, as reviewed in our case, are exceedingly rare accounting for less than 30% of all SFTs [1,6].

Clinically, presenting symptoms of SFTs are specific to their site of origin but broadly include pain, a palpable mass, and neurologic or vascular symptoms as seen in our case [4]. Urinary retention and bowel obstruction may be the presenting symptoms in patients with tumors of the abdomen or pelvis [4]. A small subset (5%) of patients, particularly those with larger SFTs, may also present with symptomatic hypoglycemia [7]. This is secondary to excess insulin growth factor 2 production by the tumor and is a manifestation of a paraneoplastic syndrome referred to as Doege-Potter syndrome [7].

CT is the preferred initial diagnostic method for identifying SFTs. At imaging, retroperitoneal SFTs typically appear on CT scans as well defined, predominantly hypervascular masses with varying degrees of cystic change and necrosis. SFTs are generally isodense to adjacent muscles on unenhanced images and typically demonstrate avid yet heterogeneous enhancement following intravenous contrast administration. In certain tumors, central areas that do not enhance or enhance less could be indicative of necrosis or cyst formation. Although rare, calcifications may appear in larger benign or malignant tumors, as seen in our case. Differential diagnosis for retroperitoneal SFTs at imaging consists of other mesenchymal neoplasms such as gastrointestinal stromal tumor, synovial sarcoma, malignant mesothelioma, leiomyosarcoma, desmoid tumor, or neurogenic tumors and lymphoma. CT in these cases not only aids in establishing the primary diagnosis, but also helps in evaluating the extent of the tumor and its invasion into nearby structures, alongside identifying locoregional and distant metastases.

The pathological diagnosis of SFT is confirmed on the basis of histological characteristics and immunohistochemistry (IHC) [8]. IHC markers CD34 and STAT6 strongly support the diagnosis of SFT; both were positive in our case [8]. The most specific genotypic marker of SFTs, however, is identification of the NAB2-STAT6 fusion gene [8]. A notable finding in our case was that despite the presence of STAT6 IHC positivity, which is considered a sensitive surrogate of the NAB2-STAT6 fusion gene, testing for the NAB2-STAT6 fusion gene at our institution was negative [8].

Although most SFTs are benign, malignant features may be detected at pathological exam in up to 20% of cases with approximately 20% also recurring within 10 years [3,9]. At our institution, we use a modified 4-variable risk stratification model to predict risk of metastatic disease in SFTs that includes patient age (<55, ≥55), tumor size (<5 cm, 5 cm to <10 cm, 10 cm to <15 cm, ≥15 cm), mitotic count (0, 1-3, ≥4), and presence of tumor necrosis ≥10% [10]. Using these validated criteria, our patient had a high risk of developing metastatic disease.

Though risk stratification can characterize the malignancy potential of SFTs, complete en bloc surgical resection remains the standard of care for all localized SFTs [11]. Preoperative embolization may also be considered given the highly vascular nature of SFTs. In our case, approximately 17 L of blood loss was reported during the successful resection. Recently, novel antiangiogenic agents have also demonstrated effectiveness as first-line therapy but research on this is still in preliminary stages and patients should be encouraged to participate in ongoing clinical trials when appropriate [12]. Anecdotal evidence also suggests that adjuvant radiotherapy has prevented recurrence [13]. Given the rarity of this tumor, however, establishing any consensus on management beyond surgical resection is difficult to prospectively assess and adjuvant management should be evaluated on a case-by-case basis in a multidisciplinary tumor board.

Patient consent

The patient reported in the manuscript signed the informed consent/authorization for the participation in the research, which includes the permission to use data collected in future research projects including presented case details and images used in this manuscript.

REFERENCES

[1] De Pinieux G, Karanian M, Le Loarer F, Le Guellec S, Chabaud S, Terrier P, et al. Nationwide incidence of sarcomas and connective tissue tumors of intermediate malignancy over four years using an expert pathology review network. PLoS One 2021;16(2):e0246958.
[2] Cardillo G, Facciolo F, Cavazzana AO, Capece G, Gasparri R, Martelli M. Localized (solitary) fibrous tumors of the pleura: an analysis of 55 patients. Ann Thorac Surg 2000;70(6):1808–12.
[3] Gold JS, Antonescu CR, Hajdu C, Ferrone CR, Hussain M, Lewis JJ, et al. Clinicopathologic correlates of solitary fibrous tumors. Cancer 2002;94(4):1057–68.
[4] Shanbhogue AK, Prasad SR, Takahashi N, Vikram R, Zaheer A, Sandrasegaran K. Somatic and visceral solitary fibrous tumors in the abdomen and pelvis: cross-sectional imaging spectrum. Radiographics 2011;31(2):393–408.
[5] Klemperer P, Coleman BR. Primary neoplasms of the pleura. A report of five cases. Am J Ind Med 1992;22(1):1–31.
[6] Miettinen M. Chapter 12: solitary fibrous tumor, hemangiopericytoma, and related tumors. In: Miettinen M, editor. Modern soft tissue pathology: tumours and

non-neoplastic conditions. New York: Cambridge University Press; 2010. p. 335.
[7] Zafar H, Takimoto CH, Weiss G. Doege-Potter syndrome. Med Oncol 2003;20(4):403–7.
[8] Geramizadeh B, Marzban M, Churg A. Role of Immunohistochemistry in the diagnosis of solitary fibrous tumor: a review. Iran J Pathol 2016;11(3):195–203.
[9] Salas S, Resseguier N, Blay JY, Le Cesne A, Italiano A, Chevreau C, et al. Prediction of local and metastatic recurrence in solitary fibrous tumor: construction of a risk calculator in a multicenter cohort from the French Sarcoma Group (FSG) database. Ann Oncol 2017;28(8):1979–87.
[10] Demicco EG, Wagner MJ, Maki RG, Gupta V, Iofin I, Lazar AJ, et al. Risk assessment in solitary fibrous tumors: validation and refinement of a risk stratification model. Mod Pathol 2017;30(10):1433–42.
[11] Rajeev R, Patel M, Jayakrishnan TT, Johnston FM, Bedi M, Charlson J, et al. Retroperitoneal solitary fibrous tumor: surgery as first line therapy. Clin Sarcoma Res 2015;5(1):19.
[12] Martin-Broto J, Stacchiotti S, Lopez-Pousa A, Redondo A, Bernabeu D, de Alava E, et al. Pazopanib for treatment of advanced malignant and dedifferentiated solitary fibrous tumour: a multicentre, single-arm, phase 2 trial. Lancet Oncol 2019;20(1):134–44.
[13] Baldi GG, Stacchiotti S, Mauro V, Dei Tos AP, Gronchi A, Pastorino U, et al. Solitary fibrous tumor of all sites: outcome of late recurrences in 14 patients. Clin Sarcoma Res 2013;3:4.