

Low-grade fibromyxoid sarcoma as a content of perineal hernia. A case report

Sarcoma fibromixioide de bajo grado como contenido de hernia perineal. Reporte de un caso

Danna Auria-Aspiazu^{1*}, Omar Parrales-Calderón² and Fernando Moncayo-Asnalema³

¹General Surgery Department, Instituto Oncológico Nacional Dr. Juan Tanca Marengo, ION SOLCA; ²Coloproctology Department, Hospital de Especialidades Dr. Abel Gilbert Pontón; ³General Surgery Department, Hospital de Especialidades Dr. Abel Gilbert Pontón. Guayaquil, Ecuador

Abstract

Background: Low-grade fibromyxoid sarcoma is a rare neoplasm with apparently benign histology but with late aggressive metastatic behavior. The perineal hernias are unusual, there are no more than 100 cases reported in the world literature.

Discussion: The presentation of this fibroblastic neoplasm is rare and makes it even more rare if it is as the content of a primary perineal hernia. **Conclusion:** It is an infrequent neoplasm that occurs in approximately 0.7% of all cases, with aggressive metastatic behavior and in need of adjuvant treatment.

Keywords: Perineal hernia. Fibromyxoid sarcoma. Low-grade fibromyxoid sarcoma. Soft tissues.

Resumen

Antecedentes: El sarcoma fibromixioide de bajo grado es una neoplasia rara de histología someramente benigna, pero con un comportamiento agresivo metastásico tardío. Las hernias perineales son de presentación inusitada y no existen más de 100 casos reportados en la literatura mundial. **Discusión:** La presentación de esta neoplasia fibroblástica es poco frecuente, y aún más como contenido de una hernia perineal primaria. **Conclusión:** Es una neoplasia infrecuente que se presenta en aproximadamente el 0.7% de los casos, con comportamiento agresivo metastásico y con necesidad de tratamiento adyuvante.

Palabras clave: Hernia perineal. Sarcoma fibromixioide. Sarcoma fibromixioide de bajo grado. Tejidos blandos.

Introduction

Fibromyxoid sarcoma is a rare neoplasm that accounts for approximately 0.7% of soft tissue tumors. First described by Evans in 1987, it is a tumor of fibroblastic origin with apparently benign clinical and

histological characteristics, but it exhibits a highly aggressive local and metastatic behavior. Its most common location is the proximal lower extremities, especially the thighs, and then the chest wall of young adults of between 25 and 46 years of age, with an average age of 35 years, and with higher predominance in males. No more than 150 cases have been

*Correspondence:

Danna Auria-Aspiazu
E-mail: dania_221@hotmail.com

Date of reception: 01-02-2020

Date of acceptance: 04-03-2020

DOI: 10.24875/CIRUE.M20000244

Cir Cir (Eng). 2021;89(2):243-246

Contents available at PubMed

www.cirugiaycirujanos.com

2444-0507/© 2020 Academia Mexicana de Cirugía. Published by Permanyer. This is an open access article under the terms of the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

reported worldwide given that, so far, it is a barely known entity^{1,2}.

Due to the low incidence of this type of tumors and its unusual form of presentation as hernial content, this case is described in order to review the international literature with regard both to its epidemiology and histopathological characteristics, and its clinical presentation and hitherto available therapeutic options.

The purpose of this article is to document a case of low-grade fibromyxoid sarcoma as the content of a primary posterior perineal hernia in a young woman. The incidence, clinical and histopathological characteristics, unusual presentation, and the therapeutic management of this type of neoplasm are described, given its highly infrequent nature according to a verification of available literature.

Clinical case

Thirty-one-year-old woman with a history of bronchial asthma and obstetric-gynecological history of four pregnancies (two abortions and two cesarean sections), who attended the emergency room for a 5-month history, after a gluteal trauma due to a fall from her own height, of left vulvar region edema, with reddening, heat, and pain since a few weeks. Abdominopelvic ultrasound reported an enlarged Bartholini gland, presence of a 1.72 x 0.59-cm fistula descending from the lip to the left buttock, and a 3.39 x 3.16-cm heterogeneous encysted image of irregular borders with a 10-cm³ volume and heterogeneous anechoic internal image on left gluteus. The patient was managed on an outpatient basis with analgesics and antibiotic therapy (ibuprofen, 400 mg/8 h PO, and azithromycin, 500 mg/12 h for 3 days), and follow-up by outpatient services.

Three days later, the patient attended the outpatient clinic with a bloody purulent collection in the left lateral wall of the vagina, left perineum, and gluteal region, which produced outflow and decompression of the cystic-looking mass on vaginal examination. Pelvic ultrasound showed a hypoechogenic image measuring 15 x 15 x 16 cm, with an approximate volume of 1,200 mL, in relation to an apparently retroperitoneal collection that extended to the left and pushed the uterus to the right, which measured 69 x 33 mm; fine echogenic endometrium without collections within and non-evaluable ovaries.

Comprehensive management was decided together with the general surgery department, which indicated

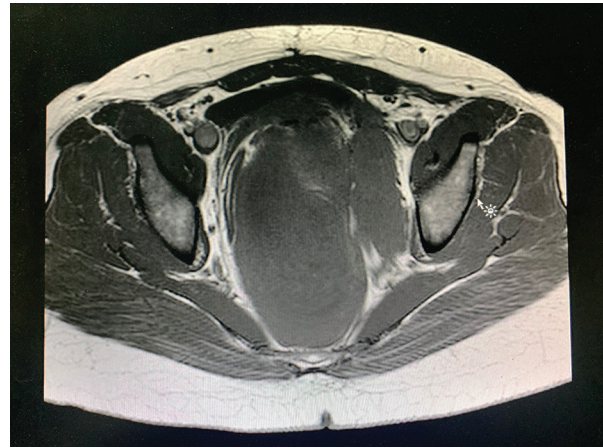


Figure 1. Abdominopelvic magnetic resonance imaging axial section showing an expansive tumor lesion of solid intensity, lobulated borders, hypointense on T1-weighted sequences.

the performance of an abdominopelvic magnetic resonance imaging in which an expansive tumor lesion was identified in the pelvic region, which extended in cephalad direction at L4 supraumbilical level, which compressed and pushed the uterine body to the left, entered through the lesser pelvis and pushed the sigmoid rectum, and slid through the left ischiopubic space into the perineal region and left paramedian gluteal region, which measured 34 cm, along a 13-cm anteroposterior axis of larger extension at the gluteal level and a 10-cm transverse axis in the pelvic cavity, occupying the pre-sacral and prevertebral space, compressing and pushing the bladder (Figs. 1 and 2). Carrying out laboratory tests and consultations with the anesthesiology and cardiology departments was decided in order for an abdominopelvic examination to be performed (Fig. 3).

Exploratory laparotomy revealed a large mass of gelatinous cystic appearance that pushed the uterus, bladder and ureter to the right, and that surrounded the left ureter at its distal portion, with pelvic floor muscular structures weakness. Obtaining a cleavage plane between the hernial sac that protruded towards the posterior perineal region was not possible; however, the mass was dissected and separated from the pelvic structures for its excision. With approximate bleeding of 900 mL and trans-surgical transfusion of two packed RBC units, the abdominal cavity was closed after placing a Hemovac-type drain by counter-opening directed to the bottom of the sac. The tumor was sent to the pathology department for examination.

During the postoperative period, the patient received antibiotic therapy, with mild transient hematuria



Figure 2. Abdominopelvic magnetic resonance imaging sagittal section showing a heterogeneous hyperintense lesion on T2-weighted sequences with paramagnetic contrast heterogeneous uptake, located at the pelvic region.



Figure 3. Hernial tumor lesion located on the perineal region and left paramedian gluteal region.

that subsided in 24 hours, and approximately 30-45-mL serous-hematic fluid daily output. She was discharged 5 days after the intervention.

Histopathological gross description reported fragments of tissue measuring 30 x 22 x 9 cm, of elastic consistency, which together weighed 1,300 g, irregular in shape and surface, grayish-white in color with violaceous areas. When sectioned, an elastic, heterogeneous consistency, yellowish-brown color and myxoid appearance were observed, which was consistent with low-grade fibromyxoid sarcoma (Fig. 4). Immunohistochemical examination reported positivity for vimentin and desmin, and negativity for S100, smooth muscle 1A4 actin, striated muscle AC, CD117, CD99, CD34, epithelial membrane antigen, and broad-spectrum keratin, which confirmed the diagnosis.

The patient is receiving adjuvant treatment with chemotherapy and radiotherapy.

Discussion

Soft tissue tumors constitute a heterogeneous group of tumors, both due to their histological presentation and for the diversity of their immunohistochemistry. Usually, they originate from multipotent primitive mesenchymal cells that can generate several cell lines

while undergoing neoplastic transformation. Low-grade fibromyxoid sarcoma accounts for about 0.7% of soft tissue tumors. First described by Evans in 1987, it is of fibroblastic origin and has apparently benign clinical and histological characteristics, but it has a highly aggressive local and metastatic behavior, with a high rate of local recurrence of approximately 33%, and 58% for metastasis. Its incidence is uncertain^{1,2}. So far, there are 26 case series in English language literature, and in 15 of them, molecular techniques have been used for their diagnosis³.

It shows predominance for males and young adults aged between 25 and 46 years. It is most often located in the lower extremities, mainly the thigh, although it can also appear on the chest wall, axilla, shoulder, gluteus, inguinal region, or the neck. A few cases have been described in the retroperitoneum, small bowel mesentery, pelvis, mediastinum, and falciform ligament. Most are sub-fascially located; it rarely has subcutaneous or dermal localization^{2,4,5}.

It presents as a slow-growing, almost painless soft tissue tumor. Sometimes it can occur with tumor hemorrhage, and it is uncommon for it to cause

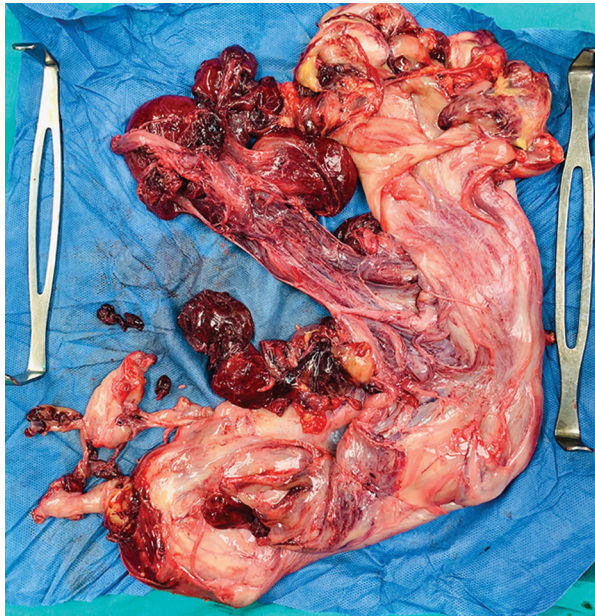


Figure 4. 30 x 22 x 9-cm tumor lesion, of irregular surface, elastic consistency, yellowish brown color and myxoid appearance.

neurological or vascular symptoms due to compression or stretching of nerves or adjacent bursae, depending on anatomical location^{4,6}.

Differential diagnosis is broad and includes myxoid variants of neurofibroma, leiomyoma, hemangiopericytoma, desmoid fibromatosis, malignant peripheral nerve tumor, spindle cell liposarcoma and angiomyxoma^{1,7}.

Histological examination usually shows fibroblasts with myxoid areas, with a swirling distribution pattern, with minimal nuclear pleomorphism, pale eosinophilic cytoplasm and slight or no mitotic activity. As for immunohistochemistry, most of them show positivity for vimentin and negativity for keratin, desmin, actin, S-100 protein, CD34, CD31 and membrane epithelial antigen^{5,6}.

Reverse transcriptase polymerase chain reaction (RT-PCR) molecular analysis and fluorescent in-situ hybridization (FISH) detection of chimeric transcription of the FUS-CREB3L2 fusion gene and FUS gene rearrangement confirm and support morphological and immunohistochemical results⁸.

The treatment of choice is surgery, although resection is usually incomplete, given that these are circumscribed but not encapsulated tumors. Chemotherapy and radiation therapy have not been shown to modify the course of the disease or to improve survival. Disseminated metastases can be managed with palliative treatment. There are no specific guidelines for metastatic and

recurrent disease, since the approach depends on patient status and histopathological results^{1,4}.

Conclusion

Low-grade fibromyxoid sarcoma is a rare tumor and can mimic other soft tissue neoplasms. High rates of relapse and metastasis are described. Final diagnosis requires histopathological examination, with immunohistochemistry and clinical correlation.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained informed consent from the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

Funding

The authors declare that no funding was obtained from any organization or institution for the performance of this article.

Conflicts of interest

The authors declare that there are no conflicts of interest.

References

1. Tinoco-Tellez L, Aguirre Trigueros J, Dominguez-Gasca L, Dominguez-Carrillo L. Sarcoma fibromixioide de bajo grado. Acta Médica. Grupo Ángeles. 2016;14:230-4.
2. Cruz-Benítez L, Cortés-Cárdenas S, Fariás-Alarcón M, Tenorio-Torres J, Ramírez-Heredia J, Conde-Vazquez E. Sarcoma fibromixioide de región sacra. Reporte de caso. GAMO. 2011;10:319-25.
3. Mengtian L, Huijiao C, Dengchao S, Min C, Zhang Z, Hongying Z. Low-grade fibromyxoid sarcoma: a clinicopathologic and molecular study of 10 genetically confirmed cases. Int J Clin Exp Pathol. 2018;11:5860-8.
4. Solano Iturri G, Blanco Sampascual S, Orive Cura V. Tumor fibromixioide de bajo grado. Gastroenterol Hepatol. 2011;34:56-7.
5. Jain S, Xu R, Prieto VG, Lee P. Molecular classification of soft tissue sarcomas and its clinical applications. Int J Clin Exp Pathol. 2010;3:416-28.
6. Scoggins CR, Pisters PW. Diagnosis and management of soft tissue sarcomas. Adv Surg. 2008;42:219-28.
7. Alfaro-Cervelló C, Benavent Casanova O, Nieto G, Mares Diago FJ, Navarro S. Sarcoma fibromixioide de bajo grado, un diagnóstico diferencial esencial en los tumores mixoides de apariencia benigna. Rev Esp Patol. 2018;51:178-82.
8. Sambri A, Righi A, Tuzzato G, Donati D, Bianchi G. Low-grade fibromyxoid sarcoma of the extremities: a clinicopathologic study of 24 cases and review of the literature. Pol J Pathol. 2018;69:219-25.