CASE REPORT

Aggressive angiomyxoma of the vulva

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ABSTRACT

Aggressive angiomyxoma is a rare, benign neoplasm occurring in 3rd to 5th decade of life that can be mistaken both clinically and on microscopy for several other conditions, it should be included as a differential diagnosis for any vaginal mass. These lesions have a predilection for female pelvic soft tissues, slow in growth, and are characterized histologically by a predominantly myxoid stroma and an abundance of thin and thick walled vascular channels. This is a deep soft tissue tumor, which as the name suggests, may have a locally aggressive course. Most tumors occur in women and are large, usually greater than 10 cm, slowly growing, and painless. Standard of care treatment for angiomyxoma has been surgery. Some authors believe that it is the only possible treatment, but surgery is often radical and can be mutilating, with massive blood loss.

Key Words: Aggressive angiomyxoma, myxoma, myxoid neoplasm, female perineum

INTRODUCTION

Aggressive angiomyxoma was first reported as a distinct variant of myxoid neoplasms in the female vagina and pelvis by Steeper and Rosai.[1] These tumors mostly occurs in 3rd to 5th decade of life and 95% in females. These lesions have a predilection for female pelvic soft parts, slow growth, frequent recurrences, and are characterized histologically by a predominantly myxoid stroma and an abundance of thin and thick walled vascular channels. Many authors have subsequently reported this lesion in female and male patients. This is a deep soft tissue tumor, which as the name suggests, may have a locally aggressive course. Most tumors occur in women and are large, usually greater than 10 cm, slowly growing, and painless. The most common location is in the perineal region and may exert pressure on adjacent organs.

CASE REPORT

A 35 year old woman, Hosp. No. – A 8332 came to our OPD on 05.08.05 with chief complaints of growth in the vulva - 2 month, bleeding from the growth - 1 month, irregular period - 6 month. She is P4+1. She chews betel nut and leaf. There is no family history of malignancy. No history of HTN, DM, TB. Tubectomy was done a year ago.

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On examination, GC is fair, pallor +, no palpable LN. P/A- no organomegaly, others NAD, groin- NAD. Local examination of the vulva revealed a 8 × 5 cms pedunculated growth [Figure 1] having its thick pedicle of about 1.5 cm thickness hanging from the clitoreal region slightly on the left side [Figure 2]. P/S/V- cervix- NAD, vaginal walls- NAD, Uterus – N/S, mobile, adnexae free. P/R- NAD.

On investigation, Hb %- 9.8 gm%, ABO gr.- O +ve, RBS- 101 mg/dl, RFT-WNL, LFT-WNL. PAP Smear-NAD, CXR-NAD, USG (W/A) – NAD. Pre operative biopsy revealed Aggressive Angiomyxoma.

She was planned for wide excision of the vulval growth; accordingly it was done under SA on 18.08.05 [Figure 3]. On sectioning, the mass had focally infiltrating margins and a rubbery glistening grey/white surface [Figure 4]. Post operative period was uneventful, and patient was discharged on 27.08.05.

Post op HPE revealed Aggressive Angiomyxoma [Figures 5 and 6], cut margins negative. IHC studies revealed Desmin

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and Vimentin +ve, ER, PR +ve and S100 protein negative. She is under regular follow-up, and doing well for the last 5 years.

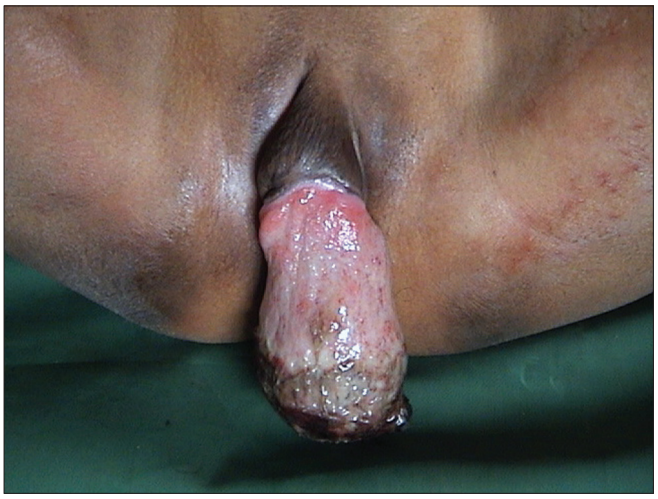

Figure 1: Clinical presentation

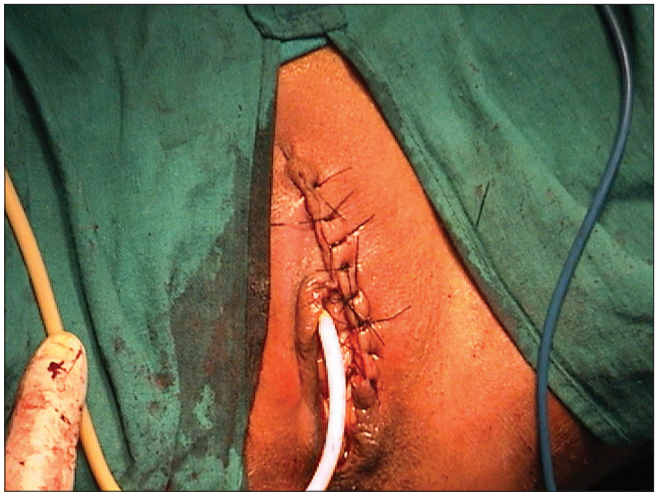

Figure 3: After the repair

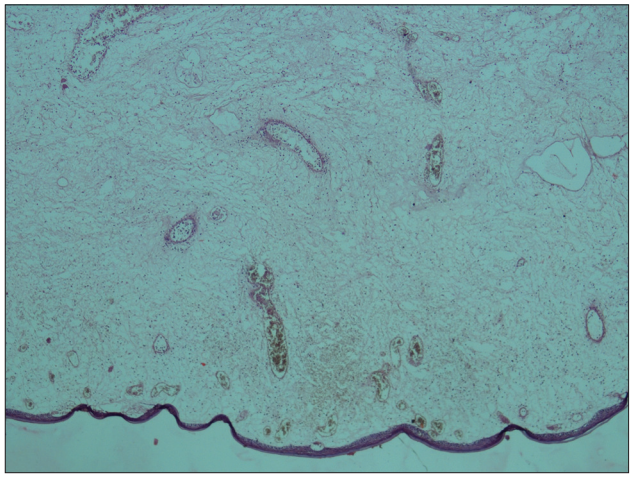

Figure 5: HPE shows thin epidermis and underlying hypo cellular myxoid tissue containing blood vessels of various calibers

DISCUSSION

Aggressive angiomyxoma is a rare, benign neoplasm that can be mistaken both clinically and on microscopy for several other conditions, it should be included

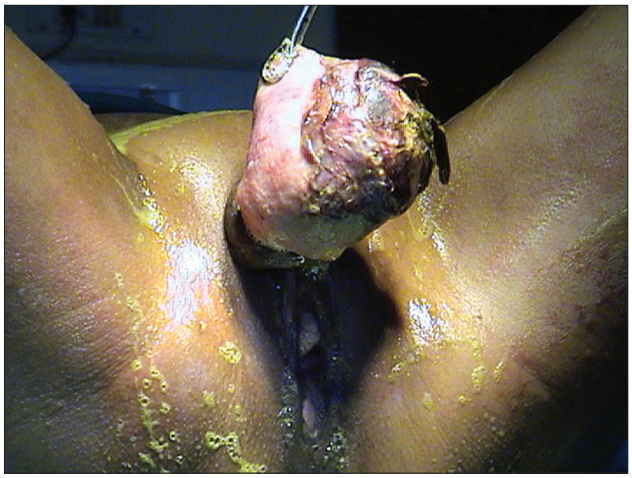

Figure 2: Growth coming out from the clitoreal region

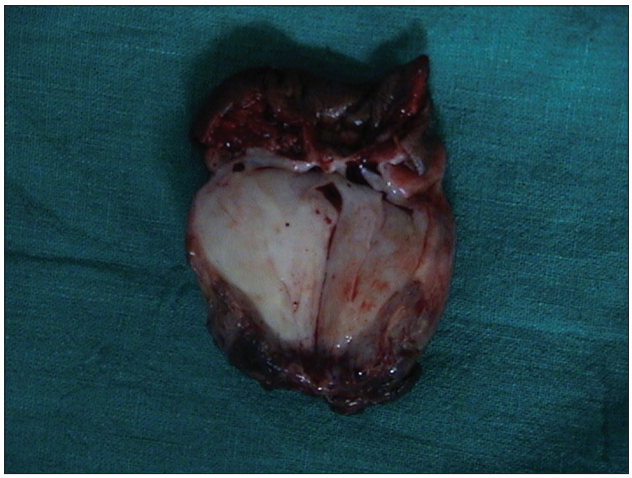

Figure 4: Specimen cut open

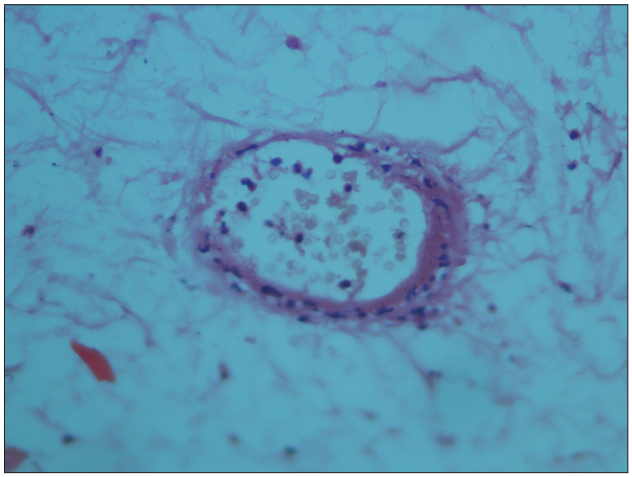

Figure 6: High power view showing hyaline thickening of blood vessels

as a differential diagnosis for any vaginal mass.[2] It is important to diagnose this condition because the tumour is locally infiltrative and requires wide excision and follow up. It occurs mainly in the female pelvis, vulva, or perineum, though some cases are described in men. Standard of care treatment for angiomyxoma has been surgery. Some authors believe that it is the only possible treatment, but surgery is often radical and can be mutilating, with massive blood loss. It also requires postoperative intensive care monitoring, and the recurrence rate after surgery remains high even if negative margins are obtained at the initial resection. A retrospective review has shown that patients having positive margins were as likely to have recurrence as those with negative margins.[3]

Immunohistochemistry of the tumor cells revealed diffuse immunoreactivity for estrogen receptors, progesterone receptors, vimentin, and CD34. No expression of S-100 protein, Bcl-2 protein, CD117 (c-kit gene product), epithelial membrane antigen, desmin, or h-caldesmon could be demonstrated.

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