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Case Report

Vulval fibroadenoma, a common tumor at an uncommon site - A case report with review of literature

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ABSTRACT

Vulval fibroadenoma is a rare benign tumour arising from anogenital mammary like glands. Vulva is an uncommon site for fibroadenoma. This is a case of 33 year old female presented with well defined mass in the right labia majora with a histopathological diagnosis of Vulval fibroadenoma confirming origin from ectopic breast tissue.

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1. Introduction

Vulval fibroadenoma is a mammary like fibroepithelial lesion. It is extremely rare having low incidence. Milk lines are precursors to the mammary glands which extends from the axilla to groin and undergoes regression during embryogenesis.¹ If they fail to regress, accessory breast structures arise from these remnants and if they persist in vulva, many physiologic and pathologic change can happen which can lead to both benign as well as malignant neoplasms.² The potential morphological variations include columnar cell, apocrine change, lactational change, pseudo angiomatous stromal hyperplasia (PASH), fibroadenoma, phyllodes tumour and multinucleated stromal giant cells.^{3,4} Vulvar Fibroadenoma occurs predominantly in women of reproductive age, but may be seen in post menopausal women and rarely in prepubertal girls. It presents as solitary cutaneous or subcutaneous nodule on labia majora and enlarges gradually.^{5,6} The clinical differential diagnosis include benign lesions like epidermal cyst, Bartholin's cyst, lipoma, and malignant lesions include extramammary Paget's disease and adenocarcinoma.⁷ The

vulvar fibroadenoma shows histologic features similar to those in the breast.

2. Case Report

This is a case of 33 year old nulliparous woman who presented with a painless swelling in right labia majora for few months, which gradually increased in size. Physical examination revealed a 2.5x2.5cm mass located in the subcutaneous area of the right labia majora. The swelling was firm, mobile, there was no ulceration, no puncta.

USG of the abdominopelvic region was unremarkable. Gross examination of the excision specimen showed a 2.5x2.5cm mass under skin with greyish white and lobulated cut surface. There was no cystic change, haemorrhage or necrosis.

Microscopy showed a biphasic tumour with epithelial and stromal proliferation in a peri canalicular and intracanalicular pattern. Epithelial element consisted of tubular branching glands lined by columnar epithelial and myoepithelial cells. Stroma was moderately cellular and fibro myxoid in nature. There was no evidence of atypia. Immunohistochemistry was done. The epithelial cells were positive for Ck7, myoepithelial cells were positive for

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p63. The glandular cells showed diffuse strong positivity for Progesterone Receptors (PR) and focal positivity for Estrogen receptors (ER). The glandular elements were positive for GATA3 and negative for PAX-8.

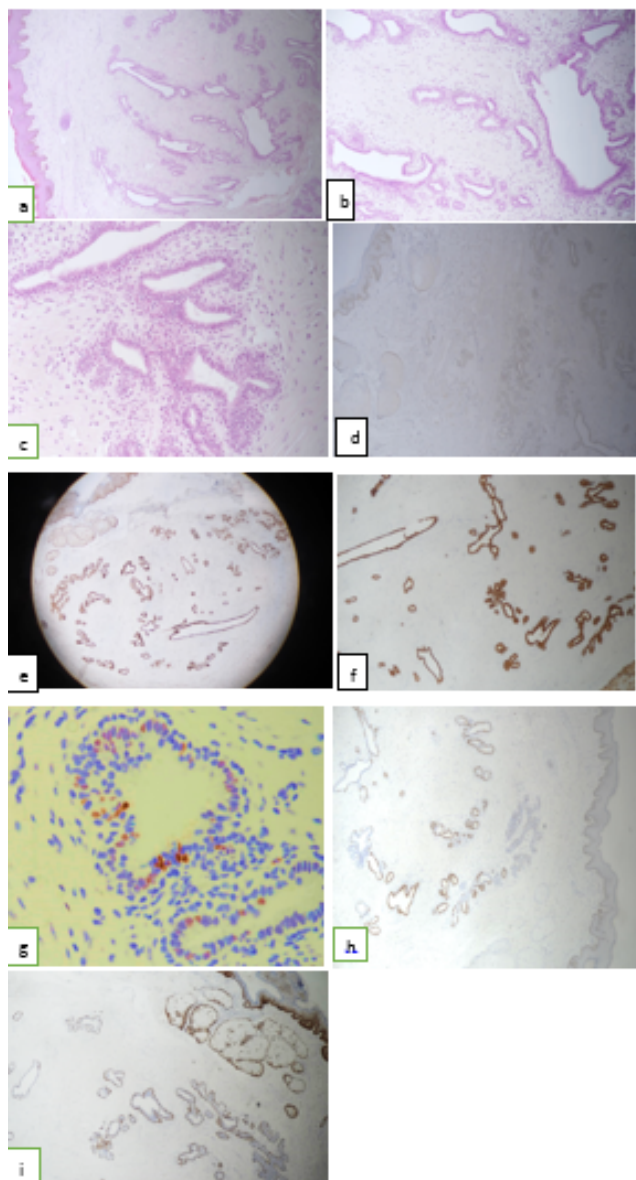


Fig. 1: a: (10x), b: (20x); c:(40x): benign dilated ducts lined by ductal and Myoepithelial cells; d: PAX8; e: GATA3; f: CK7; g: & h: ER & PR; j: p63

3. Discussion

The vulvar mammary like glands was first described by Hartung in 1872.^{8,9} Two theories have been put forth for the histogenesis, the first one states that vulvar fibroadenoma arises from ectopic mammary tissue which is derived from primitive embryological milk line. The second theory was put forth by Vander Putte in 1994¹⁰ and he suggested that

specialised glands identical to mammary glands exist in the anogenital area normally which has close relationship with eccrine glands and called them, mammary like anogenital glands. They express hormone receptors which can be detected by Immunohistochemistry. He criticized the theory of milk ridge and discussed that it cannot explain perineal ectopic breast tumours. Carter in 2008¹¹ presented an analysis of 18 reports of vulvar fibroepithelial neoplasms, showing an average patient age of 38.7 years (range, 20-60 years), average tumour size 3.0 cm (range, 0.8-6.0 cm). Ahmed¹² in his 2007 review described 10 cases, seven vulvar and three anogenital lesions with patient age ranging from 35-84 years. Vulvar fibroadenoma has an incidence of 2-6% and is difficult to differentiate it from other vulvar masses on clinical examination.¹³ Most reported cases were < 4cm diameter they are fibrous on cut section and have cystic spaces. Like the corresponding breast lesions vulvar fibroadenomas are well circumscribed and composed of glandular elements lined by bland epithelium with uniform hypocellular stroma. The glands are lined by luminal columnar cells and abluminal myoepithelial cells. In our case, histopathological features showed features of Fibroadenoma with Immunohistochemistry positive for ER, PR and GATA3.^{5,14} Vulvar Fibroadenoma has a good prognosis and rarely recurs following excision.

4. Conclusion

This case emphasizes the manifestation of ectopic mammary tissue in vulva and occurrence of a common tumour at an uncommon site. Fibroadenoma should be kept in mind as a differential diagnosis in vulvar lesions.

5. Conflict of Interest

The authors declare that there is no conflict of interest.

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