Vulval angiomyofibroblastoma

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Abstract

A 25-year old woman presented to the Gynaecology Clinic at Base Hospital, Diyatalawa with a history of a lump in the right labium majus for 3 month duration. The lump was surgically excised. The histopathological diagnosis was an angiomyofibroblastoma (AMFB) a rare benign mesenchymal tumour arising in the genital region predominantly of the pre-menopausal women.

Key words: angiomyofibroblastoma, benign vulval tumours.

Introduction

Angiomyofibroblastoma is a rare benign mesenchymal tumour which occurs in the genital region, especially the superficial areas of vulva, of pre-menopausal women1,2. It was first described by Fletcher et al in 19923. AMFB can rarely occur in the spermatic cord and the scrotum in men4. Since 1992 approximately 150 cases have been documented in the literature5. The tumour has an extremely low risk of recurrence if completely excised6.

Case report

A 25-year-old woman presented to the Gynaecology Clinic at Base Hospital, Diyatalawa complaining of a lump at vulva for 3 months. It was gradually enlarging but was not painful and was not ulcerated. She has had a vaginal birth two months back.

On examination, there was a lump in the right labium majus. It was 6 cm in diameter with an irregular surface, firm, non-tender, non-fluctuant and was not attached to the underlying structures. The overlying skin was normal and there were no abnormal blood vessels. The regional lymph nodes were not enlarged. There was no cough impulse.

The lump was surgically excised. It resembled a fibroid on macroscopic appearance. The pathological diagnosis was an angiomyofibroblastoma.

Discussion

Angiomyofibroblastoma originates from an immature multipotent cell of the connective tissue situated around the vessels, which is capable of differentiating into similar tumoral varieties7. Clinically AMFB presents as slow-growing, painless lumps with a low tendency for local recurrence, and usually are misdiagnosed as Bartholin’s gland cysts8,9. They are mostly well-circumscribed masses with a rubbery texture, measuring 0.5-12 cm in diameter7,9.

AMFB is histologically characterized by alternating hypercellular and hypocellular areas with an abundance of venular or capillary-sized blood vessels. Tumour cells are spindled, plump spindled or plasmacytoid stromal cells that cluster around blood vessels within an oedematous to collagenous matrix, sometimes forming solid compact foci. Nuclei are bland. Mitotic figures are absent or very sparse2,8.

Immunohistochemical staining is diagnostic in AMFB10. Immunohistochemically the stromal cells are reactive for vimentin and desmin, but not cytokeratin, muscle-specific actin, alpha-smooth muscles actin or S-100 protein. Both oestrogen and progesterone receptors are diffusely expressed in the stromal cells, suggesting the sex-steroid dependency of the tumour2,8,9.

The tumours may rarely be associated with a malignant counterpart (angiomyofibrosarcoma)10.

The differential diagnosis includes
1. Aggressive angiomyxoma
2. Cellular angiofibroma
3. Endothelial stromal sarcoma8.

Other uncommon sites of occurrence such as the vaginal vault and fallopian tubes have rarely been reported4,11,12,13.

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