Primary Solitary Fibrous Tumor in Pelvic Region: Case Report

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Tumor Fibroso Solitário Primário em Região Pélvica: Relato de Caso

Tumor Fibroso Solitario Primario en la Región Pélvica: Relato de Caso

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ABSTRACT

Introduction: Solitary fibrous tumor is a rare mesenchymal neoplasm, originated from CD34-positive interstitial dendritic cells and composed by the juxtaposition of spindle cells. Initial cases were primarily described in the thoracic region, the main site being the visceral pleura. However, cases of solitary extra-pleural fibrous tumors have already been described in the pelvic region demonstrating the possibility of multiple primary sites. **Case report:** A 38-year-old female patient, previously healthy and asymptomatic, seeks medical advice after a routine pelvic ultrasound that showed a heterogeneous image in the right adnexal region, paraovarian, being submitted to surgical resection of the lesion with a diagnosis of solitary fibrous tumor confirmed by immunohistochemistry. Aspects of immunohistochemical diagnosis and surgical treatment were discussed. **Conclusion:** Cases of solitary fibrous tumor in the pelvic region can be discovered through routine pelvic ultrasound. Surgical treatment, with extended resection and negative margins, should be the main objective in cases of solitary fibrous tumor in the pelvic region. Obtaining samples for immunohistochemical analysis is recommended and positivity for CD34 and STAT6 discloses the diagnosis. Relapses can occur in up to a decade of follow-up, and a long period of post-surgical follow-up is recommended.
Key words: solitary fibrous tumors; pelvic neoplasms; case reports.

RESUMO

Introdução: O tumor fibroso solitário é uma neoplasia mesenquimal rara, originada de células intersticiais dendríticas CD34 positivas e composta pela justaposição de células fusiformes. Os casos iniciais foram primariamente descritos na região torácica, e o principal sítio, a pleura visceral. Raramente são descritos casos de tumor fibroso solitário extrapleural em região pélvica, demonstrando a possibilidade de múltiplos sítios primários. **Relato do caso:** Paciente de 38 anos, sexo feminino, previamente saudável e assintomática, procurou orientação médica após uma ultrassonografia pélvica de rotina evidenciar uma imagem heterogênea na região anexial direita, paraovariana, sendo submetida à ressecção cirúrgica da lesão, com diagnóstico de tumor fibroso solitário confirmado por imuno-histoquímica. Discutem-se os aspectos do diagnóstico imuno-histoquímico e do tratamento cirúrgico. **Conclusão:** Os casos de tumor fibroso solitário em região pélvica podem ser descobertos por meio de ultrassonografia pélvica de rotina. O tratamento cirúrgico, com ressecção ampliada e margens negativas, deve ser o principal objetivo nos casos de tumor fibroso solitário em região pélvica. A obtenção de amostras para análise imuno-histoquímica é recomendada, e a positividade para CD34 e STAT6 aponta o diagnóstico. Recidivas podem ocorrer em até uma década de seguimento, sendo recomendado período longo de acompanhamento pós-cirúrgico.
Palavras-chave: tumores fibrosos solitários; neoplasias pélvicas; relatos de casos.

RESUMEN

Introducción: El tumor fibroso solitario es una neoplasia mesenquimatosa rara, originada a partir de células dendríticas intersticiales CD34 positivas y compuesta por la yuxtaposición de células fusiformes. Los casos iniciales se describieron principalmente en región torácica, siendo el sitio principal la pleura visceral. Sin embargo, ya se han descrito los casos de tumores fibrosos extrapleurales solitarios, como en la región pélvica. **Relato del caso:** Paciente femenina de 38 años, previamente sana y asintomática, acude al médico luego de una ecografía pélvica de rutina que mostró una imagen heterogénea en región anexial derecha, para ovárica. La paciente fue tratada con resección quirúrgica de la lesión y tuvo el diagnóstico de tumor fibroso solitario confirmado por inmunohistoquímica. Se discuten aspectos del diagnóstico inmunohistoquímico y del tratamiento quirúrgico. **Conclusión:** Los casos de tumor fibroso solitario en la región pélvica se pueden descubrir mediante una ecografía pélvica de rutina. El tratamiento quirúrgico, con resección ampliada y márgenes negativos, debe ser el principal objetivo en los casos de tumor fibroso solitario en la región pélvica. Se recomienda obtener muestras para análisis inmunohistoquímico y la positividad para CD34 y STAT6 apunta al diagnóstico. Las recaídas pueden ocurrir hasta en una década de seguimiento, y se recomienda un largo período de seguimiento posquirúrgico.
Palabras clave: tumores fibrosos solitarios; neoplasias pélvicas; informes de casos.

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INTRODUCTION

Solitary fibrous tumors (SFT) are rare mesenchymal neoplasms, originated from CD34-positive interstitial dendritic cells and composed by the juxtapositions of spindle cells with dense angiogenesis[1].

Although most SFT are benign, 15% can be malignant characterized by hypercellularity, > 4 mitosis per field, cellular pleomorphism and necrosis. Are more common in the fifth and six decades of life, although can occur in a wide range of ages[1].

SFT were primarily described in the visceral pleura[2]. However, extra-pleural SFT have already been described in the meninges[3], pelvis[4], bones[5], seminal vesicle[6], among others. Those occurring in the pelvis appear to have a more aggressive behavior than those in the pleura, are associated with larger sizes and relative bigger risk of malignancy, mainly for those > 10 cm[7,8].

In general, the incidence is similar for both sexes and mostly in the wide range from 20 to 70 years old and surgical resection is the treatment of choice[2].

A retrospective analysis in a large Brazilian cancer center from 1971 to 2017 described 87 cases of patients diagnosed with SFT primarily at the pelvis[9]. The literature search at PubMed from 2017 to 2022 utilizing the terms *solitary fibrous tumor* and *case report* revealed 20 articles describing SFT in the pelvis in European, Asian and North American countries; the investigation is usually initiated from an incidental post ultrasound or tomography finding and diagnosis after immunohistochemistry analysis of the resected surgical piece[1,2,4,5].

This case report describes a 38-years old Brazilian woman, healthy and asymptomatic who sought medical care after a routine pelvic ultrasound has shown a heterogeneous image of the paraovarian, adnexal right region, submitted to surgical resection and immunohistochemistry confirmation of SFT. The objective is to portray the initial clinical case, the diagnosis and treatment of the patient from the suspected diagnosis of resectable SFT at the pelvis and contribute to the correct management of this rare neoplasm.

CASE REPORT

Healthy female patient, 38-years old, sought medical care after routine ultrasound has shown solid-cystic, complex heterogeneous image at the right adnexal paraovarian, regular and well-defined margins, measuring nearly 4.1x3.2x27 cm. Uterus and ovaries were normal.

Physical rectal and vaginal examination showed palpable tumor in the right pelvic excavation, 1/3 above the vagina, defined margins, barely mobile, without infiltration to the vaginal wall.

Magnetic resonance image of the pelvis identified a well delimited multiseptated cystic lesion at the right pelvic floor in close contact with the right posterolateral wall of the bladder, whose structure was intact. Septa were thick and irregular, enhanced by paramagnetic contrast. The lesion measured approximately 4.2x3.2x3.1 cm (vol.: 21.6 cm³). Hyposignal at T1 and hypersignal at T2 C1 were shown, diffusion without fatty component or hematic content (Figure 1A, B, C). Neither upper abdomen ultrasound nor lab tests presented alterations.

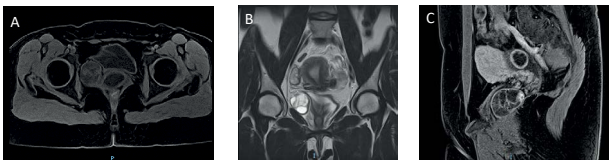

Figure 1. (A) Nuclear magnetic resonance imaging of the pelvic region – coronal cut; (B) Nuclear magnetic resonance imaging of the pelvic region – axial cut; (C) Nuclear magnetic resonance imaging of the pelvic region – sagittal cut

Note: Well-delimited multiseptated cystic lesions were identified at the right side of the pelvic floor in close contact with the right posterolateral wall of the bladder whose structure was intact.

The patient was submitted to exploratory laparoscopy with retrovesical space approach and paravesical at right where a solid tumor was identified and resected with approximately 4x4 cm, frequently hemorrhagic, dissected lengthwise which did not infiltrate the vesical wall (Figure 2). Histopathology by freezing was performed, suggestive of benign tumor, but inconclusive histogenesis. The patient had no post-operative complications.

Histological analysis revealed a short and epithelial spindle cells mesenchymal neoplasm with predominance of hypercellular solid groups focally associated with a collagenic stroma and a vascular component with rounded or angulated ectasis channels. Pseudoangiomatous cystic slit-like stroma, two mitosis in ten high power fields (HPF) appeared but without necrosis.

Immunohistochemistry (Table 1) showed that tumor cells were reactive for CD34 (Figure 3) and STAT6 (Figure 4); positive with variable intensity for estrogen receptor and diffusely positive for progesterone receptors. The neoplasm has juxtaposed morphological characteristics in different CD34 reagent fibroblastic histogenesis entities. However, the strong and extensive fusion of STAT6 is conclusive for differential diagnosis, indicating a benign SFT since the sample did not meet malignancy criteria. The expression of hormone receptors is unspecified in this context.

The Institutional Review Board of "*Hospital Universitário Oswaldo Cruz/Pronto -Socorro Cardiológico Universitário de Pernambuco – Professor Luiz Tavares*

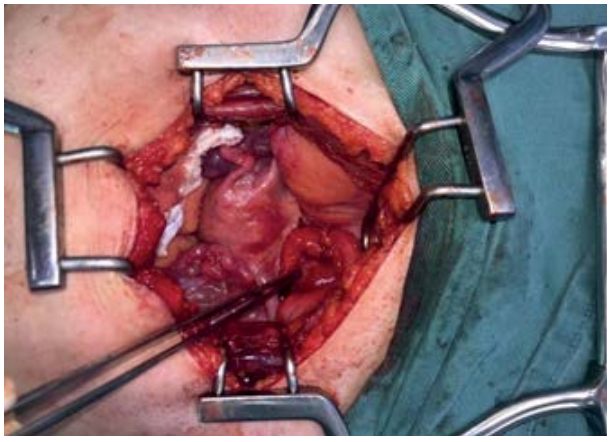

Figure 2. Paravesical tumor

Table 1. Immunohistochemistry – Markers versus expression

Marker	Expression
AE1/AE3	Negative*
SMA (smooth muscle actin), 1A4	Negative (positive internal control)
Calretinin	Negative*
CD34	Diffusely positive (Figure 3)
Desmin	Negative (positive internal control)
Inhibin alpha	Negative*
Protein S-100	Negative (positive internal control)
Estrogen receptor	Positive with variable intensity
Progesterone receptor	Diffusely positive
STAT6	Diffusely positive (Figure 4)

(*) No positive internal control or later validation of the result was found.

(Procape)" approved the study in compliance with Resolution 466/2012[10], Operational Norm 001/13 of the National Health Council of the Ministry of Health[11] report number 5,198,756 (CAAE (submission for ethical review) 54115721.9.0000.5192). The patient signed the Informed Consent Form.

DISCUSSION

SFT is a rare neoplasm initially described by Klemperer and Robin[8] in 1931. Although primarily described in the pleural cavity, it has also been described in the head and neck, extremities, mediastinum, pelvic and abdominal cavity[1-3]. They usually occur in men and women alike between 20 and 70 years, peaking in the fifth and sixth decades of life[3,4].

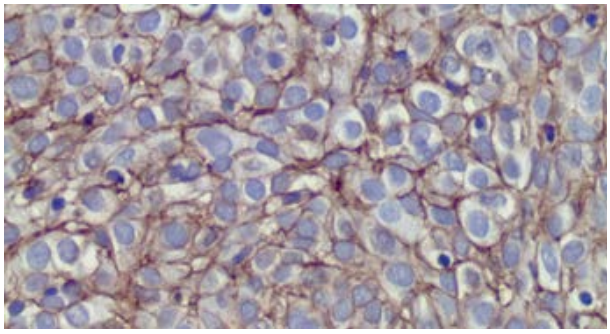

Figure 3. Immunohistochemistry for CD34
Note: Tumor cells were diffusely positive for CD34.

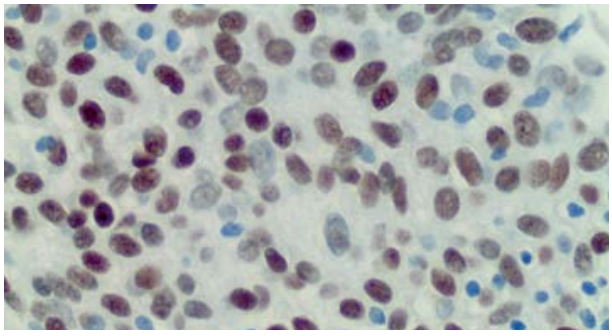

Figure 4. Immunohistochemistry for STAT6
Note: Tumor cells were diffusely positive for STAT6.

Most SFT are benign but 10% to 15% of the cases are malignant and aggressive. It is a fibroblastic or myofibroblastic origin neoplasm of the soft tissues with intermediate biologic behavior, rarely metastatic, justifying adequate surgical resection and good histopathological and immunohistochemical analysis[1].

NAB2 and STAT6 fusion proteins is a marker for this disease, in addition to the association with tumor onset. The treatment involves surgical resection when possible or administration of tyrosine kinase inhibitors as PDGFR and VEGFR[12].

Magro et al.[13] found 87 cases – 63 benign (72.4%) and 24 (27.6%) malignant – of pelvic SFT in a literature review. Aggressive tumors can relapse locally or metastasize many years after the initial treatment. The main metastatic sites are lungs, liver and bones. Large tumors > 10 cm and compromised surgical margins are dismal prognostic factors[1].

The present case reports an even more rare pelvic SFT. The symptoms, when present, are usually secondary to the compression of the bladder and/or rectum. The patient had none and was investigated only after a routine ultrasound revealed paravesical mass. As demonstrated by Chick et al.[14], the radiologic findings of CT are unspecific, frequently revealing node masses with well-circumscribed margins.

Although the preoperative diagnosis of these tumors is anticipated because it would differentiate them from stromal tumors, fine needle punctures are arguable for several tumors due to the small tissue specimen[15,16].

For resectable tumors, surgical resection with negative margins is the management of choice[17]. The patient underwent laparoscopy through transverse suprapubic incision with complete resection of the lesion with negative margins.

SFT are frequently hemorrhagic, mostly the larger (5 to 10 cm), which can make surgical approach technically difficult[18,19]. Yokoyama et al.[20] reported a case of giant SFT at the pelvis treated successfully after pre-operative embolization of the feeding arteries of the lesion. Although a vascularized tumor has been detected in the patient with local bleeding to touch, hemostasis was controlled, and pre-operative embolization was not required. She evolved without complications and was discharged the second post-operation day.

Most SFT are indolent and do not relapse or metastasize. However, relapses of 10% to 25% of pleural tumors have been reported in 10-year follow up[1]. The reasons for more aggressive tumors of some SFT are unknown and the prognostic value of biomarkers NAB2 and STAT6 is being investigated to help clinical practice but no clear prognostic value has been determined so far[21]. Due to its rarity and absence of randomized clinical trials, no global consensus exist about the adjuvant treatment with radiotherapy and/or chemotherapy for these tumors[9].

The patient is currently asymptomatic after nine months of follow-up, a short period in comparison with the literature which reports the likelihood of later tumor relapse[1].

CONCLUSION

SFT at the pelvic region due to its rarity may be an incidental finding of routine ultrasound and symptomatology is related to local tumor effects. Immunohistochemistry was the most specific recommended method of diagnosis, mainly when positivity for CD34 and STAT6 is found. Surgical treatment with extended resection and negative margins was successful and should be the major objective of treatment and histopathological diagnosis of patients with resectable tumors. Preoperative embolization of tumor feeding arteries has been described but unnecessary for the current case. The patient evolved without complications, discharged at the second postoperative day and 9-month relapse-free follow up. However, continuous follow up is required due to reports of 10-years relapse post-surgical resection.

CONTRIBUTIONS

Both authors contributed substantially to the study design, acquisition, analysis and interpretation of the data, wording, critical review and approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

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None.

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