Phyllodes Tumour of the Vulva: Report of a Case and Review of the Literature

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Citation

Abstract
Phyllodes tumour occurring in the vulva is rare with only a few cases reported in the literature. We report a case of a 33-year-old lady who presented with a small vulval swelling in which subsequent excision of the lesion showed a benign phyllodes tumour. She was followed-up by re-excision which did not reveal any residual lesion and she is well without recurrence 6 months after surgery.

INTRODUCTION
The mammary ridges appear during the fourth week of intrauterine life and extend bilaterally from the axilla to the groin and medial thigh. Supernumerary nipples and breasts can, although rarely, appear from extra mammary buds that develop along these lines. Mammary-like tissue of the vulva can present as a small nodular swelling and a variety of benign breast diseases such as fibroadenoma may arise in this ectopic tissue. Phyllodes tumours are relatively uncommon in the breast with an overall incidence of approximately 1.5% of that of breast carcinoma. However, phyllodes tumour arising in mammary like tissue of the vulva is rare.

CASE REPORT
We report a case of a 33-year-old lady who presented to the Gynaecology Outpatient Clinic of the University Hospital Aintree with a 2cm mobile labial swelling that had been present for 18 months. The patient complained that this was gradually getting larger but was not painful. There were no other abnormalities on both general and pelvic examination. The lesion was subsequently removed under general anaesthesia. Postoperatively, she made an uneventful recovery and was discharged home. The patient is well with no evidence of recurrence 6 months after surgery.

Macroscopic examination of the resected lump revealed an opened light brown cystic lesion measuring 10x8x7mm, all of which was processed for microscopic examination. Histologically, the neoplasm had a biphasic pattern and was composed of a leaf-like configuration of tissue covered by epithelium and projecting into cystic spaces. The stroma was composed of bland spindle-shaped cells embedded in a loose myxoid stroma (figure 1). There was no evidence of pleomorphism or necrosis and mitotic figures did not exceed 2 mitoses/10 high power fields. Normal breast ducts were seen elsewhere in the specimen. On immunohistochemistry, the stromal cells were positive for CD34, vimentin and bcl-2. They were negative for p53 and Ki67. The epithelial cells were positive for bcl-2 and MNF-116 and were weakly positive for p53 and Ki67 (<1% of cells). Oestrogen and progesterone receptors were strongly positive in 40% and 85% of epithelial cells respectively. The epithelial cells also showed moderate positivity with c-Kit (CD117). The tumour was reported as a benign phyllodes tumour.
DISCUSSION

Phyllodes tumour of the vulva is rare with only a few case reports published in the literature. The microscopic features together with the immunohistochemical profile of this neoplasm in this unusual site are similar to its counterpart in the breast. Review of previous case reports revealed that the average age at presentation ranged between 20 and 39 years and most of the patients presented with a painless cystic vulval swelling or nodule. Normal breast ducts were seen adjacent to the lesion in the majority of the cases. Only one case recurred after 8 months and this was attributed to its large size (4 cm). Although most phyllodes tumours in the breast are benign, histologically benign and borderline cases may tend to recur if they are not completely excised. In our case no comment could be made on completeness of excision because the cyst was received open. However, re-excision did not reveal any residual tumour.

In these tumours the epithelial cells are usually positive with epithelial markers (AE1/AE3, CAM 5.2, MNF-116), bcl-2, oestrogen and progesterone receptors. The stromal cells usually express bcl-2, vimentin, actin, desmin and CD34, and they lack p53 and c-Kit (CD117) expression in benign tumours. Due to the rarity of cases of phyllodes tumour in the vulva, prognosis is uncertain. However, a favourable outcome is expected in our case.

ACKNOWLEDGEMENT

We wish to thank Dr W. Taylor, Consultant Histopathologist, for his help in reviewing the manuscript.

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