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Giant Pseudoangiomatous stromal hyperplasia (PASH) of the breast - An uncommon benign breast lesion.

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Abstract

PASH is an uncommon benign lesion of the breast , characterized by a dense, collagenous proliferation of mammary stroma, forming anastomosing slit like spaces lined by slender spindle cells. They are lined by discontinuous, often attenuated, inconspicuous cells without atypia or mitotic activity set in a hylinized collagenous stroma.

It is a rare benign condition identified during a histological examination of tissue in a twenty six year old lady who underwent lumpectomy for fibroadenoma. We have reported this case for its rarity and also for stressing upon the need to distinguish this benign lesion from low grade angiosarcoma , as the correct identification of PASH of the breast has important treatment implications for patients.

Here we describe a case of 26 yr- old female with typical clinical and surgical presentation of fibroadenoma.

Keywords: pseudoangiomatous stromal hyperplasia, fibroadenoma, angiosarcoma

Introduction

Pseudoangiomatous stromal hyperplasia (PASH) of the breast is a rare benign proliferation of mesenchymal stromal cells with irregular slit –like formation resembling vascular structures (1) . PASH may present in a wide clinicopathologic spectrum , ranging from focal minor incidental microscopic changes to clinically symptomatic breast masses. (4) But in majority of cases, this lesion is a focal microscopic finding in breast biopsies performed for benign or malignant diseases. (1) The lesion is pale, fibrous and has a homogenous cut surface and is typically well-circumscibed. Its ramifying slit like spaces lined by flattened myofibroblastic cells are opt to be mistaken for vascular spaces, leading to an erroneous diagnosis of angiosarcoma. (1) Most of the reported cases of PASH have been small to moderate sized lesions however , the exact etiology and pathogenesis of this tumor – like lesion is still unknown. It presents mainly in premenopausal women. Hormonal factors are thought to play a developmental role. This report deals with the rare occurance in

common benign breast lesions and its importance in distinguishing it from angiosarcoma. (1,2)

Case Report

A 26 yr old female was admitted with a painless lump in her right breast since 3 months. There was asymmetric enlargement of right breast. Examination revealed a large lump measuring 11cm x 8x 6.5 cm, firm to hard in consistency and mobile. It was non tender and overlying skin appeared to be normal. There was no evidence of axillary lymphadenopathy. Mammography revealed enlarged and dense mass in right breast. Cytological examination reveled inadequate cellular material.

Clinical diagnosis was giant fibroadenoma of the right breast. Patient underwent surgical excision. Gray white, homogenous firm mass was excised.

On gross examination:– excised specimen of breast-lump measured 11cm x8x 4cm, it was well circumscribed and encapsulated. Cut surface was pale tan pink to gray coloured and homogenous. Fig 1and 2

Histopathological examination revealed encapsulated tumor mass with a complex pattern of interanastomosing empty, slit like spaces. The spaces are lined by spindle cells simulating endothelial cells and surrounded by dense collagen fibres. The intervening stroma consisted of dense hylanized collagen. (fig. 1,2,3) Mitosis, tufting, atypia and pleomorphism were absent.

Discussion

Palpable breast masses are common and usually benign, but efficient evaluation and prompt diagnosis is necessary to rule out malignancy. (1)

PASH was originally reported by Vuitch et al in 1986, is nowadays a well-recognized change in the breast. The etiology and pathogenesis of mass forming PASH is not known. However, hyperplastic breast tissue from premenopausal women commonly has foci of PASH, so it is likely that the development of a discrete tumor with this pattern represents an exaggerated form of stromal hyperplasia. (2 3 4)

The pathogenesis of this lesion is attributed to hyperplasia of stromal myofibroblasts in response to hormonal stimuli. (5)

Vuitch and colleagues have suggested that mass forming PASH represents an exaggeration of normal physiological events that histologically resemble breast stromal cells in the luteal and secretory phases of the menstrual cycle. Hormonal factors are thought to play a developmental role. (1234)

According to Powell et al, foci that eventually create discrete masses probably escape normal physiological control mechanisms, cease cycling with the remaining breast and aquire the capacity for independent myofibroblastic proliferation.(1,3)

Although these lesions do not occur commonly, they represent a diagnostic challenge. Piccoli et al reported 13 cases of PASH in biopsy specimens. Developing asymmetric tissue on mammography, suggesting PASH is commonly confirmed on histopathology. (3)

PASH is associated with other benign entities, which includes proliferative and nonproliferative fibrocystic changes, fibroadenomas, gynacomastia and sclerosing lobular hyperplasia. (3)

In the past, PASH has been considered a variant of breast hamartomas, but a distinctive feature is absence of fatty tissue in PASH.(4)

Pathologically, the gross appearance of PASH is of a well-circumcribed tumor with smooth external surface resembling a capsule. The cut surface usually consists of homogenous fibrous tan or white and rubbery tissue. Although conventional PASH is clinically inapperant , rarely, when extensive ,it alone may produce a

clinically palpable tumourous mass or even massive breast enlargement. (5) The main diagnostic importance of this lesion is its potential confusion with low – grade angiosarcoma. Our patient presented at 26 years of age with large, palpable breast lump.

Histologically, the most striking finding was a complex pattern of slit-like spaces lined by plump myofibroblastic cells separated by crackled collagenous stroma. The spaces were almost always empty, only rarely containing few red blood cells. True capillaries with RBCS could be seen. Present case had similar morphological features.

Factors helpful in distinguishing PASH include, narrow slits with its peculiar lobulocentric distribution which merges with interlobar stroma rather than infiltrate and destroying it as seen in angiosarcoma which has more aggressive, infiltrative pattern. The lining cells of anastomosing spaces in PASH are lined by bland myofibroblastic cells which lack atypia, whereas in angiosarcoma , lining cells show marked atypia as well as presence of mitotic figures.Tumourous form of PASH can be confused with entities associated with increased cellularity such as cystosarcoma phylloides. But lack of stromal pleomorphism , mitoses and foci of PASH are distinguishing features

The recommended treatment for PASH is wide local excision. The recurrence rates of PASH after the excision ranges from 15 – 22%. (6)

Conclusion

This case is of interest because of its giant clinical presentation and uncommon microscopic incidental finding in breast lesion. Even though it behaves in a benign fashion, it has a propensity for growth over time or occasionally rapid growth. Excision is still the treatment of choice and is often a necessity for differentiating the condition histologically from angiosarcoma or other breast diseases. (2) The prognosis for patients with PASH is good and there are no reports of distant metastasis or death related to PASH.(7)

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Legend Figure

Fig. 1 Gross photograph illustrating a well encapsulated large tumour mass .Cut section was grey, opaque with homogeneous appearance with a few slits.

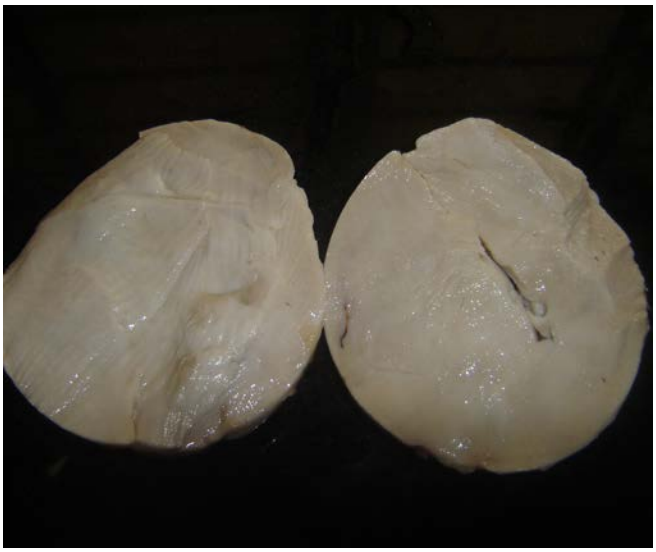

Fig. 2 and 3 Microphotographs to show interanastomosing network of slitlike pseudovascular spaces present within increased collagenised stroma.

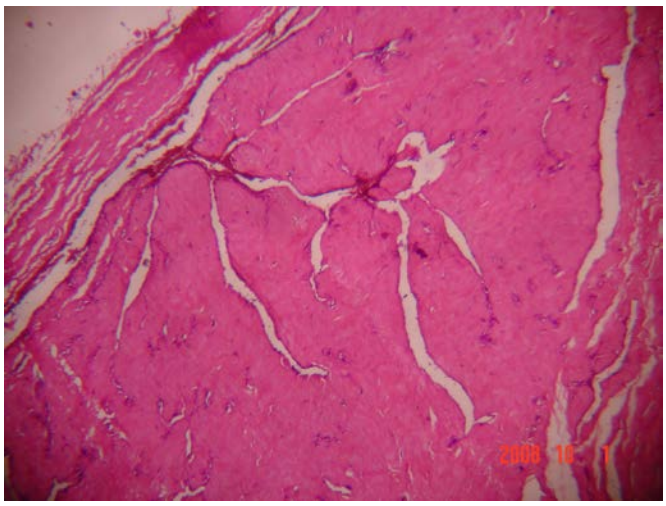

Figure 3

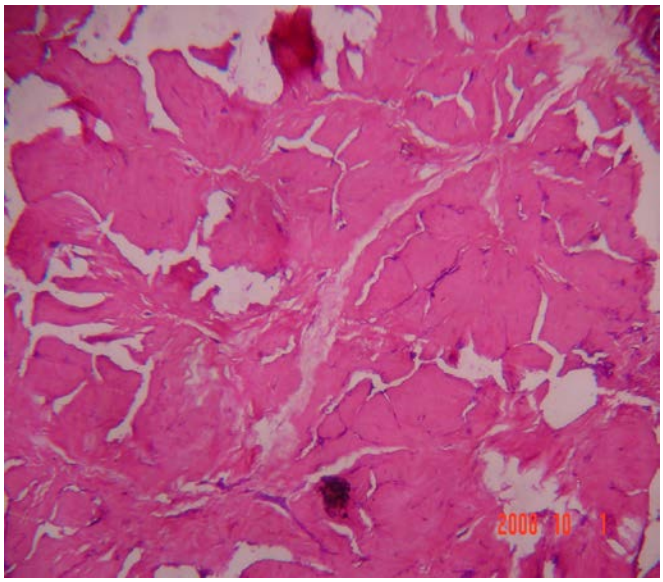

Figure 3