Kathmandu University Medical Journal (2005), Vol. 3, No. 4, Issue 12, 423-424

Case Note

Pedunculated angiomyxoma of the vulva - A rare clinical entity

Dash S[1], Awasthi RT[2], Bandana K [3]
[1] Lecturer, [2] Professor and HOD, [3] Medical Officer, Department of OBG, Manipal Teaching Hospital, Pokhara, Nepal.

Abstract
Background: Aggressive angiomyxoma is an uncommon mesenchymal tumor that preferentially involves the pelvic and perineal regions of females. Since its initial description in 1983, approximately 65 cases have been reported in the English literature.
Case: A 33-year-old parous woman with a slow growing pedunculated soft tissue tumour of the vulva which later underwent surface ulceration. Pathological findings in the case were suggestive of an aggressive angiomyxoma, a distinctive soft tissue non-malignant tumour, which could, at times be locally aggressive.

Keywords: Aggressive Angiomyxoma, Pedunculated Vulval Tumours

Clinical presentation
Mrs. S.N, a 33 year old P2L2, presented to us with complaints of a mass arising in the genital region interfering with her locomotion. She also complained of ulceration and bleeding from the mass. The patient first noticed the mass after the delivery of her last child, five years ago. The mass initially, to start with, was the size of a pea-nut with a thin stalk. It gradually increased in size and length over the past five years, with a history of surface ulceration 6 months ago due to local trauma. The ulcer on the mass was non-healing, painless and it gradually increased in size to its present size. The patient was widowed one year back, her husband succumbing to AIDS. Her previous two deliveries were normal deliveries and there was no practice of any form of contraception. There is no other significant past, personal or family history.

Examination findings
The patient was of average build and nourishment. General examination revealed no significant abnormality other than the presence of pallor. Breasts were normal and there was no swelling in the region of the thyroid gland. Per abdomen examination was normal. On examination of the genitalia, there was a 7x7 cm pedunculated mass arising from the anterior half of the right labium majus. The pedicle of the mass measured approximately five cm (picture-1). The most dependant portion of the mass showed a 4x4 cm ulcer (picture-2), with regular margins and an indurated base. The floor of the ulcer was covered with unhealthy pale granulation tissue. The whole mass was soft and spongy in consistency, non-tender, with no palpable expansile cough impulse.

Fig. 1: Pedunculated soft-tissue tumour arising from the right labium majus

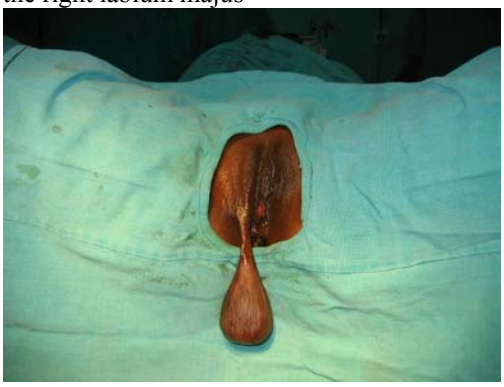

Fig 2: Excised specimen showing surface ulceration in the dependent area of the tumour.

Dr. Soumyaroop Dash
Lecturer, Department of Obstetrics & Gynaecology,
Manipal Teaching Hospital, Pokhara
Email: sdash2k1@yahoo.co.in

On the basis of the above findings, the patient was planned for local excision of the mass under short GA Pre-operative investigations revealed a Hb of 9.8 and a normal bleeding and clotting time. She was sero-negative for HIV by the dot ELISA test. The patient underwent an un-complicated simple local excision of the tumour with ligation of the stalk. The specimen was labeled and sent for histo-pathological examination. The histo-pathological report showed a well differentiated tumour of mesenchymal origin, consistent with the diagnosis of CUTANEOUS VULVAL ANGIOMYXOMA.

Discussion

Aggressive angiomyxoma (AAM) is a distinctive neoplasm seen in the female genital tract. Since its initial description in 1983, approximately 65 cases have been reported in English literature. The term 'Aggressive' has been added to this mesenchymal tumour to denote its propensity for local aggression, reticent recurrences and lack of metastatic potential. The soft tissues of the pelvis, perineum, vulva, buttocks, retro-peritoneum and the inguinal region are usually involved [1,2,3]. This tumour is more common in females, between 16 and 70 years (median ;34 years), and about one-fourth of these tumours are pedunculated [3].

The line of differentiation of this tumour is not firmly established, but a fibroblastic / myo-fibroblastic origin has been proposed [4]. The histo-morphology of this tumour can be easily confused with angio-myofibroblastoma and leiomyoma of the vulva with myxomatous degeneration [4,5].

The definitive treatment of AAM in the perineum, especially pedunculated tumours, is only a local excision. Surgery, whenever offered for symptom control, should be done with minimal morbidity. Non-pedunculated tumours and peri-rectal tumours usually require wide local excision [2,6].

The tumour usually recurs in 30 to 40 % of patients undergoing local excision, within a period of 10 months to 7 years [2]. A possible estrogen-progesterone dependency of this tumour (by the virtue of positive estrogen / progesterone receptors) may cause the tumour to grow or recur during pregnancy [7,8]. With repeated recurrences, a potential medical treatment with GnRH analogues may obviate the need for repeated surgeries for a recurrent aggressive angio-myxoma of the vulva [9].

References

1. Sanghi PS, Desai S, Pradhan SA, Chinoy RF : Aggressive angio-myxoma of the female perineum : A case report. Indian J Pathol Microbiol 40 (3) : 385-7, 1997
2. Amezcua CA, Begley SJ, Mata N et al : Aggressive angiomyxoma of the female genital tract : a clinico-pathologic and immuno-histochemical study of 12 cases. Int J Gynecol Cancer 15(1) ; 140-5, 2005
3. Fetsch JF, Laskin WB, Lefkowitz M et al : Aggressive angiomyxoma : a clinico-pathological study of 29 female patients. Cancer 78(1) : 79-90, 1996
4. Granter SR, Nucci MR, Fletcher CD: Aggressive angiomyxoma : reappraisal of its relationship to angiomyofibroblastoma in a series of 16 cases. Histopathology 30(1) ; 3-10, 1997
5. Nemoto T, Shinoda M, Komatsuzaki K et al : Myxoid leiomyoma of the vulva mimicking aggressive angiomyxoma. Pathol Int. 44(6); 454-9, 1994
6. Behranwala KA, Thomas J: Aggressive' angiomyxoma : a distinct clinical entity. Eur J Surg Oncol 29(7); 559-63, 2003
7. Havel G, Burian P, Kortz M, Mark J: Aggressive angiomyxoma of the vulva: an unusual, deceptive and recurrence-prone tumour with evidence of estrogen receptor expression. Case report. APIMS 102(3); 236-40, 1994
8. Htwe M, Deppisch LM, Saint-Julien JS: Hormone dependent, aggressive angiomyxoma of the vulva. Obstet Gynecol 86(4 Pt 2); 697-9,1995
9. Fine BA, Munoz AK, Litz CE et al: Primary medical management of recurrent angiomyxoma of the vulva with a gonadotropin-releasing hormone agonist. Gynecol Oncol 81(1); 120-2, 2001