Vulva Plexiform Fibrohistiocytic Tumor: Case Report

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Tumor Fibro-histiocítico Plexiforme de Vulva: Relato de Caso

Tumor Fibrohistiocítico Plexiforme de la Vulva: Reporte de Caso

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ABSTRACT

Introduction: Plexiform fibrohistiocytic tumor is a rare clinical condition, with about 150 cases described in the literature. **Case report:** 23-years-old female patient with nodular lesion in vulva without malignancy characteristics on ultrasound image. She underwent clinical follow-up and after an increase in the size of the lesion, surgical resection was opted. Histopathological findings suggested plexiform fibrohistiocytic tumor, with free surgical margins. In follow up, she was referred to oncological evaluation for screening complementary exams and rule out morphologically similar diagnoses. After the complementary exams confirming the initial diagnosis of plexiform fibrohistiocytic tumor and without evidence of metastatic disease after complete surgical excision with free margins, the patient is in clinical follow-up. **Conclusion:** Reporting the case is extremely important to disseminate the clinical presentation, diagnostic steps, proposed treatment and share information about the clinical evolution presented.

Key words: Histiocytic Sarcoma; Vulvar Neoplasms; Case Reports.

RESUMO

Introdução: O tumor fibro-histiocítico plexiforme é uma condição clínica rara, com cerca de 150 casos descritos na literatura. **Relato do caso:** Paciente, 23 anos, sexo feminino, portadora de lesão nodular em região vulvar sem características de malignidade em exames de ultrassonografia. Realizou acompanhamento clínico, tendo apresentado aumento do tamanho da lesão inicial, sendo optado então por realização de ressecção cirúrgica da lesão. Em estudo histopatológico, evidenciou-se lesão compatível com tumor fibro-histiocítico plexiforme, com margens cirúrgicas livres. No seguimento, a paciente foi encaminhada para avaliação oncológica com a intenção de realizar exames complementares de rastreio e descartar diagnósticos morfologicamente semelhantes. Diante dos exames complementares confirmando o diagnóstico inicial e sem evidência de doença metastática após excisão cirúrgica completa com margens livres, foi iniciado seguimento clínico. **Conclusão:** Relatar o caso é de extrema importância para divulgar a apresentação clínica, etapas diagnósticas, tratamento proposto e compartilhar informações acerca de evolução clínica apresentada.

Palavras-chave: Sarcoma Histiocítico; Neoplasias Vulvares; Relatos de Casos.

RESUMEN

Introducción: El tumor fibrohistiocítico plexiforme es una condición clínica poco frecuente, con unos 150 casos descritos en la literatura. **Reporte del caso:** Paciente de 23, sexo feminino, años con lesión nodular en una región vulvar sin características malignas en los exámenes de ultrasonido. Se sometió a un seguimiento clínico, habiendo presentado un aumento en el tamaño de la lesión inicial, y se decidió realizar la resección quirúrgica de la lesión. En un estudio histopatológico, se evidenció una lesión compatible con tumor fibrohistiocítico plexiforme con márgenes quirúrgicos libres. Fue referida para la evaluación oncológica, realizó pruebas complementarias para el cribado y para descartar diagnósticos morfológicamente similares. En vista de las pruebas complementarias que confirman el diagnóstico inicial y sin evidencia de enfermedad metastásica después de una escisión quirúrgica completa con márgenes libres, se inició el seguimiento clínico. **Conclusión:** Informar del caso es extremadamente importante para difundir la presentación clínica, etapas diagnósticas, tratamiento propuesto y compartir información sobre la evolución clínica presentada.

Palabras clave: Sarcoma Histiocítico; Neoplasias de la Vulva; Informes de Casos.

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INTRODUCTION

Described for the first time by Enzinger and Zhang in 1988, based in the analysis of 65 cases, the plexiform fibrohistiocytic tumor (PFT) is a mesenchymal neoplasm with approximately 150 cases described hitherto[2]. It is a clinical entity of rare presentation affecting children and young adults with mean age of approximately 14.5 years at the onset and more prevalence in females. So far, no evidences of prevalence for a specific ethnicity exist[1,3].

It is a slow-growing, low degree tumor, usually asymptomatic, commonly located in the upper extremities as shoulders, forearm, fingers, hands, head and neck. The recurrence varies from 12.5% to 40% with lymph nodes and pulmonary metastasis[1,3,4]. Although affects predominantly the subcutaneous adipose tissue, this malignant tumor can affect the bone[5].

After clinical follow-up, most of the patients fare well, the physicians should prescribe long-term follow-up to help them[6].

Because of its rarity, the objective is to report the clinical presentation, diagnostic phases, treatment proposed, in addition to information about the clinical evolution of a case of PFT in order to share with the scientific community and help healthcare professionals in differential diagnosis and clinical conduction of this pathology.

CASE REPORT

Female, 23 years-old, nulligravida, without previous comorbidities, non-smoker and social drinker. Two years ago, during the initial evaluation, noticed nodular lesion in the vulva with 0.5 cm of diameter and painless. Sought medical care and underwent ultrasound of the vulvar region, showing heterogeneous and well demarcated hypoechoic nodular image with nearly 10 mm x 3 mm x 3 mm. The ultrasound was suggestive of fibrosis node. Based in the finding, it was decided for outpatient follow-up of the lesion. At the time, lab tests without alterations, denied weight loss, bleedings, changes of menstrual cycle or other symptoms associated. Morbid and family history indicated deceased maternal grandmother due to face neoplasm, but unable to report specific etiology.

After the initial exam, the lesion evolved with fast growing, painless and when palpation revealed nearly 0.5 cm in the greatest diameter, the patient sought medical consultation once again when new US of the vulvar region revealed hypoechoic, heterogeneous node in the subcutaneous tissue of larger dimensions in comparison with the previous, measuring nearly 3.0 x 0.6 x 1.1 cm, but without malignant characteristics and absence of inguinal lymphonodomegalia.

She was submitted to surgical resection of the lesion upon gynecology outpatient differential diagnosis of the etiology. Hepatologic evaluation showed lesion consistent with surgical free-margins PFT.

Referred for oncologic evaluation, immunohistochemistry study of the lesion was performed (Figure1), confirming the initial diagnosis of PFT, showing myxoid or hyalin areas with discreet leukocyte infiltrate, immunoexpression for CD68 and vimentin with absence of expression for factor XIIIa, CD34 and protein S-100. Computed tomography (CT) of the chest without significant alterations and of abdomen and pelvis with small area of cutaneous/subcutaneous densification in the vulvar region that may possibly correspond to the surgical excision.

Based in the complementary exams confirming the initial diagnosis and without evidence of metastatic disease after complete margin-free surgical excision, clinical follow-up commenced. After two months of the diagnosis, patient asymptomatic, good healing of the surgical site, absence of new signs and symptoms or laboratory alteration.

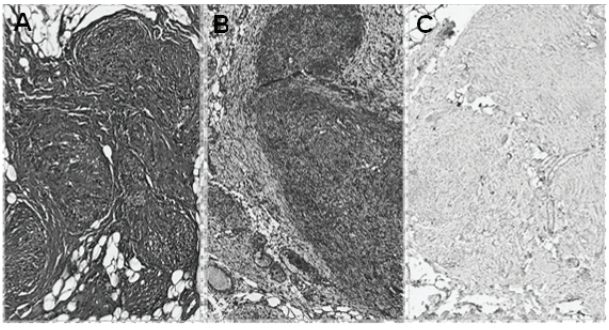

Figure 1. Immunohistochemistry: plexiform fibrohistiocytic tumor: myxoid or hyalin areas with discreet leukocyte infiltrate **(A)**. immunoexpression for CD68 and vimentin **(B)**. absence of expression for factor XIIIa, CD34 and protein S-100 **(C)**

DISCUSSION

The mechanism of onset of PFT is still unknown with cases described related to local trauma and the possibility of congenital etiology[3,7].

The tumor has a slow growth pattern from months to years, typically as a soft tissue node, rarely larger than 3 cm in its greatest diameter involving the subcutaneous adipose tissue and may extend to the dermis, skeletal muscle or both. Can affect any body regions, but there is description of substantial impact in shoulders, forearm, fingers, hands, head and neck[1,4]. In this case, the tumor was localized in the vulvar region.

The diagnosis is confirmed by histological test and immunohistochemistry profile. Histologically, PFT is

characterized by multiple small nodules arranged in a plexiform pattern. Three distinct cell types are noted in variable amounts: spindle fibroblast-like cells, mononuclear histiocyte-like cells and osteoclast-like giant cells[1,3,4].

The immunohistochemical test is useful to support the diagnosis and rule out similar morphologic diagnoses[7]. PFT reacts with CD68 antibodies, staining mainly the multinucleated gigantic and histiocytic cells, and the fibroblastic cells display reactivity to vimentin, in addition to focal reactivity for muscle actin[3,4], not showing expression of cytokeratins, protein S-100, CD34 and factor XIIIa[2,8]. In this study, immunoexpression for CD68 and vimentin with absence of expression for factor XIIIa, CD34 and protein S-100 were observed, ensuring the diagnosis of PFT.

The histogenesis of PFT is quite complex. Previously, it was defined as a variation of the malignant fibrous histiocytoma and currently the World Health Organization (WHO) classifies it as an intermediary malignancy of the so-called fibrohistiocytic tumors rarely metastasizing[9]. Regardless of being included in the subgroup of fibrohistiocytic tumors, it is believed that the cellular origin of the tumor are myofibroblasts with potential of transformation in fibroblasts or histiocytic-like cells. Ultrastructural studies and immunohistochemistry indicate (myo) fibroblastic and histiocytic cellular properties[5].

PFT can appear in x-rays as a discreet mass with soft parts edema, without calcifications or subjacent bone alterations. Ultrasound can show a discreet hypoechoic mass[10], consistent with the case of this study, with a hypoechoic node lesion measuring approximately 10 mm x 3 mm x 3 mm detected in the initial exam.

A few other pathologies should be included in the differential diagnoses, the main are plexiform neurofibroma, Schwannoma plexiform, cellular neurothekeoma, fibrous hamartoma in childhood, deep benign fibrous histiocytoma, dermatofibroma and myofibromatosis[1-3].

Even with low malignant potential, the treatment of choice for PFT is free-margins[10] complete surgical resection as extensive as possible, this conduct was followed for the patient in question, the margins of the lesion removed were disease-free. The rate of local recurrence is considered high between 12.5% and 40%. There were reports of some patients with local relapse with pulmonary metastasis and regional lymph nodes[1,3].

At the diagnosis and later long-term follow-up, the patients should be submitted to proper exams of chest imaging to rule out pulmonary metastases which, though rare, have been described in younger women[7]. Therefore, it was decided to follow the specialized literature with tomography exams to screen secondary lesions, ruled out after the assessment. The follow-up must include yet a thorough evaluation of the primary excision site because of high relapse rates[3,7].

Povýsil and Habanec[11] reported a six-years follow-up period without recurrence. Yalcinkaya et al.[5] described a 27 months follow-up after initial resection, the patient feeling well and without recurrence.

The follow-up of the patient of this study after surgical resection is still short, if compared with other publications that showed the importance of a longer follow-up which is still more necessary, considering the younger age.

The Institutional Review Board of "*Fundação Centro de Controle de Oncologia do Amazonas*" (FCecon), CAAE: 18447619.4.0000.0004 approved the study. The patients signed the Informed Consent Form (ICF) authorizing the publication.

CONCLUSION

Because of its rarity, the case was described to contribute for the scientific knowledge, in addition to disclosing the indolent clinical condition, diagnostic phases, established treatment and strict later follow up and collaborate for the conduction of new studies. For the patients, follow-up is essential for the purpose of early diagnosis either locally, relatively high or nodal or remote metastasis, a little rarer, but possibly occurring as younger are the patients.

CONTRIBUTIONS

Aluisio Juliano Baumgartner and Fernanda Stefania Bastos Garcia contributed for the study design and planning, collection, analysis and interpretation of the data, wording, critical review with intellectual contribution. William Hiromi Fuzita, Lia Mizobe Ono and Caroline Souza dos Anjos contributed for the wording and critical review with intellectual contribution. Érica Condé Marques e Oliveira contributed for the study design and/or planning, collection, analysis and interpretation of the data. All the authors approved the final version to be published.

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DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

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