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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Case report

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Abstract

Background: 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 radical reliable approach for abdominal-pelvic tumors in lower quadrants. We present the case of a patient with a presacral STF extending to iliac vessels, which required the Karakousis's approach for its surgical resection.

Case Presentation: Forty-seven-year-old man with pain in lumbosacral region radiating to left leg since 5 months. MRI and CT show evidence of a 10cm-retroperitoneal mass (presacral space) extending to left iliac vessels. The early diagnosis corresponded to a sarcomatous retroperitoneal tumor. It was decided to carry out the abdominal scan through the Karakousis's approach for surgical resection with permanent neurovascular control. The immunohistochemistry and histopathological study revealed a CD-34 positive spindle-cell neoplasia, vimentin, Bcl-2, and β-catenin, compatible with a SFT. It was categorized as low risk for developing metastasis and death from disease, according to the new criteria for malignancy. At present, the patient is asymptomatic and disease-free at 17 months after surgery.

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.

Background

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 safely and effectively the radical treatment of tumors located in abdominal lower quadrants, 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 TFS extending to iliac vessels, which required the Karakousis’s approach for its surgical resection.

Case Presentation

A 47-year-old man with no clinical or surgical history, consulted for pain in the lumbosacral region radiating to the left leg since 5 months. Magnetic resonance imaging (MRI) is taken and shows evidence
of a left retroperitoneal mass (presacral space), which encapsulates the homolateral internal and external iliac vessels; besides, it is in contact with the gluteal vessels and the homolateral sciatic nerve (Fig. 1).

The computed tomography (CT) shows a lump of 9.4 x 9.8 x 11 cm in the anteroposterior, transverse, and craniocaudal sense, respectively. It shows fat planes of separation with sigmoid rectum and bladder, originating an important displacement of these organs to the right (Fig. 2).

With the digital rectal exam, a posterolateral extrarectal mass is detected. Besides, a videocolonoscopy was made presenting no alterations. The laboratory test results were within normal ranges, including the prostate-specific antigen, alpha-fetoprotein, CEA, and CA 19 - 9.

A CT guided biopsy from the lump is made, which shows fibroconnective tissue with myxoid changes in positive immunohistochemistry (IHC) for vimentin, suggesting possible mesenchymal origin.

In the multidisciplinary institutional committee, the case is discussed and on suspicion of a retroperitoneal sarcoma, it is decided a surgical rescue.

An exploratory laparotomy is made through the Karakousis’s left abdominoinguinal approach. In the exploration, we find a tumor of about 10 cm in all its dimensions, lateralizing bladder, and rectum to the right side. The retroperitoneal space is entered, identifying the left iliac vessels and the homolateral ureter medialized; one then proceeds to extract the tumor, detaching it from the colon, rectum, and anterior side of the bladder. Once identified the ureter and the contralateral iliac vessels, one then proceeds to detach the tumor from the sacrum. During the lateral detachment, it is observed that the tumor extends to the left obturator fossa, compromising the homolateral obturator nerve, which made difficult its resection.

Subsequently, one manages to separate the sacrum, where the tumor extended to the sciatic notch (Fig. 3). The operation time was 5 hours, with an estimated blood loss of 150 cc. Two units of red blood cells were required. Immediate postoperative follow-up is done in the intensive care unit. Later he goes to the common room, where he completes the hospital stay 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. 4). There was no evidence of necrosis and the mitotic activity was low (1 mitosis / 10 HPF).

Scheduled clinical controls were made. At present, the patient is asymptomatic with no evidence of local relapse or distant at 17 months after the surgery (Fig. 5).

**Discussion**

SFTs predominantly affect adults, equally distributed between men and women, between the fifth and sixth decade of life [6]. They can be difficult to diagnose given that they are great simulators of soft-part tumors; they are usually located in deep somatic soft tissues and within body cavities, specially pleura and pelvis [7]. On the other hand, retroperitoneal SFTs are even rarer and only about 50 cases are described in the literature [8]. In our case, the age at presentation was < 55 years, with a retroperitoneal
localization, specifically in the presacral space, where some isolated cases have been reported, which is why the true incidence is still 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 [9].

Most patients with STFs 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 in lumbosacral region radiating to lower extremity.

The STF diagnosis is based on histopathological studies. Nonetheless, when these have an extrathoracic localization, they constitute a diagnostic challenge, due to their variable histological characteristics [10]. 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) [10]. However, the imaging techniques may help suspect the nature of the tumor, for its great vascularization; besides, they allow the study of adjacent structures for a more effective surgical planning [11].

The imaging techniques of choice are CT and MRI, in which STFs 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 [12]. In this patient, the findings suggested the presence of a malignant mesenchymal tumor, although without demonstrating a definite diagnosis. This explains the reason why most abdominal-pelvic STFs 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%) [13]. Our case presented a similar behavior.

The molecular genetic analysis has proved to be vital for the differential diagnosis of soft tissue tumors. Recently, STFs have been associated with a fusion of genes NAB2-STAT6 that have demonstrated to be very specific and sensitive [14].

The clinical course of STFs 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 STFs present a metastatic rate from 20–30%, and given the fact that these tumors have unpredictable behavior, new risk stratification models have been developed in recent years.
A 3-level model incorporated the age (< 55 or ≥ 55 years), tumor size (< 5 to ≥ 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 presented a score of 3, having then a low risk of developing metastasis and death by disease.

The treatment of choice is surgical resection, being 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 [18]. In this case, surgical resection STF was successful using this approach.

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 [19].

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

**Conclusion**

Presacral STFs are a rare entity, with scant evidence with regard to this location and long-term treatment. The definite STF 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.

**Abbreviations**

STF: Solitary fibrous tumor; MRI: Magnetic resonance imaging; CT: Computed tomography; IHC: Immunohistochemistry; GIST: Gastrointestinal stromal tumor.

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**Availability of data and materials**
All relevant data are included in the published report.

**Ethics approval and consent to participate**

The patient provided informed consent for this treatment strategy.

**Consent for publication**

Not applicable.

**Competing interests**

The authors declare that they have no competing interests.

**Authors' contributions**

DPP and RPH did the literature search and prepared the draft manuscript. GS contributed to be involved in the pathological diagnosis of the patient. GV, AD and FM contributed to the manuscript review. DPP and RPH wrote the final version of the manuscript. All authors read and approved the final manuscript.

**References**


**Figures**
Abdominal and pelvic MRI. A: In sequence T2, a lump with presacral location is seen, with solid areas of intermediate signal intensity. B: In sequence T1, heterogeneous lesion, well-defined borders, and hyperintense, with some trabecular areas. C: It is noteworthy the development of pelvic collateral circulation and left gluteal vessel ingurgitation
Figure 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. B: The lump presents zones with early marked enhancement, which begins at the periphery. Besides, fat planes of rectum separation are observed.
Figure 3

A: Karakousis's left abdominoinguinal approach. B: Identification of elements with vessel loops. Iliac artery (red), iliac vein (blue) and ureter (white). STF (black arrow). C: Lump of 11 x 8 x 8 cm, weighing 315 gr, and light beige colored.
Figure 4

A: HE, 10x, 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).

Figure 5
Abdominal and pelvic CT at 17 months after surgery. A and B: The retroperitoneum does not present compatible images with adenomegalies, with conservation of peritoneal fat planes, without evidence of local or distant relapse.