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CASE REPORT

A Rare Case of Solitary Fibrous Tumour of the Pelvis in an 18-Year-Old Young Man: CT and MRI Features with Pathologic Correlations

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Raffaele Ascione[1,*]
Felice Crocetto[2,*]
Andrea Ponsiglione[1]
Savio Domenico Pandolfo[2]
Annarita Gencarelli[1]
Luigi Insabato[1]
Massimo Imbriaco[1]
Ciro Imbimbo[2]

[1]Department of Advanced Biomedical Science, University "Federico II", Naples, Italy; [2]Department of Neurosciences, Human Reproduction and Odontostomatology, University "Federico II", Naples, Italy

*These authors contributed equally to this work

Abstract: Solitary fibrous tumors (SFTs) are mesenchymal neoplasms of fibroblastic origin, even if commonly seen in the pleura, they can occur anywhere in the body. SFT presents as a slow growing, often asymptomatic mass, generally affecting middle-aged adults regardless of the sex. We report a rare case of an 18-year-old man referred to our institution to perform computed tomography (CT) and magnetic resonance imaging (MRI), to investigate a pelvic mass incidentally discovered at abdominal ultrasound examination. A well circumscribed, heterogenous and hypervascular lesion was described at imaging, with absence of calcifications, hemorrhage, necrosis nor cystic degeneration. The mass removal was performed via the Da Vinci-assisted robotic surgery. Histopathological evaluation confirmed the diagnosis of SFT. CT and MRI can aid the identification of SFT, providing useful information which needs to be supported by histopathological analysis.

Keywords: solitary fibrous tumor, computed tomography, magnetic resonance imaging, robotic surgery

Introduction

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms of mesothelial or submesothelial origin representing less than 2% of all soft-tissue tumors.[1] While SFTs were originally thought to be exclusively located in the pleura, the pericardium, or the peritoneum, recent studies have shown that they can occur anywhere in the body.[2–6] SFT manifests as a slow growing, often asymptomatic mass which affects middle-aged adults regardless of gender. Symptoms include pain, neurologic or vascular alterations. Tumors occurring in the abdomen or pelvis give symptoms due to the compression of the neighboring organs.[7] Even if the majority of extra-pleural SFTs have a benign prognosis, 10–15% of these recur or become malignant.[7–9]

We hereby report a rare case of pelvic SFT occurring in a young 18-year-old man, as assessed by computed tomography (CT) and magnetic resonance imaging (MRI).

Case Presentation

An 18-year-old man with no relevant clinical history or any symptoms underwent an abdominal ultrasound examination which revealed the presence of an unexpected pelvic mass. The patient was then referred to our institution to perform pelvic CT and MRI examinations. CT scan showed a large, well-defined hypervascular

Correspondence: Andrea Ponsiglione
Department of Advanced Biomedical Sciences, University "Federico II", Via S. Pansini, 5, Naples 80131, Italy
Email a.ponsiglionemd@gmail.com

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tumor of about 5 x 4 cm located in the cave of Retzius (Figure 1). Subsequent contrast-enhanced MRI well depicted the heterogenous mass, showing intermediate signal on T1 and T2-weighted (T1w and T2w) images and high-signal intensity on T2w-STIR sequences, with areas of signal void preferentially located in the peripheral part of the tumor. After paramagnetic contrast agent administration, the lesion showed early, strong and persistent enhancement (Figure 2).

Given the size of the lesion and in order to exclude malignant entities such as angiosarcoma or malignant fibrous histiocytoma, a surgical approach was deemed necessary and the patient underwent a mass removal procedure via the Da Vinci-assisted robotic surgery. After resection, the removed mass of about 5.6×4.4×1.8 cm was subjected to pathological examination. Macroscopically it was apparently capsulated, with a smooth and shiny surface. In section the tumor appeared solid and brownish with whitish fibrous shoots. Microscopically, the neoplasm was characterized by high cellularity and comprised spindle-like cells with soft nuclei and no nucleoli, which were interspersed into a fibrous matrix. The mass was highly vascularized with 1 mitosis per 10HPF. The proliferative activity was evaluated through Ki67 and was approximately 10%. Immunohistochemical analysis revealed that the tumor had a strong positivity for STAT-6 and CD34, low positivity for bcl-2, and negativity for smooth muscle-actin, S-100 protein and desmin, confirming the diagnosis of solitary fibrous tumor (hemangiopericytoma) (Figure 3). The patient did not suffer from any complications after surgery and is currently in good health.

Discussion

SFTs are mesenchymal tumors of fibroblastic origin, composed of spindle-shaped cells that produce fibrous collagen

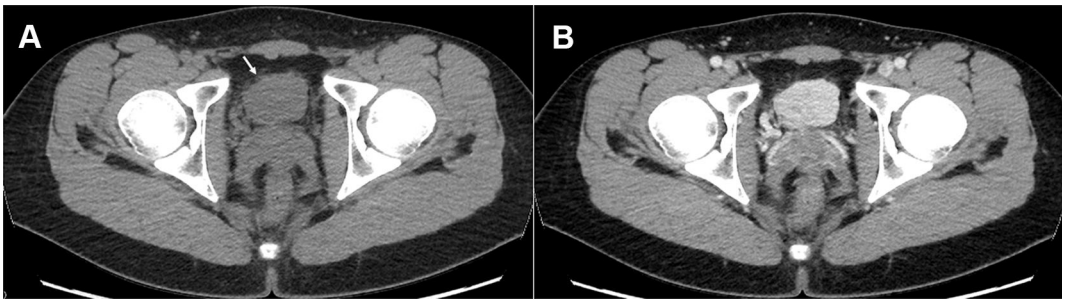

Figure 1 Computed tomography: homogenously hypodense pelvic mass (arrow) on axial unenhanced image (**A**), which appears hypervascular on axial enhanced scan (**B**).

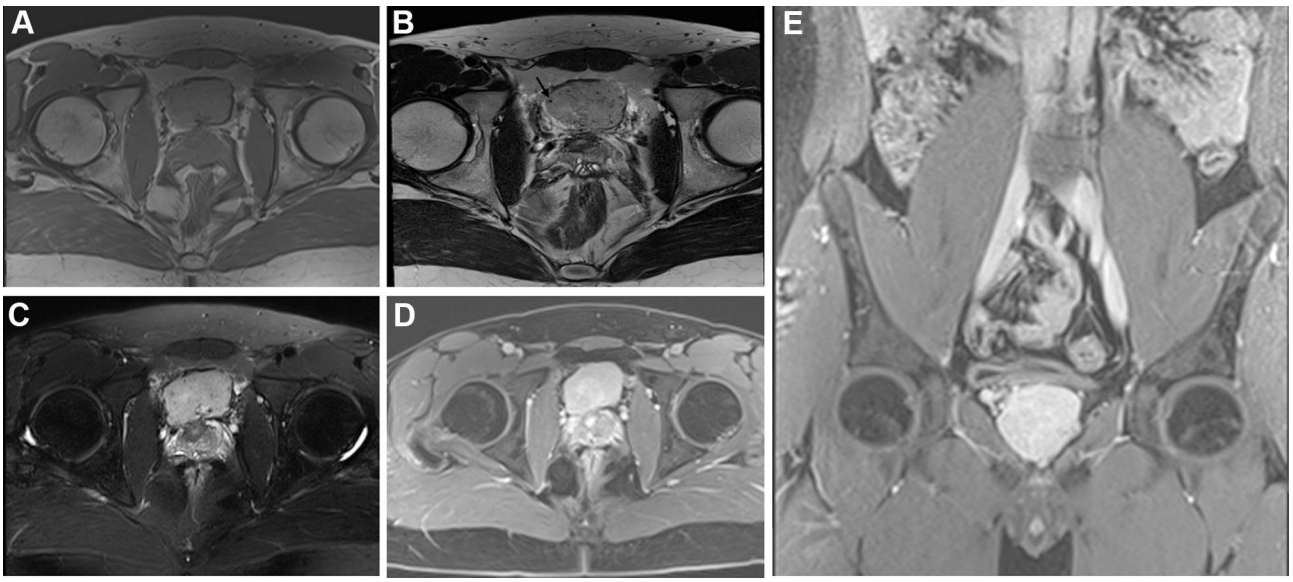

Figure 2 Magnetic resonance imaging: the mass shows intermediate signal intensity on axial T1- (**A**) and T2- (**B**) weighted images while high-signal intensity on axial T2-weighted-STIR image (**C**) with areas of signal void preferentially located in the peripheral part (arrow). Post-contrast images on arterial phase in axial plane (**D**) and delayed phase in coronal view (**E**) show an intense and persistent enhancement.

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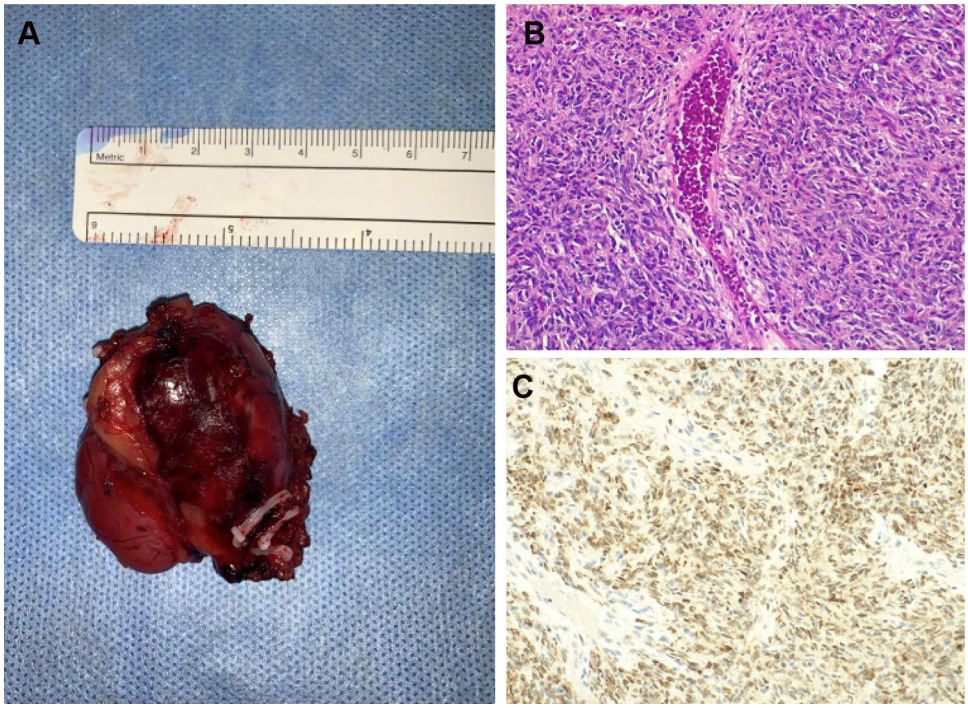

Figure 3 Gross features of the lesion specimen are shown (**A**). Hypercellular areas consisting of bland and uniform oval to spindle cells with minimal cytoplasm, small elongated nuclei and indistinct nucleoli were evident in the hematoxylin and eosin stain, 20 x magnification (**B**). Strong positivity for STAT-6 was detected, 20 x magnification (**C**).

and mucoid material.[7] At CT scans they manifest as well-circumscribed, hypervascular masses that may determine mass effects on neighboring structures. At MRI images, SFTs generally show intermediate signal intensity on T1w images and heterogeneous low signal intensity on T2w images.[7,10,11] In particular, pelvic SFTs generally appear as hypervascular neoplasms, with various degrees of intratumoral cystic change, necrosis, or hemorrhage.

Wignall et al[5] reported 34 cases of SFTs, mainly located outside the thoracic cavity. The most common location was the soft tissues and muscles (50%), followed by the pelvis (29%).[5]

Tan et al[12] previously described the case of a large pelvic mass located anteriorly to the sacrum in a 76-year-old man whose imaging features steered toward a diagnosis of angiomyxoma or liposarcoma. However, histological analysis revealed the presence of a SFT.[12]

Zhao et al[13] reported the case of a 66-year-old man with a pelvic mass detected during a screening examination. CT and MRI showed an inhomogeneous, mixed soft tissue tumor of the pelvis that was misdiagnosed as a cystadenoma originated from the seminal vesicle due to its imaging features. Nevertheless, the diagnosis based on pathological assessment was a SFT arising from the seminal vesicle.[13]

Differential diagnosis for hypervascular, intra-pelvic lesions includes high grade sarcoma, angiosarcoma, hemangioendothelioma, angiomyxoma and solitary vascular metastatic lesions.[7,10] As previously reported, the presence of large collateral feeding vessels can be a useful distinguishing imaging feature of SFTs, even if not specific.[5,14] They can be easily depicted at MRI as areas of signal void, mainly located in the periphery of the tumor. Another important differential diagnosis includes peripheral nerve sheath tumor (PNST), that generally presents as a T2w hyperintense and hyper-vascular lesion, making differentiation from SFT challenging.[15] However, malignant PNSTs may infiltrate into adjacent organs and tissues as well as erode adjacent bone. Other pelvic tumors, such as malignant fibrous histiocytoma and mesothelioma can share similarities with SFTs but are characterized by a predominant fibrotic component.[7,10]

In our case, no calcifications were observed at CT scan, which may be suggestive of tumor malignancy, as previously observed for the sarcomatous degeneration of malignant peritoneal SFTs and malignant pleural mesothelioma.[16] Neither hemorrhage, necrosis nor cystic degeneration were detectable. Therefore, on the basis of CT and MRI features, we hypothesized the presence of a benign entity such as a SFT, even if these tumors are unusual for such a young

male patient.[7] Complete surgical resection with negative histologic margins was performed, and final histopathological examination confirmed our diagnosis.

The prognosis of patients with SFTs who undergo surgical treatment is good; however, aggressive SFTs can recur, which makes patient follow up highly recommended.[1]

Conclusion

The radiological appearances of pelvic SFTs are variable and can mimic those of other intra-pelvic tumors. CT and MRI can aid the differential diagnosis of these lesions, providing useful information which needs to be supported by histopathological analysis.

Abbreviations

SFTs, solitary fibrous tumors; CT, computed tomography; MRI, magnetic resonance imaging; T1w, T1-weighted; T2w, T2-weighted.

Ethics Approval and Consent to Participate

This case report was approved by the local institutional review board (Ethical Committee "Carlo Romano," University of Naples Federico II). The patient has provided written informed consent for the case details and accompanying images to be published.

Disclosure

The authors report no conflicts of interest for this work.

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