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Vulvar Aggressive Angiomyxoma- A Rare Diagnose in Post Menopause

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Abstract
Aggressive angiomyxomas are rare, locally invasive mesenchymal tumours, usually occurring in the pelvis or perineum of young women. They can be locally aggressive and have high risk of local relapse. The authors present a case of a postmenopausal woman with a five-year, slow growing, large, pedunculated mass on the right labia majora. Histological exam after surgical excision concluded it was an aggressive angiomyxoma. There was no local relapse after two years of follow-up.
Keywords: *Aggressive angiomyxoma, vulvar, genital, menopause.*

Introduction

Aggressive angiomyxoma is a rare locally aggressive and infiltrative soft tissue tumor, which usually occurs in the pelvic-perineal region (main site is the perineum, followed by the pelvic cavity and vagina) in reproductive age [1-3]. It is a slow growing neoplasm with a significantly higher incidence in females (female-to-male ratio of 6:1)[4]. They have a prominent myxoid matrix with extensive vascularization, frequently with estrogen and progesterone receptors and have high risk for local recurrence [1,4-6]. Since the major part of the neoplasm is often concealed within the deep soft tissue (only twenty-five percent are pedunculated) and do not cause rectal, urethral, vaginal or vascular obstruction, most tumors are large at the time of surgery. Surgical resection, followed by long-term surveillance is recommended. Treatment of recurrence is advised but no treatment has demonstrated to be clearly better than others. Repeated surgeries are associated with increased morbidity [7,8].

Case

The authors report a case of a 61-years-old woman, with two gestations with vaginal birth, menopause at 53 years old, which did not use replacement hormone therapy. The patient attended our Gynaecology department because of a large, painless neoplasm that had been growing for five years on her right labia majora. For family reasons, the patient had not had a gynaecological exam since that time. Genital exam revealed a round pedunculated tumoral mass, coloration

similar to the patient's skin, with 15-20 cm of extension and long base (Figure 1). The rest of the exam was normal. Cervical cytology was negative for intraepithelial lesion or malignancy. The situation was discussed with the patient and a complete surgical resection was performed in the operation room without major incidents. There was profuse bleeding during the procedure, which was controlled with suture and pressure. Histopathology revealed spindle-shaped cells with stellate cells in a myxoid background, many capillary-sized vessels and perivascular collagen condensation (Figure 2). Immunohistochemical staining was positive for vimentin, desmin, estrogen and progesterone (Figure 3). Complete histological exam concluded it was an aggressive angiomyxoma. She had no local relapse of the disease at follow-up two years after surgery.

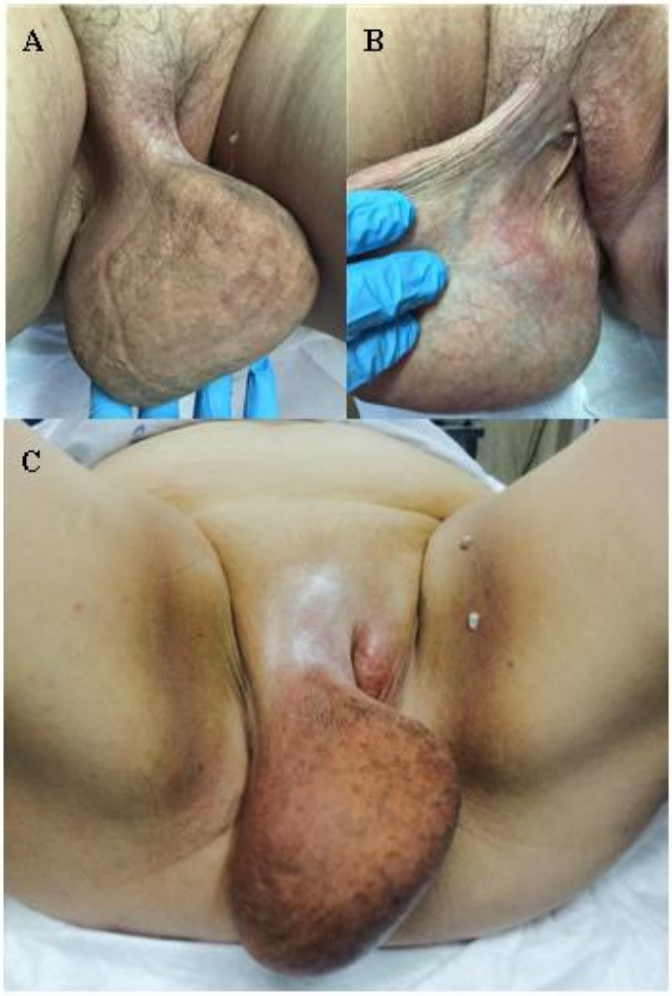

Figure 1 - Vulvar neoplasm observed during gynaecological exam

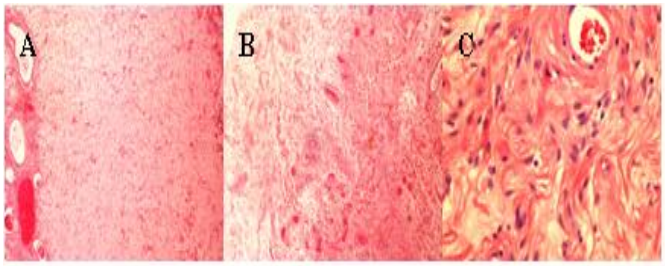

Figure 2 - Histopathology revealed spindle-shaped cells with stellate cells in a myxoid background, many capillary-sized vessels and perivascular collagen condensation (A,B: H&E 4x; C: H&E 20x).

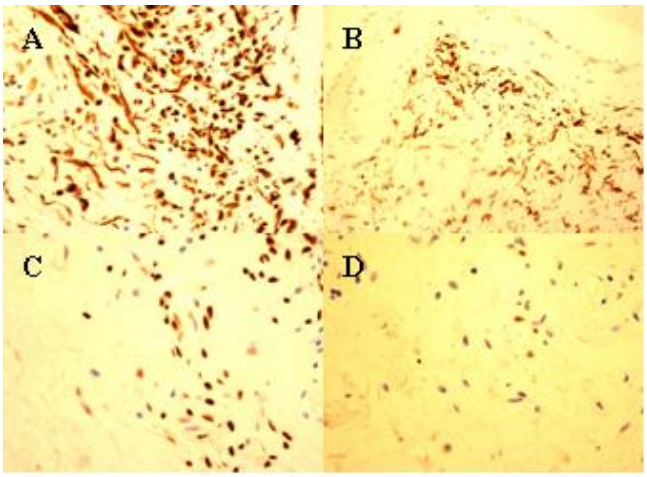

Figure 3 - Immunohistochemical staining was positive for vimentin (A, 20x), desmin (B, 20x), estrogen (C, 20x) and progesterone (D, 20x)

Discussion

Aggressive angiomyxoma occurs mostly in the pelvic and perineal region of women in reproductive age [1-3]. They present as a soft slow growing mass with no capsule. It can invade the paravaginal or pararectal spaces or extend retroperitoneally. Differential diagnosis with other mesenchymal tumors can be difficult and consists on the microscopic identification of stellate cells and a myxoid matrix with positive immunohisto-chemical staining for desmin, vimentin and smooth muscle actin [9]. These neoplasms are rare in menopause. Treatment consists of surgical resection of the lesion and long term follow up. Relapse of the disease is frequent in up to 50-70% of the cases [10].

The case we present is uncommon in nowadays-developed countries with easy access to medical

consultations. These neoplasms are rare and when pedunculated like this one, are uncomfortable and frequently detected and treated at an early stage, not reaching such bulky sizes.

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References

1. Han-Geurts IJ, van Geel AN, van Doorn L, M den Bakker, Eggermont AM, Verhoef C. Aggressive angiomyxoma: Multimodality treatments can avoid mutilating surgery. Eur J Surg Oncol. 2006;32:1217–21.
2. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. N Engl J Med. 1999;341:1772.
3. Blandamura S, Cruz J, Faure Vergara L, Machado Puerto I, Ninfo V. Aggressive angiomyxoma: A second case of metastasis with patient's death. Hum Pathol. 2003;34:1072–4.
4. Lin HC, Liu CC, Kang WY, Ke HL, Li CC, Wu WJ, Huang CH. Huge aggressive angiomyxoma: a case report and literature review. Kaohsiung J Med Sci. 2006;22:301–304.
5. Fishman A, Otey LP, Poindexter AN, Shannon RL, Gritanner RE, Kaplan Al. Aggressive angiomyxoma of the pelvis and perineum. A case report. J Reprod Med. 1995;40:665–669.
6. Wolf CA, Kurzeja R, Fietze E, Buscher U. Aggressive angiomyxoma of the female perineum in pregnancy. Acta Obstet Gynecol Scand. 2003;82:484–485.
7. Chen H, Zhao H, Xie Y, Jin M. Clinicopathological features and differential diagnosis of aggressive angiomyxoma of the female pelvis - 5 case reports and literature review. Medicine Clinical Case Report. 2017;96:20.
8. Das BP, Baruah D, Medhi KB, Talukder B. An aggressive angiomyxoma of vulva - A rare entity - A case report. J Midlife Health. 2016;7(3):140–143.
9. McCluggage WG, Patterson A, Maxwell P. Aggressive angiomyxoma of pelvic parts exhibits estrogen and progesterone receptor positivity. J Clin Pathol. 2000;53:603–605.
10. Bagga R, Keepanasseril A, Suri V, Nijhawan R. Aggressive Angiomyxoma of the Vulva in Pregnancy: A Case Report and Review of Management Options. MedGenMed. 2007;9(1): 16.