

Review Article

Solitary Fibrous Tumor of the Female Pelvis – Clues to the Radiological Diagnosis

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Abstract:

Solitary fibrous tumors were initially described in the pleural cavity and they were thought to have mesothelial origin. Nowadays, these tumors are known to have origin in the mesenchymal cells and they are ubiquitous, with both pleural and extrapleural distribution. There are many extrapleural localizations, being the pelvis one of the rarest and the most challenging to diagnose. Usually, solitary fibrous tumors are clinically insidious and manifest as a slow-growing large tumor. At imaging, these tumors have different characteristics, and computed tomography and magnetic resonance play an important role in their diagnosis. Solitary fibrous tumors show heterogeneous enhancement on contrast-enhanced computed tomography, intermediate signal on T1-weighted images, hypointense on T2-weighted images and have intense heterogeneous enhancement in dynamic sequences. When they appear in the female pelvis, it is essential to recognize their radiological characteristics, in order to differentiate it from others tumors, like pure stromal ovarian tumors or Brenner tumors. This study reviews the female pelvis solitary fibrous tumors' imaging findings and its differential characteristics from more common tumors, by describing illustrative confirmed cases.

Keyword: Solitary fibrous tumor, female pelvis, MRI, CT.

Introduction:

Solitary fibrous tumors (SFTs) were thought to have mesothelial origin with exclusive involvement of the serosa surfaces, like pleura, peritoneum and pericardium⁽¹⁾. Histogenesis, tumor distribution and histologic heterogeneity of SFTs are better understood thanks to advances in pathology, and now they are known as ubiquitous mesenchymal neoplasms. SFTs can be benign or malignant, and account for less than 2% of all soft-tissue tumors⁽²⁾.

Because SFTs can arise from any part of the body, extrapleural SFTs are more common than pleural SFTs, and they usually appear as a large, slow-growing soft-tissue mass^(3,4). Symptomatic SFTs occur due to compression of the parenchyma and adjacent viscera. Magnetic resonance (MR) and other cross-sectional imaging studies, like Computed Tomography (CT) are fundamental for the detection and characterization of the SFTs⁽⁴⁾.

In this study, we review CT and MR findings of a series of cases of SFTs histologically confirmed.

Body Text

We review the solitary fibrous tumors of the female pelvis diagnosed in our institution in last five years. All of the selected cases were surgically removed and were histological confirmed as fibrous tumors.

Result and discussion

Clinical Features, Pathology and Management

Normally, SFTs have no sex predilection and present in middle-age patients⁽⁵⁾.

Pelvic SFTs manifest clinically as slow-growing mass, often asymptomatic, but can present as pain and palpable mass, or with symptoms regarding the compression of adjacent organs, like bowel obstruction, constipation or urinary symptoms^(4,6). In a few cases of SFTs, the patients present symptomatic hyperglycemia, because of excessive production of insulin

growth factor by the tumor⁽⁴⁾.

The mesenchymal origin of the SFTs is confirmed by using immunohistochemical analysis with positive staining for CD34 and bcl-2⁽⁶⁾. Histologically, SFTs result from spindle cells within a background of patternless collagen stroma, show highly vascularization, and have a predisposition to undergo necrosis and myxoid degeneration⁽⁵⁾. At pathology, malignant SFTs show marked cellular atypia, high mitotic activity, and infiltrative greater cellularity⁽⁶⁾. Previously, many SFTs were errantly characterized as hemangiopericytomas⁽⁷⁾.

The treatment of choice for SFTs is surgical excision and the 5-year survival rate is close to 100%⁽⁴⁾. However, there had been some reports of higher local recurrence rate with extrathoracic SFTs⁽²⁾.

Imaging Findings

Since these patients have insidious and non-specific symptoms, cross-sectional imaging studies play an important role in the diagnosis of extrathoracic Solitary Fibrous Tumors, like the pelvic ones, because regularly these tumors are an unexpected finding.

Computed tomography (CT) is usually the first modality of choice for investigation of these patients because it is an accessible and widespread exam modality, and it encompasses the thorax, the abdomen and the pelvis.

On non-enhanced-CT the density of the SFTs depends on the amount of collagen: they appear hypodense when lesions have few collagen⁽⁵⁾. Extrapleural SFTs are well-circumscribed masses on CT that can displace the adjacent organs and viscera⁽³⁾ and cause related symptoms, like urinary or bowel obstruction^(3,4,6,8) in pelvic SFTs, when the bladder, the uterus (Fig. 1) or the rectum are displaced. On contrast-enhanced CT, pelvic SFTs can appear as well-circumscribed hypervascular masses with hypoenhancing and non-enhancing areas of necrosis or cystic changes (Fig. 2)⁽⁹⁾. Calcifications are usually a rare feature of these tumors⁽⁶⁾, but they could be seen within SFTs⁽⁹⁾.

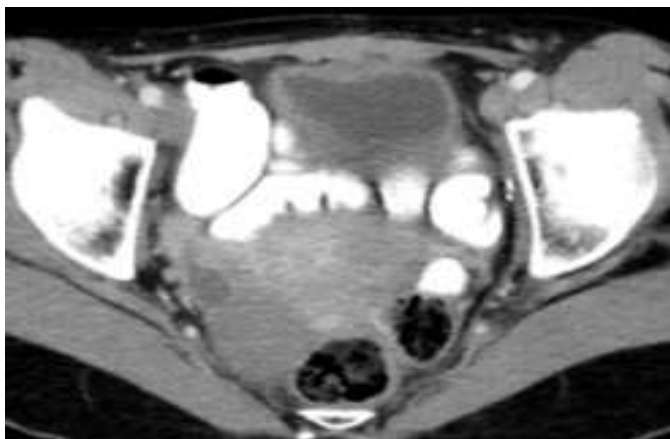


Figure 1. Computed Tomography findings in Solitary Fibrous Tumor histologically confirmed – Axial contrast-enhanced CT shows a right posterior pelvic tumor with poorly enhancement.



Figure 2: Computed Tomography findings in Solitary Fibrous Tumor histologically confirmed – Coronal plane shows the relationship between the tumor and the uterus.

Regarding the potential malignancy of the SFTs, the pattern of enhancement, namely the intense heterogeneous enhancement, is not accurate to differentiate malignant from benign SFTs⁽⁹⁾. At magnetic resonance (MR) imaging, SFTs appear as an isointense lesion on T1-weighted images (Fig. 3) and a heterogeneous hypointense lesion on T2-weighted images⁽⁴⁾. However, the reality is that these tumors may show heterogeneous mild hypointense signal or homogeneous isointense signal on T1 (Fig. 4) and T2-weighted images, and they can present with some linear areas of hyperintensity on T1 and T2 sequences^(6,8). These various imaging patterns reveal the heterogeneity of this group of tumors.

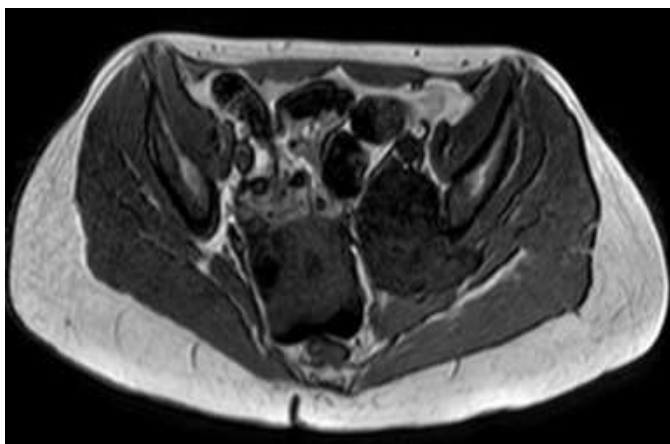


Figure 3: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – T1-weighted images: axial plane T1-weighted image shows a markedly hypointense pelvic tumor.

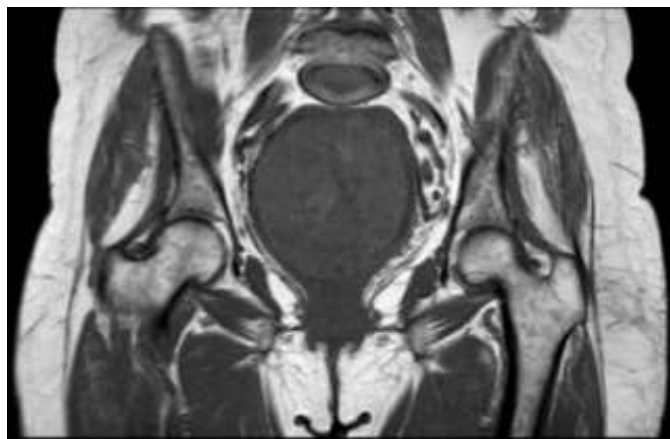


Figure 4: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – T1-weighted images: coronal plane T1-weighted image shows an isointense tumor.

On T2-weighted images, most SFTs have heterogeneous hypointense signal with some intra or extra-tumoral flow voids (Fig. 5), related to the prominent vascular structures^(8,9). Fibrous tissue, collagen content, low cellularity and reduced proton mobility appear as hypointense areas within SFTs on T2-weighted images, and these features are responsible for the “chocolate chip cookie” appearance^(5,9) (Fig. 6).

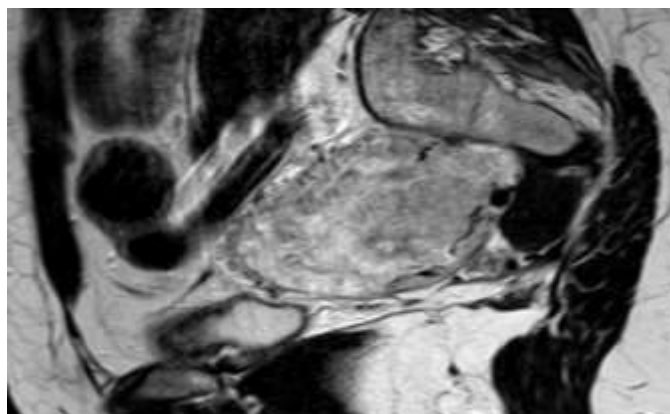


Figure 5: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – T2-weighted images: sagittal plane T2-weighted image shows a heterogeneous pelvic tumor with peripheral flow voids signals, representing vascular structures.

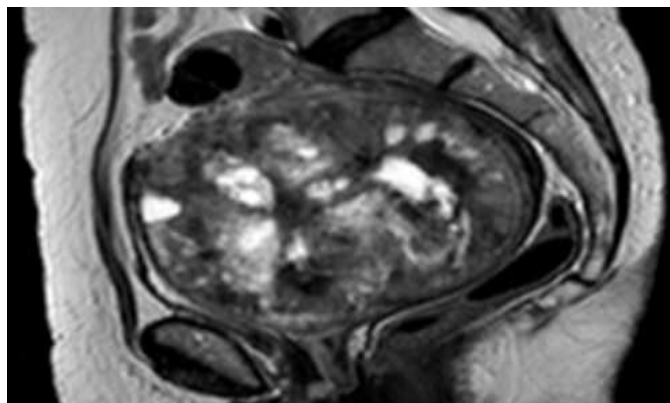


Figure 6: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – T2-weighted images: axial plane T2-weighted images shows a heterogeneous tumor with areas of hyperintensity and areas of hypointensity.

After administration of gadolinium, SFTs typically show intense enhancement related to its high vascularized⁽⁴⁾. The tumor enhancement on dynamic sequences can be heterogenous with hypoenhancing central areas^(4,6) (Fig. 7) or

show some homogeneity⁽⁶⁾.

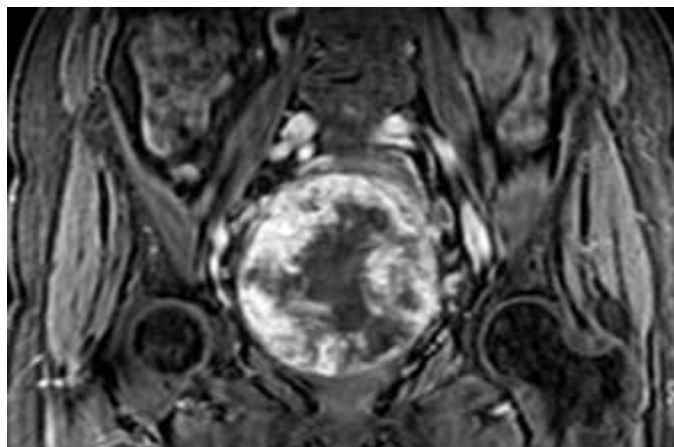


Figure 7: Magnetic Resonance in Solitary Fibrous Tumors histologically confirmed – Coronal plane T1- post-gadolinium image showing the intense and heterogeneous enhancement of the tumor.

SFTs' behavior on dynamic post-contrast images depend on the tumor components, and those which have more fibrous and collagenous stroma show progressive enhancement in the venous and late phases on post-gadolinium-T1-weighted images^(4,8). On the other hand, we can differentiate hypercellular areas that enhance moderately after gadolinium administration, from hypocellular components, such as necrosis, cystic or myxoid, that do not enhance on post-gadolinium-T1-weighted images⁽⁴⁾.

SFTs features on diffusion-weighted images and ADC maps are rarely referred in the literature. We found that SFTs usually show low restriction on b800 DWI-weighted images, with corresponded low-attenuation values on ADC map (Figs. 8A and 8B), reflecting the hypercellularity that some of these SFTs show.

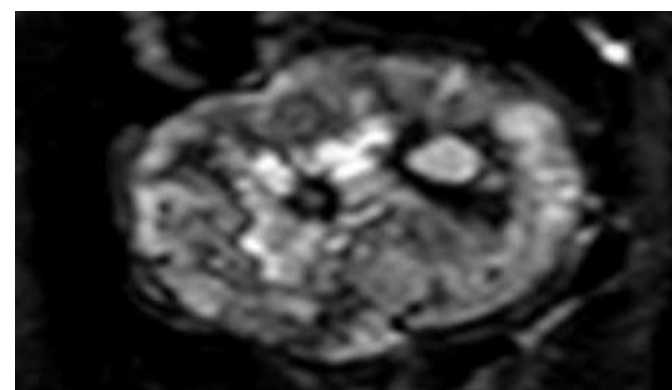
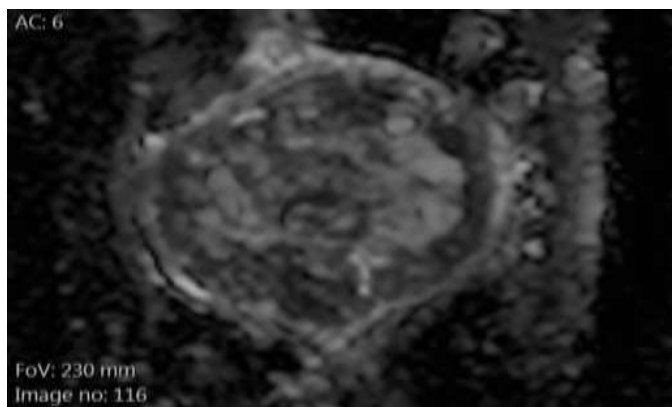


Figure 8A and 8B: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – (A) DWI image showing a

tumor with low signal intensity on b800 DWI image; (B) ADC map showing a tumor with low-value signal on the ADC map.

Conclusion

The differentiation between pelvic SFTs and other pelvic tumors rely on some imaging characteristics.

Usually, SFTs are hypervascular tumors and can be confused with some rare retroperitoneal tumors, such as hemangioendotheliomas, angiosarcomas and angiomyxomas⁽⁴⁾. Regarding its fibrous components, the differential diagnosis include mesothelioma, malignant fibrous histiocytoma, desmoid tumor, ovarian Brenner tumor, fibroma, cellular fibroma, and uterine leiomyoma⁽⁴⁾.

In the female pelvis, the primordial differential diagnosis are ovarian tumors, namely stromal tumors (fibroma, cellular fibroma and thecoma), Brenner tumors and epithelial tumors (adenofibromas). Beak sign, lateral displacement of the iliac vessels (Fig. 9) and posterior or posterolateral displacement of the ureter are imaging features of an ovarian lesion, and can be the clues to differentiate these ovarian tumors from SFTs. Fibroma and cellular fibroma are ovarian pure stromal tumors that show hypoenhancement on contrast-enhanced images⁽¹⁰⁾ and Brenner tumors are epithelial-stromal tumors that show more enhancement and produce androgen hormones⁽¹⁰⁾.

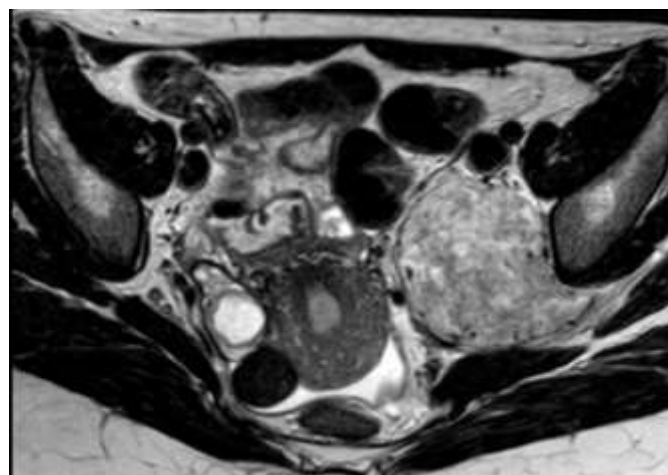


Figure 9: Magnetic Resonance in Solitary Fibrous Tumor histologically confirmed – Axial plane T2-weighted image showing the right ovary with some follicles and a left-sided solitary fibrous tumor that deviates the iliac vessels anteriorly, reflecting the posterior and not ovarian origin of the tumor.

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