J Surg Med. 2024;8(12):211-214.
https://doi.org/10.28982/josam.8108

JOSAM

Journal of Surgery and Medicine

e-ISSN: 2602-2079

Case report
https://jsurgmed.com/

A rare entity: Giant pelvic dedifferentiated solitary fibrous tumor

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Informed Consent
The authors stated that the written consent was obtained from the patient presented with images in the study.

Conflict of Interest
No conflict of interest was declared by the authors.

Financial Disclosure
The authors declared that this study has received no financial support.

Published
2024 December 19

Abstract

Solitary fibrous tumors are a rare soft tissue tumor of mesenchymal origin with variable behavior. The clinical presentation of solitary fibrous tumors is related to the local mass effect, i.e., the size and localization of the lesion. Although they can occur anywhere in the body, they often originate on lined surfaces within a cavity [pleura, peritoneum, dura]. Such tumors occur in only four to six people out of ten million people annually. Since it is such a rare entity, solitary fibrous tumors may not be considered in the differential diagnosis list when it occurs in uncommon localizations. Solitary fibrous tumors are the "great simulator" of soft tissue tumors. The diagnosis is difficult to make and often requires a multidisciplinary approach integrating clinical, radiological, immunohistochemical and genetic factors. Radiological imaging is usually the first step in the detection of these lesions. Computed tomography and magnetic resonance imaging methods allow us to determine the localization of lesions, assess their size and internal structure, as well as their relationship with adjacent anatomical structures and detect distant metastases. The most important treatment method is excisional surgery. Although radiotherapy and chemotherapy are thought to be effective in reducing recurrence, the global efficacy of these treatment methods has not been clearly demonstrated. In addition, randomized prospective studies with large patient populations evaluating the efficacy of other treatment modalities, such as immunotherapy and targeted therapy, are needed. We reported the CT and MRI findings of a 63-year-old patient who presented with complaints of abdominal distension and was diagnosed with pelvic dedifferentiated solitary fibrous tumor.

Keywords: solitary, fibrous, tumor, giant, pelvic

Introduction

Solitary fibrous tumors [SFTs] are a mesenchymal tumor occurring mainly in the pleura [1]. SFTs occur equally in men and women between the ages of 40 and 70 [2]. The rare tumor accounts for less than 2% of all soft tissue tumors [3]. Up to 6% of SFTs are localized in the pelvic region [4].

The symptoms of SFTs show variability according to the localization of origin [5]. Most SFTs can be asymptomatic at presentation, but large sizes can cause abdominal pain, bowel obstruction, and urinary symptoms [6].

In this article, we aimed to present CT and MRI findings of a rare case of giant pelvic located dedifferentiated SFT.

How to cite: Aytaç A, Kasapoğlu H, Keyik BY, Başbuğ M, Turan G. A rare entity: Giant pelvic dedifferentiated solitary fibrous tumor. J Surg Med. 2024;8(12):211-214.

Case presentation

A 63-year-old male patient was admitted to the emergency department with abdominal pain. There was nothing noteworthy in the patient's background. Abdominal examination revealed tenderness in the pelvic region and a contrast-enhanced abdominal CT scan was performed. A heterogeneous dense mass filling the pelvis and extending to the midline of the abdomen was observed (Figure 1). Hydroureteronephrosis secondary to obstructive retention in the right renal collecting system was detected (Figure 2). Contrast-enhanced MRI was performed to elucidate the origin of the mass lesion. A retroperitoneal mass was seen, with an intermediate signal on T1-weighted sequence, a hyperintense signal on T2-weighted sequence, lobulated contour, including cystic-necrotic areas, and well contrasted (Figure 3-6). Diffusion-weighted images [DWI] showed diffusion restriction (Figure 7). No pathological lymphadenopathy was detected. With these findings, a retroperitoneal mesenchymal tumor with malignant features was considered. A diagnostic laparotomy was performed. Histopathology was reported as dedifferentiated SFT (Figure 8). The patient was followed up with neoadjuvant chemo-radiotherapy before surgery.

Figure 1: Sagittal section contrast-enhanced abdominal CT image: There is a heterogeneous and densely contrasted mass lesion, approximately 19x17x10 cm in size [within the stars], filling the retrovesical space and extending cranially up to the supraumbilical level. The bladder is displaced anteriorly [blue arrow]. Fat planes cannot be seen between the prostate gland [red arrow].

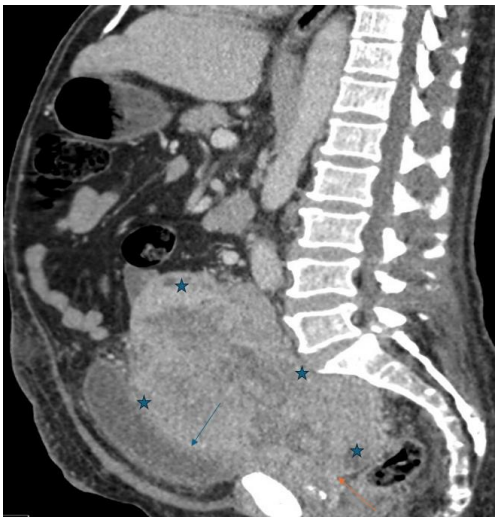


Figure 2: Coronal section contrast-enhanced abdominal CT image: There is grade 1-2 dilatation of the intrarenal collecting system on the right [blue arrow] due to compression of the heterogeneous mass lesion [stars] filling the pelvis.

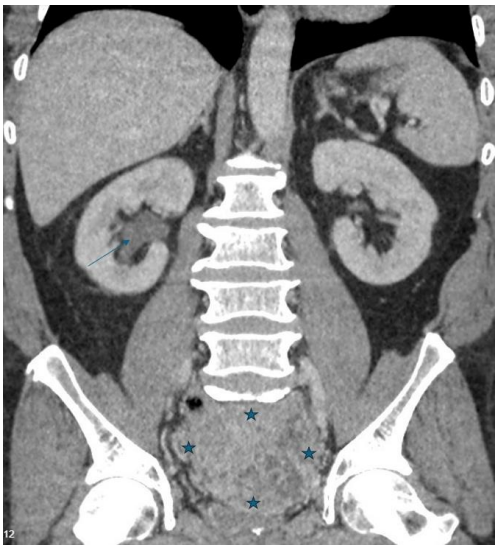


Figure 3: Sagittal section pelvic MRI T2A image: There is a large mass lesion [in stars] filling the pelvis in the retrovesical area with cystic/necrotic [blue arrows] hyperintense foci, solid components are hypointense, and the bladder is displaced anteriorly [red arrow].

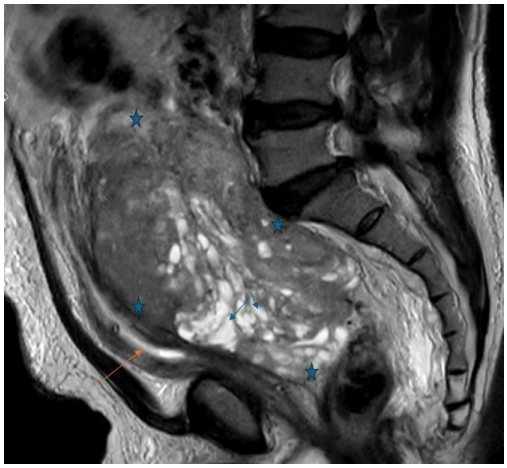


Figure 4: Axial section pelvic MRI T1A image: Mildly heterogeneous mass lesion with solid components with intermediate signal in the pelvis [within the stars].

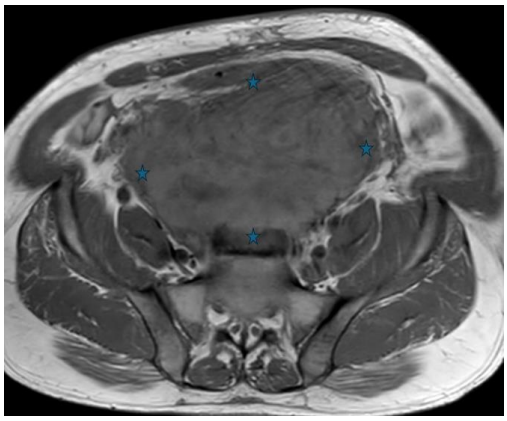


Figure 5: Coronal section pelvic MRI fat-suppressed T2A image: Hypointense thin fibrous capsule is observed at the margins of the lesion [blue arrows].

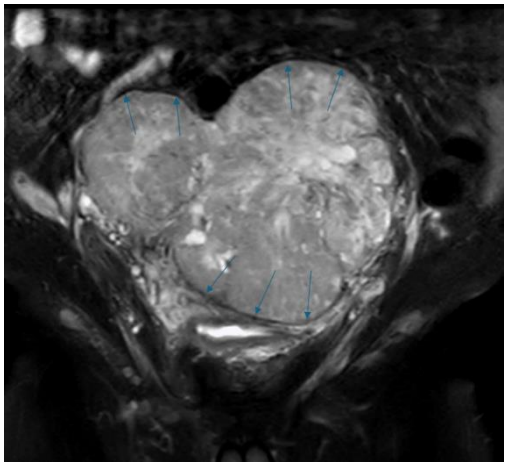


Figure 6: Sagittal section pelvic MRI postcontrast fat-suppressed T1A sagittal image: The mass lesion is well circumscribed and lobulated contoured with intense contrast enhancement in the solid components.

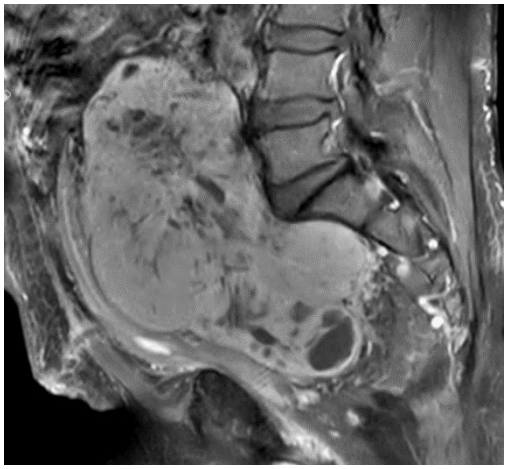


Figure 7: On diffusion weighted images, the mass is hyperintense on 1.b:1000 image, 2. ADC map as hypointense [ADC value average 0.90 0.95x0.001xmm²/sec] and diffusion restriction was observed, more prominent in some regions.

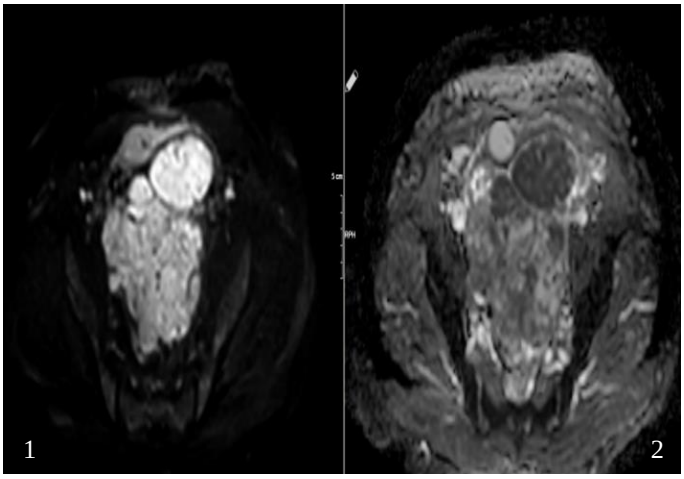
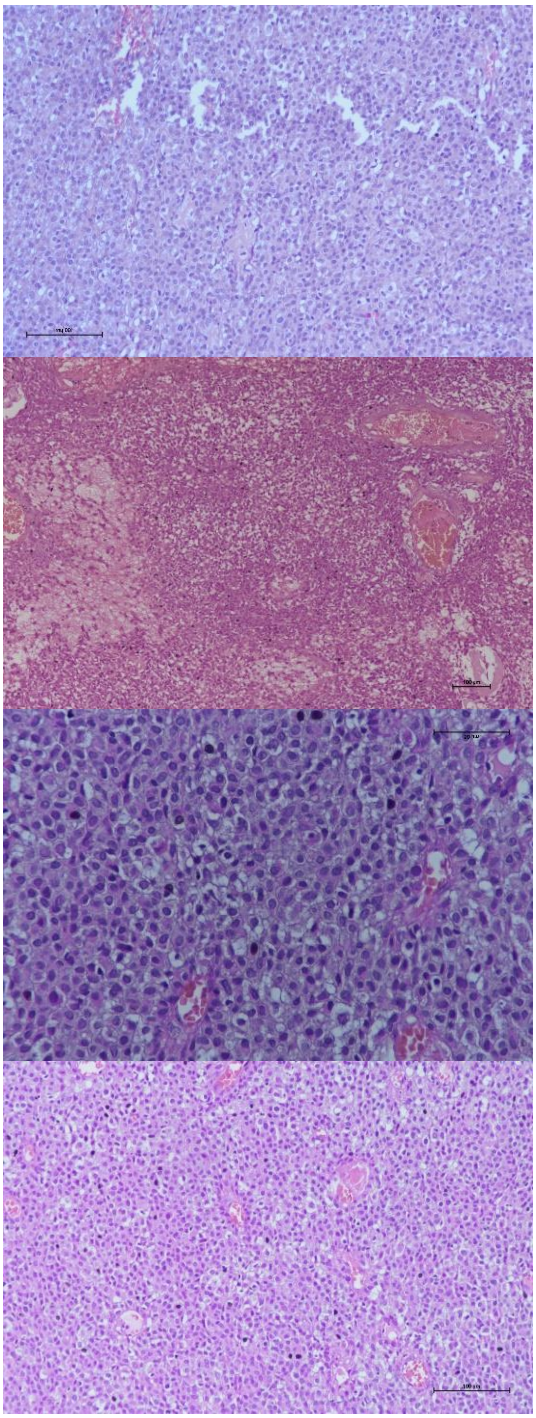


Figure 8: The histopathologic specimen images of the lesion is presented.



Discussion

SFT consists of spindle cells of fibroblastic origin [6]. Although most of them are benign, 12-22% may be malignant [7]. Histopathologically, malignant SFTs have hypercellularity, atypia, ≥4/10 mitosis rate, necrosis and infiltrative borders [8,9]. The definitive diagnosis of SFTs is currently supported by immunohistochemical staining and genetic studies [8,9]. In this case, BCL2, CD34, CD99, STAT6, ki67 positivity was detected.

The radiological appearance of SFTs is variable and nonspecific [9]. On CT scans, the majority of SFTs show marked contrast enhancement in the arterial and portal venous phase [9,10]. If the fibrous component is predominant, contrast enhancement becomes more prominent in the delayed phase [10]. Large collateral feeding vessels may be seen in one-third of all cases [10]. Heterogeneous contrast enhancement and cystic, necrotic, and hemorrhagic components are more common in aggressive SFTs [5,10]. These findings may help predict tumor behavior and guide treatment management.

MRI is a complementary examination in terms of characterizing the lesion and revealing its extension and relationship with surrounding structures [11]. SFTs show variable signal characteristics on MRIs according to the different amounts of solid and cystic components within them [10,12]. Fibrous components show hypointense signals in T1AG and T2AG series, while myxoid and cystic components show hyperintense signals in T2AG. Hemorrhagic areas may also show hyperintense signals on T1AG [11]. Radiological differentiation is difficult due to overlapping appearances in benign or malignant lesions [13]. In recent years, studies have reported that a low apparent diffusion coefficient (ADC) value in DWI may be beneficial in terms of malignant transformation, but the threshold value has not yet been established [13,14]. In addition, the differential diagnosis of SFTs with malignant features is broad [such as angiosarcoma, leiomyosarcoma, desmoid tumor, mesothelioma and uterine neoplasms] [15]. In our case, the definitive diagnosis could be made histopathologically.

Conclusion

In conclusion, CT and MRI help differentiate SFT from gynecological and rectal masses, which are more common in the pelvic region, and allow for preoperative evaluation of the relationship and invasion of surrounding organs. Histopathological evaluation is required for definitive diagnosis.

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