A rare vulval lump: a case of superficial angiomyxoma

Widanage N¹, Abeygunasekera N², Wasalaarachchi KCK³, Yapa C⁴

Key words

Vulva, lump, superficial, angiomyxoma

Introduction

Patients with vulval lumps are frequently encountered in sexually transmitted diseases (STD) clinics. A wide variety of conditions including venereal as well as non-venereal causes like condylomata lata, genital warts, Bartholin's abscesses, nodular scabies, sebaceous cysts, seborrhoeic keratosis, skin tags and malignancies can present to STD clinics as vulval lumps. It is essential for the clinicians who are managing patients in STD clinics to have a good understanding of the possible differential diagnoses of vulval lumps.

Here, we describe a rare case of superficial angiomyxoma of the vulva presenting to STD clinic, Kalubowila. Less than 20 cases of superficial angiomyxoma of the vulva have been reported world wide as of now.(1, 2)

Case

A 42 year old married lady was referred from the gynaecology ward with two progressively enlarging painless lumps in the vulva of 2 years duration. During the last two months there had been a rapid increase in the size of lumps. There was no vaginal discharge, abdominal pain or dyspareunia. However, she did report of having similar lesions in the same site thirteen years ago which had been surgically removed. Her menstrual cycles were regular with no inter-menstrual bleeding or post-coital bleeding. She has had two normal vaginal deliveries and there was no history of abortions. She denied ever having

premarital or extra marital sexual exposures and the marital partner was asymptomatic.

Examination revealed two large soft, non-tender pedunculated masses arising from the left labia majora measuring 16cm x10cm and 12cmx 8cm. In addition to these two large lumps there were several much smaller lumps (figure 1). There were a few ulcers and hypopigmented areas on the surface of the larger lesions. STI screening was negative and cervical Papanicolaou smear did not show any abnormality.

Figure 1 : Large pedunculated vulval lumps

The lumps were surgically excised. The biopsy report stated an oedematous stroma with hypocellular fibrous tissue and myxoid appearances with scattered thin walled blood vessels. There was no cellular atypia or mitoses seen. Foci of adipose tissue were also seen predominantly in the pedicle sections of the excision margin. The conclusion was a benign fibrovascular lesion suggestive of superficial angiomyxoma (figure 2).

¹Registrar in Venereology, ²Consultant Venereologist, STD clinic, ³Consultant Pathologist, ⁴Consultant Obstetrician & Gynaecologist, Teaching Hospital, Colombo South, Kalubowila.

Figure 2 : Microscopic appearance; Oedematous stroma with hypocellular fibrous tissue and thin walled blood vessels.

Discussion

Three types of angiomyxomas have been identified; superficial angiomyxoma (SAM), aggressive angiomyxoma (AAM) and angiomyofibroblastoma (AMFB). SAMs are rare benign cutaneous tumours arising from the dermis or subcutis. It was first described in 1988.(3) SAM is a distinctive clinicopathologic entity that should be included in the differential diagnosis of myxoid cutaneous tumours. (3)

A study done on 17 cases of SAM arising in the genital region found that the predominant reason for seeking medical attention was a slow growing painless mass. Also this study reports the sites of involvement in females were the labium majus (n=6), vulva (n=4), groin (n=2) and mons pubis (n=1) of the 13 females in the study.(4) Most cases occur in adults as an asymptomatic solitary papule or nodule with equal sex incidence.(5) However, literature review revealed a case of SAM in a three year old girl in Japan.(6)

SAMs are usually less than 3cm and have a wide anatomical distribution with a predilection for the trunk, head/neck area and genital skin.(5) The patient we encountered had two large lesions measuring 16cm x10cm and 12cm x 8cm. Giant superficial angiomyxomas measuring over 10cm have been reported and they can mimic aggressive

angiomyxoma.(2) As this vulval lump was fairly large the possibility of whether it was an AAM was queried. AAM and SAM can be differentiated histologically and with immuno-histochemical studies. AAMs are often larger, ill-defined measuring 10cm or more with extension into deeper tissue and composed of thick walled blood vessels.(5) SAMs are not locally destructive and composed of numerous small blood vessels with sparse spindle shaped cells in a prominent myxoid stroma.(5) Actin and desmin stains are positive in almost all cases of AAMs whereas SAMs are desmin negative.(7) Tumour cells of AAMs are often positive for oestrogen and progesterone receptors.(5) Immuno-histochemical studies were not performed in this case. Local recurrence is common and is seen in up to 30% of cases when there is incomplete excision of SAMs.(5) This patient too, had similar lesions in the past.

A case of SAM complicated with *condylomata accuminatum* and *staphylococcus hominis* has been described.(8) It highlighted the importance of early treatment of SAM as they are composed of small blood vessels in a myxoid stroma where surface ulceration can lead to various secondary infections. Even though there were small ulcers noticed over the lumps in this patient there was no evidence of secondary infection. In patients with multiple lesions the possibility of Carneys' complex comprising of myxomas, lentiginous pigmentation and endocrine abnormalities should be considered.(9) However, in our patient there was no clinical evidence of endocrine abnormalities or skin pigmentations.

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