Case Report/Caso Clínico

Solitary fibrous tumor of the uterine cervix – about a case
Tumor fibroso solitário do colo do útero – a propósito de um caso

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Abstract
Solitary fibrous tumors are rare mesenchymal neoplasms. Although first reported in the pleura, they were described at extrapleural sites, but they are uncommon in the female genital tract (less than 30 cases reported). We present a case of solitary fibrous tumor arising from uterine cervix with a benign course, as well as its histopathological and immunohistochemical features that helped establishing the diagnosis.

Keywords: Solitary fibrous tumor; Female; Genital neoplasms; Uterine cervix.

INTRODUCTION
Solitary fibrous tumors (SFTs) are uncommon mesenchymal neoplasms with an incidence rate of 2.8 per 100,000¹.

Although originally reported for the first time in 1931 by Klemperer and Rabin in the pleura, extra pleural locations of these tumors have been described all over the years². Nevertheless, their occurrence in the female genital tract is extremely rare (about 30 cases described in the literature)¹,³.

In the past, STF have been given many different names including benign mesothelioma, localized mesothelioma, solitary fibrous mesothelioma and localized fibrous tumor⁴. Fortunately, the World Health Organization (WHO) classification of soft tissue sarcomas published in 2013, helped further differentiation within the sarcomas. As so, SFT are classified as fibroblastic/myofibroblastic tumors and sub classified as intermediate behavior (rarely metastasizing)⁵.

Due to the low incidence of STF, most data is provided by small series and case reports which made diagnostic and treatment strategies more difficult to establish⁶.

We report one of the single cases described in the literature about STF of the uterine cervix.

An informed consent was obtained from the patient and approved by the Hospital Ethics Committee.

CASE REPORT
A 68-year-old, G3P2 white woman was referred to our institution for abnormal cervical cytology (ASC-US, atypical squamous cells of undetermined significance).

The patient had a history of hypertension, diabetes, hypercholesterolemia and an orthopedic hand prosthesis (due to traumatic amputation).

She denied any kind of abdominopelvic discomfort, dyspareunia, abnormal uterine bleeding or abnormal discharge.

The colposcopy was unsatisfactory due to atrophy and the high risk human papilloma virus (HPV) test was negative.

Physical examination revealed a bulky lesion with 4 cm that was palpable in the posterior surface of the cervix, protruding into the vaginal wall. This accidental lesion detectable during the vaginal touch was not visible neither during speculum examination nor during colposcopy.
On vaginal ultrasound (Figure 1) a lobed solid lesion was identified, measuring 3.7 x 3.5 x 2.9 cm, with central and peripheral moderated vascularization, localized in the posterior surface of the uterine cervix. The lesion was regular and isoechoic when compared to the myometrium.

Laboratory data was unremarkable.

The pelvic magnetic resonance imaging (MRI) showed a vascularized mass, hyper intense in T2, with 4 x 4.7 cm, from the cervix and protruding into the posterior vaginal wall (Figure 2). There was no significant additional findings and no evidence of extra-pelvic neoplastic sites nor metastasis.

A biopsy was performed and revealed a mesenchymal tumor without cytological atypia. An immunohistochemical panel analysis was done and showed positivity for CD 34, CD 99 and vimentin; and negativity for estrogen receptors, EMA (epithelial membrane antigen) and desmin. As so, these histological and immunohistochemical features suggested a benign SFT of the uterine cervix.

After discussing the case it was decided to propose a total hysterectomy with bilateral adnexectomy to the patient, which was accepted.

On macroscopic examination of the tumor it was possible to identify a well delimited, elastic mass with 4.5 x 4.4 x3.5 cm. Microscopically, the neoplasm was characterized by spindle cells on a myxoid stroma without cytological atypia and with a low mitotic rate. Hemangiopericytoma-like vessels were also described and the surgical margins were free of tumor.

Currently the patient is being followed-up and continues asymptomatic (13 months after the surgery). Imagological data shows no evidence of disease, neither the physical exam.

**DISCUSSION**

SFT are tumors with a mesenchymal origin, but their etiology is unknown.

They have an equal gender distribution and may be diagnosed in all ages, although commonly presenting between 20 and 70 years.

These tumors could be found in almost any site of the body, with intra-thoracic being the most common primary location.

On the other side, the most common extra pleural location overall is the abdominopelvic cavity. These tumors generally present with a palpable mass or with lo-
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TABLE I. REPORTED CASES OF UTERINE CERVIX SOLITARY FIBROUS TUMOR

<table>
<thead>
<tr>
<th>References</th>
<th>Age (years)</th>
<th>Tumor size</th>
<th>Pathology</th>
<th>Recurrence/metastasis</th>
<th>Follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nowakowski et al. 2014</td>
<td>45</td>
<td>15 cm</td>
<td>Benign</td>
<td>No</td>
<td>8m AWOD</td>
</tr>
<tr>
<td>Sidebotham et al. 2009</td>
<td>14</td>
<td>2 cm</td>
<td>Benign</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Hasegawa T et al. 1998</td>
<td>78</td>
<td>6 cm</td>
<td>Benign</td>
<td>No</td>
<td>132m AWOD</td>
</tr>
<tr>
<td>Rahimi et al. 2010</td>
<td>68</td>
<td>3 cm</td>
<td>Benign</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Present case</td>
<td>68</td>
<td>4 cm</td>
<td>Benign</td>
<td>No</td>
<td>6m AWOD</td>
</tr>
</tbody>
</table>

AWOD: alive without disease; NA: data non available
SFT treatment. These antiangiogenic drugs could be a promising treatment strategy for SFT.\(^5\)\(^15\)

SFT mostly have an indolent course and benign behavior. But even those tumors considered histologically benign may aggressively recur. Obtaining adequate negative margins has been shown to decrease the rate local disease recurrence and improve survival (10-year survival rate of 89%). Long-term follow-up in small case series have demonstrated local recurrence as low as 8% after complete resection, however, true recurrence rates may be higher as recurrence has been already reported to occur 17 years following resection\(^1,15,16\).

Nevertheless in 10-20% of patients occurs local recurrence or metastasis, with risk factors including positive surgical margins and tumor size greater than 10 cm, as well as malignant features such as more than 4 mitoses per 10 high-power fields, increased nuclear pleomorphism, increased cellularity, and presence of necrosis.\(^3,15,17\) Some investigators also described older age (≥ 55 years), paraneoplastic syndrome, and incomplete tumor resection as indicators of poor outcome.\(^1,18\)

The scarce cases of pelvic SFTs results in absence of a well-defined follow-up protocol. Therefore, close long-term follow-up is recommended with three monthly clinical examination, ultrasound and a yearly cross-sectional MRI or CT scans imaging or as indicated by the clinical symptoms.\(^1,3,7,18\)

Thus we present a rare tumor with a rarer location, which imposed a clinical challenge, since SFT of the female genital tract has only been subject of a few reports, possibly being under-recognized. It could be easily mistaken with fibroid, but the biopsy performed helped in the differential diagnosis previous to the treatment and also helped planning the surgical approach.

This is the 5\(^{\text{th}}\) case of SFT of the uterine cervix reported in the literature.

REFERENCES


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