CASE REPORT

Aggressive Angiomyxoma of Vulva Which Grew with Pregnancy and Attained a Huge Size Rarely Seen in Literature

Sinha Vandana · Dave Kalpana S. ·
Bhansali Ronak P. · Arora Ruchi S.

Received: 1 September 2012 / Accepted: 1 December 2012 / Published online: 26 March 2013
© Federation of Obstetric & Gynecological Societies of India 2013

Introduction

Aggressive Angiomyxoma (AA) of the pelvis and perineum was identified as a distinct clinicopathologic entity in 1983 [1] with less than 250 cases reported since then. The tumor usually arises in women of the reproductive age group with peak incidence in the fourth decade. Few cases of its occurrence outside the pelvis have been reported.

Case Report

A 43-year-old female came to our OPD with complains of a growth over her private parts for the past 10 years and difficulty in walking for the past few days. She had delivered by Cesarean Section 9 months ago. The tumor increased gradually for 9 years, but grew suddenly during and after her pregnancy.

On examination, there was a huge mass arising from the left labia majora of about 55 × 45 × 45 cm with a broad

pedicle (Fig. 1). The skin over the tumor was smooth, intact, and without any obvious nodularity. The right labia majora had multiple warts of 2–12 mm in size, also involving the peri-anal region.

Pelvic examination was normal. There were no palpable lymph nodes. Biopsies from the large mass diagnosed angiomyxoma and biopsies from the warts diagnosed them as fibroepithelial polyps.

Her routine investigations, Pap Smear were normal. CT scan showed the presence of 53 × 43 × 46 cm sized heterogeneously enhancing soft tissue density lesion involving the left labia and perineum.

A wide excision of the tumor with adequate tumor-free margins and primary closure was done. There was no significant blood loss during the surgery. Histopathologic examination of the excised mass confirmed angiomyxoma, which was further confirmed by immunohistochemistry. The tumor was an infiltrating, diffuse, very low cellular spindle cell tumor with extensive myxoid areas and thick and thin blood vessels with foci of lymphocytes around the blood vessels. The base of the resection was free of tumor. She is symptom free for the past 8 months.

Discussion

Aggressive Angiomyxomas are benign tumors. The term “aggressive” refers to the tumor’s locally infiltrative and recurrent nature, but with rare distant metastases [2].

Sinha V. (✉)
c/o Dr. Dhiraj Saxena, Department of Hyperbaric Medicine,
Apollo Hospitals, Bhat, Plot-1A, GIDC Estate, Gandhinagar,
Gujarat 382428, India
e-mail: drvandanasinha@rediffmail.com

Sinha V., Fellow · Dave K. S., Professor and Head of
Department · Bhansali R. P., Assistant Professor ·
Arora R. S., Junior Lecturer
Department of Gynecology, Gujarat Cancer & Research Institute,
Civil Hospital Campus, Asarwa, Ahmedabad 380016, Gujarat, India

Fig. 1 Angiomyxoma of vulva arising from left labia majora with multiple warts on right labia majora

In our case, AA showed the typical insidious growth pattern. Patients can present with an asymptomatic perineal nodule or a pelvic mass diagnosed on imaging studies. Our case also reflects the possible hormone dependence of this tumor as its growth was pregnancy related. Estrogen receptor positivity was also seen in the immunohistochemistry.

Pre-operative diagnosis helps in planning the extent of excision and patient counseling. However, the rarity of these neoplasms and a lack of typical features make it a difficult pre-operative diagnosis. They are misdiagnosed as

labial, Bartholin's, or Gartner's Cyst and are diagnosed histologically after surgical excision.

Magnetic Resonance Imaging accurately detects the trans-levator spread and relation of the mass with the anal sphincter, urethra, bladder, and pelvic side-wall.

Tumor recurrences are high and can be seen even within 6 months from initial resection. There is no proven therapy for recurrences. Radiotherapy, selective angiographic embolism, hormone antagonists like tamoxifen, or GnRH analogs are also treatment options, but their role is still unclear.

Conclusion

Angiomyxomas are rare tumors. Complete surgical excision is necessary to prevent recurrence. Patient counseling, individualization of each case, and long-term follow-up are essential.

References

1. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of 9 cases of a distinctive type of gynecologic soft tissue neoplasm. *Am J Surg Pathol.* 1983;7: 463–75.
2. Sereda D, Sauthier P, Hadjeres R, et al. Aggressive angiomyxoma of the vulva: a case report and review of the literature. *J Low Genit Tract Dis.* 2009;13(1):46–50.