Radiological Case Report / Caso Clínico

Pelvic Solitary Fibrous Tumor: A Case Report with Imaging and Anatomopathological Review of the Literature

Tumor Fibroso Solitário Pélvico: Relato de um Caso Clínico com Revisão Imagiológica e Anatomopatológica da Literatura

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Abstract

Nowadays, routine pelvic imaging in female patients, primarily conducted through ultrasound (US), is a commonly used method to search for uterine and adnexal abnormalities. Occasionally, during these examinations, non-gynecologic findings may be identified. Even with more advanced techniques, such as magnetic resonance imaging (MRI), determining the exact diagnosis may be challenging. In this article, by presenting a clinical case with mention to the ultrasound, MRI, surgical specimen and pathological findings, we focus our attention on a solitary fibrous tumor of the pelvis, a lesion that, although rare, should be considered in the differential diagnosis of pelvic asymptomatic lesions.

Keywords

Solitary fibrous tumors; Pelvic neoplasms; Magnetic resonance imaging; Tomography X-ray computed; Ultrasound.

Resumo

Atualmente, a ecografia é amplamente utilizada na realização de exames de imagem de rotina da pelve feminina, visando investigar alterações no útero e anexos. Por vezes, são ocasionalmente identificados achados não ginecológicos, conhecidos como incidentalomas. Apesar do avanço dos métodos de imagem, nomeadamente através da Ressonância Magnética (RM), o diagnóstico preciso de incidentalomas pélvicos pode ainda ser desafiante. Neste artigo, ao apresentar um caso clínico com menção a achados ecográficos, de RM, da peça cirúrgica e do diagnóstico anatomopatológico, focamos a nossa atenção no tumor fibroso solitário da pelve, uma lesão rara que deve ser considerada no diagnóstico diferencial de lesões pélvicas assintomáticas.

Palavras-chave

Tumores fibrosos solitários; Neoplasias pélvicas; Ressonância magnética; Tomografia computadorizada por raios X; Ultrassom.

Case Presentation

A 44-year-old woman came to our institution for further workup of a pelvic mass found during a routine endovaginal US scan. The patient was asymptomatic, with no relevant laboratory anomalies and her past medical history was unremarkable. US revealed a round and well-circumscribed mass, with both solid and cystic components, in close relation but separate from the ipsilateral adnexa (Fig. 1A). It measured around 5.1 cm. A MRI was performed for further evaluation, which confirmed the extra-adnexal origin of the pelvic mass, located anterior-externally to the left ovary and adjacent to jejunal bowel loops and the bladder dome (Fig. 1B,C). It also confirmed both the solid component, mildly hyperintense on T2-weighted images (T2WI), and the cystic areas, highly hyperintense on T2WI (Fig. 1B,C). No intrinsically high-signal foci on T1-weighted images (T1WI) was identified (Fig. 1D). The lesion exhibited restriction with high signal intensity in Diffusion-Weighted Imaging b800 (DWI) and low values on ADC map (Fig. 1E,F). A unique acquisition after intravenous contrast administration was performed, on portal-venous phase, which revealed solid areas displaying significant enhancement (Fig. 1G). A small amount of ascites was present (Fig. 1B,C). No internal calcifications, pelvic lymphadenopathy or other relevant findings were present.

Due to the anatomic close relation of the mass to the small bowel, its round and well-defined limits and the presence of heterogeneous enhancement due to cystic change, the diagnosis of a jejunal gastrointestinal stromal tumor (GIST) was suggested. The patient underwent surgical excision after proper staging with abdominopelvic computed tomography (CT) scan and chest x-ray (not shown) that did not reveal signs of metastatic disease. The pathological study of the surgical specimen was not concordant, revealing a pelvic omental Solitary Fibrous Tumor (SFT) (Fig. 2,3,4,5). The peritoneal fluid cytology was negative for neoplastic cells.

Discussion

Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms that were originally described in 1931 by Klemperer and Rabin in a series of pleural neoplasms.[1] Only in 1991, the first case of extra-thoracic SFT was published.[2] Since then, SFTs have been reported at almost every anatomic site, involving mostly the pleura (30% of cases), meninges (27% of cases), abdominal cavity (20% of cases), trunk (10% of cases), extremities (8% of cases) and head & neck (5% of cases).[3] They exist on a spectrum from benign to malignant lesions.


Abbreviations: computed tomography (CT); Diffusion-Weighted Imaging (DWI); gastrointestinal stromal tumor (GIST); magnetic resonance imaging (MRI); T2-weighted images (T2WI); T1-weighted images (T1WI); ultrasound (US).
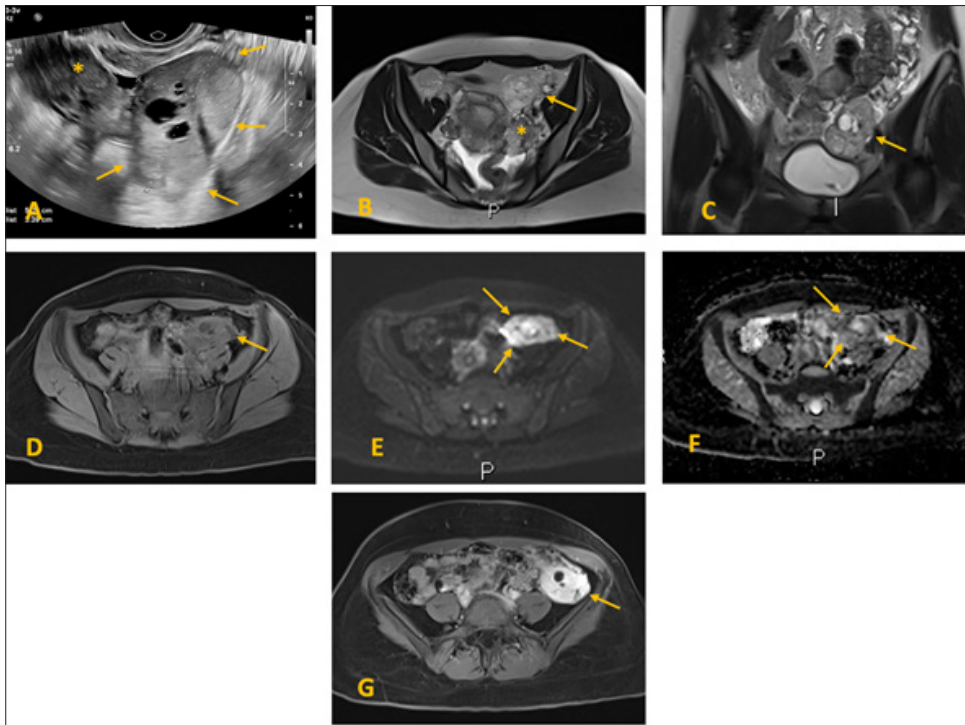
Figure 1 – Pelvic Solitary Fibrous Tumor in a 44-year-old woman. On (A) Endovaginal US, a round and well-circumscribed heterogeneous mass (arrows) in close relation but separate from the ipsilateral adnexa (*) was revealed; (B) Axial and (C) Coronal T2WI, confirming the extra-adnexal (ovary highlighted with *) origin of the lesion (arrows) and showing its both solid and cystic components; (B) small amount of pelvic ascites is seen; On axial unenhanced fat-suppressed T1WI (D), the lesion is heterogeneously iso and hypointense (arrow); it has high signal intensity in DWI b800 (E) and low values on ADC map (F); (G) Axial contrast-enhanced fat-suppressed T1WI on the portal-phase showing avid enhancement of the lesion.
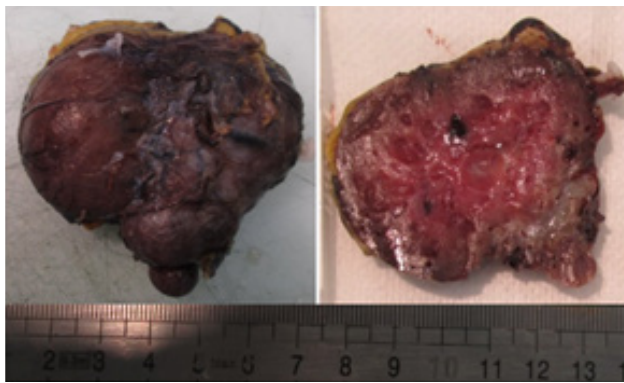

Figure 2 – Gross appearance of the surgical specimen: it is a well-defined brown mass (left); the cut surface was homogeneous purple-brownish and spongy, with some hemorrhagic and cystic areas (right).
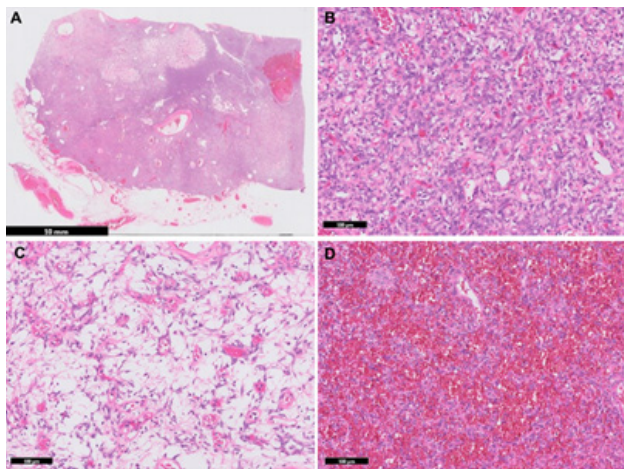
Figure 3 – Histology of the surgical specimen. (Low magnification, A) Histologically, it was a well-defined but non-encapsulated cellular proliferation localized in the fibroadipose tissue; (Higher magnification, B, C and D) It is a heterogeneous lesion, with areas with high cellularity (B), areas with loose connective tissue with few cells (C) and areas with hemorrhage (D).
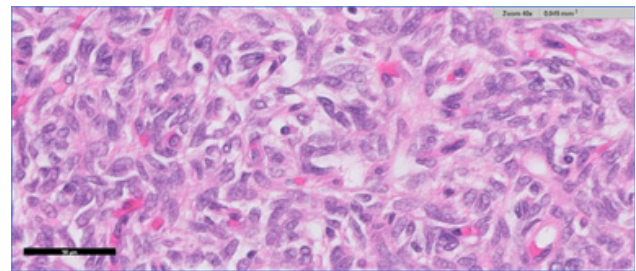
Figure 4 – Histology of the surgical specimen. At higher magnification the cells have a spindly cytoplasm, with an oval to elongated nucleus, with no pleomorphism and no nucleolus. No mitotic figures are observed.
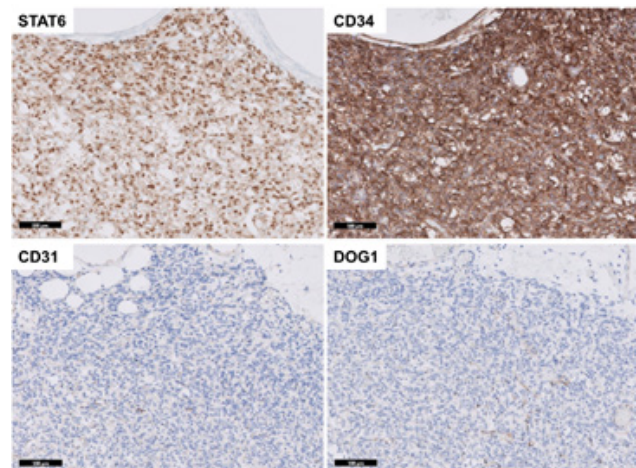
Figure 5 – Immunohistochemistry analysis. The tumor cells display diffuse and intense nuclear expression of STAT6, diffuse and intense expression of CD34 and do not show any expression of CD31 nor DOG1.

Regardless of their tissue of origin, SFTs commonly occur between the 5th and 6th decades of life, being rare in children. They have no gender predilection and no established risk factor is associated with their development. The majority of these neoplasms are slow-growing and are typically discovered incidentally during imaging studies. When symptoms occur, they are due to the mass itself (palpable

mass) or its pressure effects (e.g., ureteral compression, urinary frequency).[3,4,5] There is an association between large SFTs and paraneoplastic syndromes, (e.g. Doege-Potter syndrome, a refractory hypoglycemic state resulting from excessive secretion of insulin like growth factor-2) although these are less likely with abdominopelvic SFTs compared to pleural ones.[4]

Regarding pathologic features, classic SFTs appear as well-circumscribed and partially encapsulated lesions with a multinodular, whitish, firm-cut surface. These tumors are highly vascular and have a propensity to undergo hemorrhage, necrosis, and myxoid degeneration. Histologically, SFTs are characterized by a variable admixture of two architectural patterns: one composed of haphazardly arranged spindle-shaped to oval cells lying in a variably collagenous stroma (patternless); the other composed of spindle-shaped or oval cells interspersed in a network of branching and often hyalinized staghorn small vessels (hemangiopericytoma). Most SFTs have low mitotic counts and show little nuclear pleomorphism.[3,4] Increased mitotic activity ($\geq$4/10HPFs or > 2mitoses/2 mm2) and nuclear pleomorphism are associated to malignancy.[3,4]

Nowadays, even in malignant cases of SFTs, accurate diagnosis is achievable by identifying their hallmark feature -NAB2-STAT6 gene fusion-, which is best detected by multiplexed sequencing assay. Immunohistochemical detection of STAT6 is also a highly sensitive (98%) and specific (85%) marker. Other sensitive immunohistochemical markers classically used, although non-specific, are CD34, CD99 and BCL-2.[3] They are not reactive to S-100 and DOG1, differentiating them from Schwannomas and GISTs, respectively.[3]

Imaging features of SFTs in the abdomen or pelvis tend to be nonspecific, implying biopsy or complete excision and histopathologic examination to establish the diagnosis. Nevertheless, some unifying features suggest the diagnosis: commonly, we find a single, well-defined, round or lobulated large mass that displaces, rather than invading, adjacent structures, such as the bowel, urinary bladder, ureter and uterus.[4,5,6]

On unenhanced CT, small lesions demonstrate homogeneous soft tissue attenuation and larger lesions are heterogeneous due to intra-lesional low attenuation areas that represent myxoid/cystic degeneration. Tumor calcifications are rare and can be seen in large-sized masses.[4,6]

On MRI, these lesions are hypo to isointense on T1WI and show variable signal intensity on T2WI, linked to the relative amount of mature fibrous tissue and myxoid/cystic degeneration. Tumors with a predominance of collagen and fibroblasts have low signal on T2WI, while degenerated lesions have high T2 signal.[4,6] A key imaging finding at MRI is large vessels associated with flow voids that are best appreciated at T2WI.[7]

Another unifying feature is that SFTs show intense heterogeneous enhancement on both contrast-enhanced CT and MRI. This is attributed to the hypervascular areas that exhibit marked early enhancement and the areas of necrosis or cystic/myxoid degeneration that show no enhancement. Mild enhancement in the arterial phase with increasing enhancement in the delayed phase correlates with the fibrous or collagenous stroma.[4,5,6]

Although US is nonspecific in evaluating appearances of SFTs, it can serve as a tool, particularly in the pelvis, for excluding lesions of genital origin. When the mass is closely related to one of the ovaries or the uterus, gentle pressure can be applied with the transducer, preferably through an endovaginal approach, to separate the mass from the ovary/uterus and establish its extraovarian origin.[8]

Fluorodeoxyglucose positron emission tomography (PET) PET/CT is being used for the assessment of multifocal disease, distant metastases and disease recurrence after resection. This technic highlights the hypercellular areas within the tumor that appear as increased uptake foci.[4,5]

Until today, many groups have proposed models of stratifying the malignant potential of SFTs based on histopathology, which could help guide management and follow-up on this rare entity that exists on a spectrum from benign to malignant. Nevertheless, even when these tumors are considered benign with histologic assessment, they can recur locally or metastasize. Extrathoracic site is an independent predictor of poor prognosis, being associated with a greater risk of recurrence. Due to their rarity and lack of randomized control trials, there is no global consensus on treatment and post-treatment surveillance of SFT, though surgical excision with clear margins and imaging follow-up is recommended. Adjuvant radiotherapy has been suggested to improve the local control of the tumor.[3,4]

Conclusion

The female pelvis is a complex anatomical region with diverse spaces and organ systems. Although primarily focused on gynecological conditions, pelvic imaging studies might uncover non-gynecologic findings, namely solitary fibrous tumors. These are rare slow-growing masses that have unifying, although non-specific, imaging features. The definitive diagnosis is made by pathological examination. Post-treatment imaging surveillance is advised, as local recurrence or distant metastasis can occur.

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