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Case report

Surgical resection of a presacral solitary fibrous tumor with extension to iliac vessels using Karakousis's abdominoinguinal approach: Report of a rare case

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ABSTRACT

Introduction and importance: The solitary fibrous tumor (SFT) is a rare tumor of mesenchymal origin, with a reported incidence of 2.8 cases per 100,000 tumors and with distinctive histopathological and immunohistochemical characteristics. It was initially described as a pleural lesion and subsequently, it was found in different organs and tissues. The abdominoinguinal incision described by Karakousis allows a safe and radical approach for lower quadrants abdominopelvic tumors.

Case presentation: A 47-year-old man was referred to us with a 5-months history of lower backache radiating to the left lower limb. MRI and CT revealed a retroperitoneal mass of 10 cm extending to left iliac vessels. The initial diagnosis corresponded to a sarcomatous retroperitoneal tumor. It was decided to perform an abdominal exploration using Karakousis's approach for surgical resection. The immunohistochemistry and histopathological study revealed neoplasia compatible with a SFT. It was categorized as low risk for developing metastasis and death from disease, according to the new malignancy criteria. Currently, the patient is asymptomatic and disease-free at 19 months after surgery.

Clinical discussion: Most patients with SFTs present symptoms derived from the tumor growth and the compression on adjacent structures with clinical manifestations that are frequently insidious and precede the tumor discovery. The diagnosis is based on histopathological studies. Nonetheless, when they present an extrathoracic location, they represent a diagnostic challenge, due to their variable histological characteristics.

Conclusion: Presacral SFT is a rare entity, with a scant incidence reported regarding this location and long-term treatment. Surgical resection is needed as the immediate treatment.

1. Introduction

The solitary fibrous tumor (SFT) is a rare mesenchymal neoplasia, with a reported incidence of 2.8 cases per 100,000 tumors [1]. Although it is mainly developed in the pleura, it has also been found in various extrathoracic sites [2]. The 30% appear in the abdominal-pelvic region and frequently, they have a silent evolution with low risk of recurrence and metastasis, being the surgical resection the treatment of choice [3,4].

The abdominoinguinal approach described by Karakousis in 1980, allows a safe and effective radical approach to tumors in the lower quadrants of the abdomen, based on the correct exposure of iliac vessels, warranting the vascular control and, in turn, allows access to retroperitoneal space by contiguity [5].

We present the case of a patient with a presacral SFT extending to iliac vessels, which required the Karakouis's approach for its surgical resection. This case report is reported in line with the SCARE Guidelines [6].

2. Case presentation

A 47-year-old man was referred to us with a 5-months history of lower backache radiating to the left lower limb. This patient did not have any other significant comorbidities and he was evaluated outside

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with magnetic resonance imaging (MRI) which demonstrated a left retroperitoneal mass (presacral space), which extended to the ipsilateral internal and external iliac vessels; it was also in contact with the gluteal vessels and the left sciatic nerve (Fig. 1). The laboratory test were within normal ranges, including blood glucose, prostate-specific antigen, alpha-fetoprotein, CEA, and CA 19-9. With the digital rectal exam, a posterolateral extrarectal mass is detected. Besides, a videocolonoscopy was made presenting no alterations. The computed tomography (CT) showed a mass of 9.4 × 9.8 × 11 cm in the anteroposterior, transverse, and craniocaudal sense, respectively. It showed fat planes of separation with sigmoid rectum and bladder, originating an important displacement of these organs to the right (Fig. 2).

A CT guided biopsy of the mass was performed, which reported fibroconnective tissue with myxoid changes in positive immunohistochemistry (IHC) for vimentin, suggesting possible mesenchymal origin.

The case was discussed in the multidisciplinary institutional committee, and on suspicion of a retroperitoneal sarcoma, it was decided a surgical rescue.

An exploratory laparotomy was performed using the Karakousis's left abdominoinguinal approach (Fig. 3). In the exploration, we found a tumor of about 10 cm in all its dimensions, lateralizing bladder, and rectum to the right side. The left iliac vessels and the medialized ipsilateral ureter were identified; and the tumor was released, separating it from the colon, rectum, and anterior face of the bladder. Once the ureter and the contralateral iliac vessels were identified, the tumor was separated from the sacrum. During the lateral release, it was detected that the tumor extend to the left obturator fossa, compromising the homolateral obturator nerve, which made difficult its resection. Subsequently, it was possible to separate from the sacrum, where the tumor extended to the sciatic notch (Fig. 4). The operative time was 5 h, with an estimated blood loss of 150 cm^3. One unit of red blood cells was required. The immediate postoperative follow-up was carried out in the intensive care unit. Later he went to the common room, where he completed the hospitalization for five days.

The histopathological study and the IHC revealed a positive fusocellular neoplasia for CD34, vimentin, Bcl-2, and B-catenin, compatible with a SFT (Fig. 5). There was no evidence of necrosis and the mitotic activity was low (1 mitosis/10 HPF).

Periodic clinical controls were made. At present, the patient is asymptomatic with no evidence of local relapse or distance at 19 months after the surgery.

3. Discussion

SFTs predominantly affect adults, equally distributed between men and women, between the fifth and sixth decade of life [7]. They can be difficult to diagnose given that they are great simulators of soft-tissue tumors, and are more frequently located in deep somatic soft tissues and within body cavities, specially pleura and pelvis [8]. On the other hand, retroperitoneal SFTs are even rarer and only about 50 cases are described in the literature [9]. In our case, the age of presentation was <55 years, with a retroperitoneal location, specifically in the presacral space, where some isolated cases have been reported, so its true incidence is unknown. According to the literature, presacral tumors represent a heterogeneous group of neoplasias, predominantly benign, with an incidence of 1: 40,000 admissions and of 0.014% from the documented rectoscopies [10].

Most patients with SFTs present symptoms derived from the tumor growth and the compression on adjacent structures; they usually present as a slow-growing and not very painful mass, with clinical manifestations that are frequently insidious and precede the tumor discovery [1]. In this report, the patient presented as a manifestation, pain inlower backache radiating to the left lower limb.

The SFT diagnosis is based on histopathological studies. Nonetheless, when they present an extrathoracic location, they represent a diagnostic challenge, due to their variable histological characteristics [11]. Besides, other presacral tumors present similar histopathological findings, as the hemangiopericytoma, malignant mesothelioma, synovial cell sarcoma, leiomyomatous tumor, granulosa cell tumors and gastrointestinal stromal tumor (GIST) [11]. However, the imaging techniques may help to suspect the nature of the tumor, due to its great vascularization, and also allow studying its relationships with neighboring

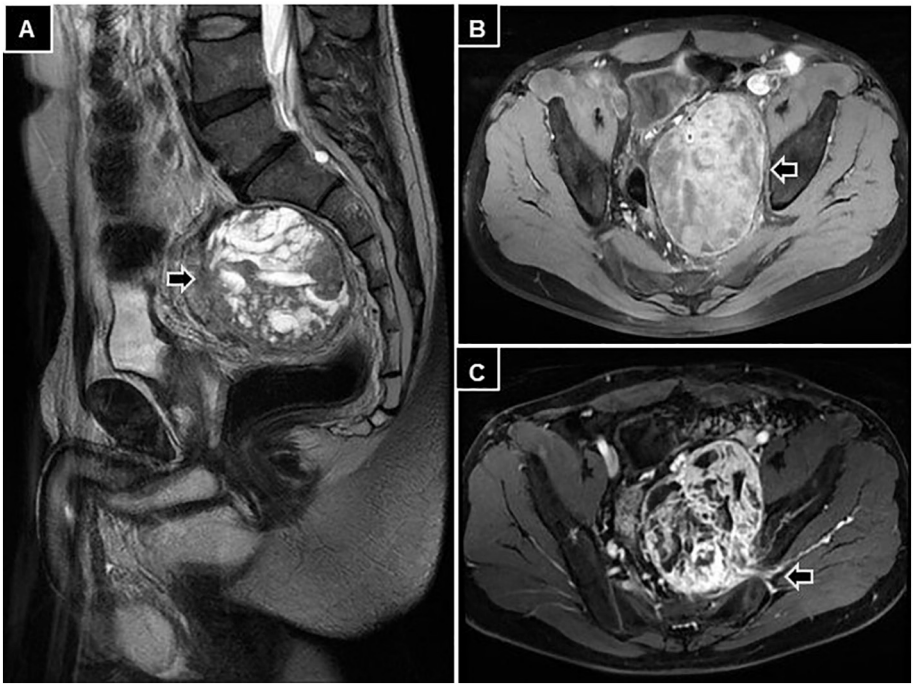

Fig. 1. Abdominal and pelvic MRI. A: In sequence T2, a mass with presacral location is seen, with solid areas of intermediate signal intensity (black arrow). B: In sequence T1, heterogeneous lesion, well-defined borders, and hyperintense, with some trabecular areas (black arrow). C: It is noteworthy the development of pelvic collateral circulation and left gluteal vessel ingurgitation (black arrow).

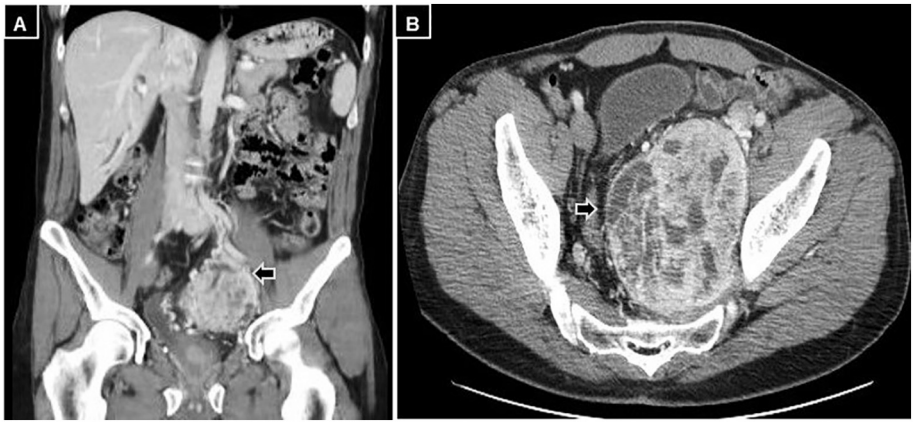

Fig. 2. Abdominal and pelvic CT in arterial phase. A: An increase of highlighted areas to the tumor center alternating with others without contrast uptake suggesting collagenization spots is observed (black arrow). B: The tumor presents zones with early marked enhancement, which begins at the periphery. Besides, fat planes of rectum separation are observed (black arrow).

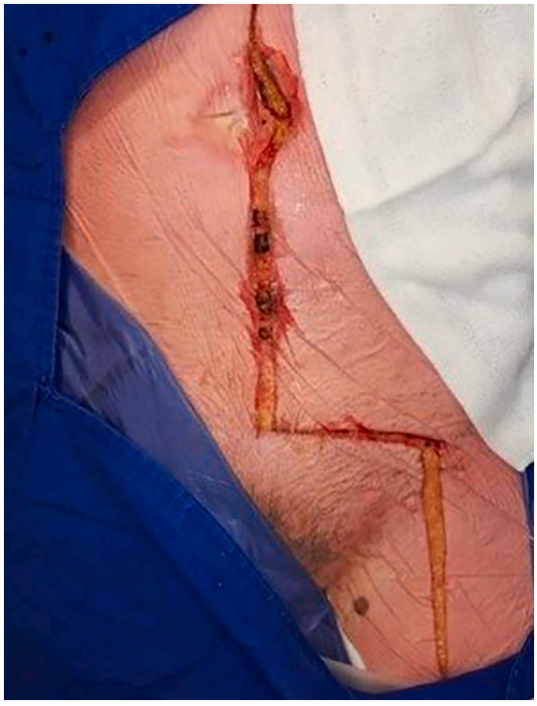

Fig. 3. Karakousis's left abdominoinguinal approach.

structures for better planning of the surgical act [12].

The imaging techniques of choice are CT and MRI, in which SFTs are often heterogeneous, with hypervascular areas that show an intense enhancement, hypercellular areas that show a moderate enhancement, and cystic areas with myxoid degeneration or necrosis that show no enhancement at all. However, other hypervascular tumors with fibrous content also show these characteristics, including most of the sarcomas, GISTs, and malignant peripheral nerve sheath tumors, for this reason, it is difficult to distinguish these tumors with only CT or MRI [13]. In this patient, the findings suggested that it was a malignant mesenchymal tumor, but without demonstrate a definitive diagnosis. This explains the reason why most abdominal-pelvic SFTs are diagnosed postoperatively [3].

The histopathological study shows a profusion of vascular elements, surrounded by asymmetrically distributed spindle cells, with a predominance of collagen and reticulin fibers; immunohistochemically, they frequently present Positivity for CD34 (80–90%), CD99 (70%), Bcl-2 (30%), epithelial membrane antigen (30%), and smooth muscle actin (20%) [14]. Our case presented a similar behavior.

The clinical course of SFTs has been difficult to predict in terms of the histological characteristics; although most tumors are benign, 5–10% recur or metastasize, typically to the lungs, liver, and bone, including some cases with a benign histologic appearance [15]. The traditional criteria of malignancy include a great size ($\geq$10 cm), dissemination at presentation, pleomorphism, necrosis, and mitotic rate $\geq$ 4 per HPF. The malignant SFTs present a metastatic rate from 20 to 30%, and given the fact that these tumors have unpredictable behavior, new risk stratification models have been developed in recent years [14]. A 3-level model incorporated the age (<55 or >55 years), tumor size (<5 to $\geq$15 cm, with 5 cm increments) and mitotic count (0.1–3 or >4/10 HPF), for the general risk of metastasis and death [16]. Subsequently, it was validated, with the addition of necrosis as the fourth criterion (<10 or >10%) increasing the percentage of correctly categorized patients as low-risk and identifying with greater accuracy a high-risk group with the rapid development of metastasis [17]. The case described here was categorized with a score of 3, when the definitive diagnosis of SFT was obtained in the postoperative period, demonstrating that this patient had a low risk of developing metastasis and death from disease.

The treatment of choice is surgical resection. However, the main difficulty to treat these huge retroperitoneal pelvic masses is that the best approach is still not well defined. Generally, most authors use a midline abdominal incision. This incision does not allow a good exposure of the caudal portion of the external iliac vessels, as the presence of tumor hinders further visibility [18]. The Karakousis's abdominal-inguinal approach the only one that warrants the radical resection of retroperitoneal tumors with pelvic extension by allowing an exposure in continuity from the abdominal aorta to iliofemoral vessels, thus facilitating the en-bloc resection and dissection of the tumor mass with vascular control and preservation of vascular-nervous package [19]. Our case demonstrates that the use of this approach guarantees safe resections along the iliac vessels and although the dissection appears to be extensive, blood loss was controlled since the preservation of the vascular vessels was achieved, with successful resection of the SFT.

This approach must be present in every surgeon's arsenal, allowing a resectability rate of up to 95% of abdominal lower-quadrant tumors which are otherwise considered unresectable [18].

Due to the rarity of presacral SFTs, as well as the confusion regarding the histopathological diagnosis, the treatment guidelines remain unclear at present.

4. Conclusion

Presacral SFTs are a rare entity, with scant evidence with regard to

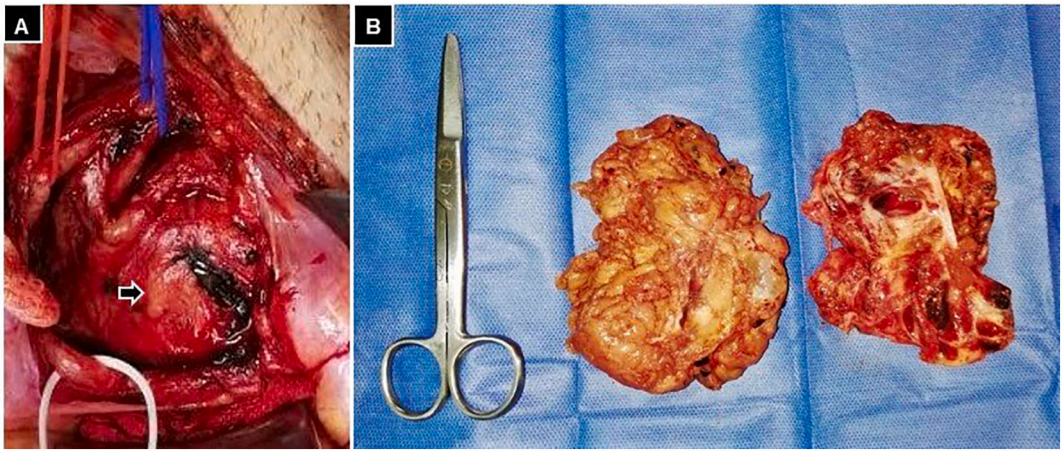

Fig. 4. A: Identification of elements with vessel loops. Iliac artery (red), iliac vein (blue) and ureter (white). STF (black arrow). B: Tumor of 11 × 8 × 8 cm, weighing 315 g, and light beige colored.

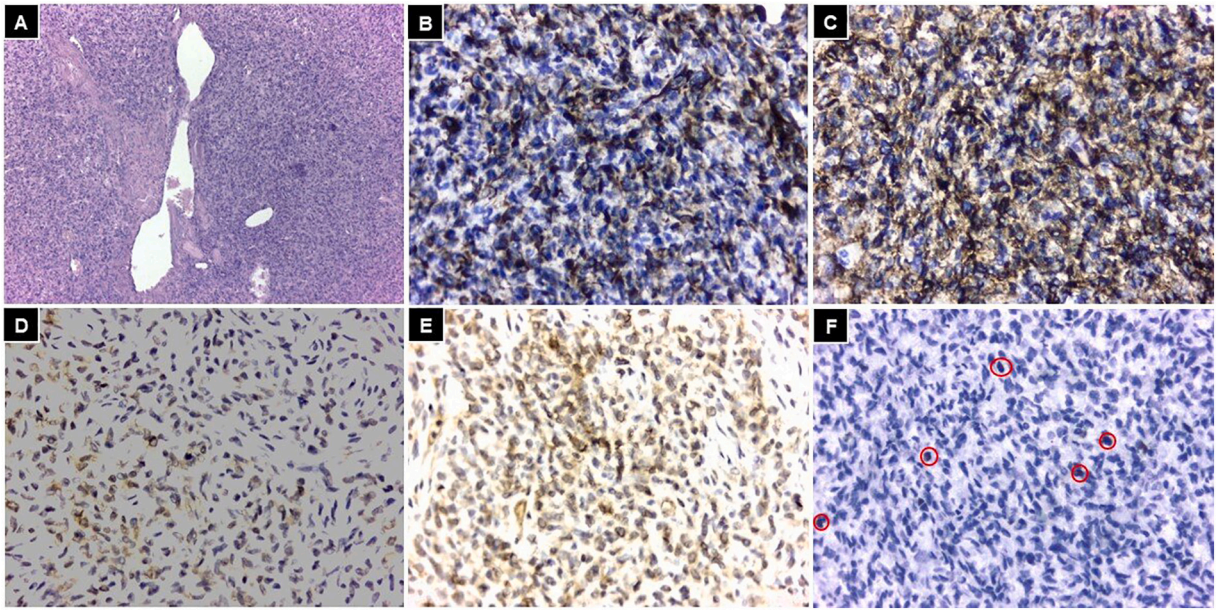

Fig. 5. A: HE, 10×, spindle cells with focal nuclear pleomorphism and a prominent vascular branching pattern, similar to hemangiopericytoma are seen. B: Positive IHC for vimentin. C: Positive CD34. D: Focal positive Bcl-2. E: Positive β-catenin. F: Ki67 < 10%, indicating that this is a benign lesion or has low-grade malignancy (positive nuclei in red circles).

this location and long-term treatment. The definite SFT diagnosis must be establish by immunohistochemistry and its immediate treatment is surgical resection, being the Karakousis's approach a technique that warrants safety and neurovascular control.

Ethical approval

This is a case report study and ethical approval not required.

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CRediT authorship contribution statement

Diana A. Pantoja Pachajoa, and René M. Palacios Huatuco did the literature search, prepared the draft manuscript, and wrote the final version of the manuscript.

Gabriela Sambuelli contributed to be involved in the pathological diagnosis of the patient.

Germán R. Viscido, and Facundo I. Mandojana contributed to the surgical treatment of the patient, and the manuscript review.

Alejandro M. Doniquian did the literature review, and approved the final manuscript.

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Research registration number

Not applicable.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Patient perspective

Not applicable.

Declaration of competing interest

There is no conflict to be declared.

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