

Breast Pseudoangiomatous Stromal Hyperplasia challenges

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ABSTRACT

Pseudoangiomatous stromal hyperplasia (PASH) is a rare proliferative breast disease. It is characterized by fibrosis of the mammary stroma surrounding breast ducts and lobules with formation of anatomizing capillary-like spaces lined by myofibroblastic cells. Commonly, patients present with a palpable breast mass or continuous breast enlargement. We report a case of a 25 year old lady who presented with painless unilateral swelling of her right breast. Ultrasonography showed heterogeneous, hypoechoic benign appearing breast tissue. Histopathology revealed dense collagenous stroma separated by slit like spaces that were lined by bland spindle cells showing no cytological atypia. With immunohistochemistry, these cells were positive for CD-34 and SMA and negative for the endothelial marker CD-31. Some of these cells were also positive for ER and PR.

KEY WORDS: Benign breast, PASH, Fibroadenoma.

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INTRODUCTION

Pseudoangiomatous stromal hyperplasia (PASH), first reported by vuitch et al., is a benign proliferative disease of the breast.¹ It is characterized by proliferation of stromal spindle cells with an

intervening network of complex anastomosing channels lined by slender spindle cells.² PASH usually presents as painless, single or multiple, palpable breast masses, or as continuous breast enlargement.³ In this report, we present a case of a 25 year old lady who presented with painless single swelling of her right breast.

CASE REPORT

A 25 year old unmarried woman presented to the surgical services of Sultan Qaboos University Hospital with a two week history of swelling in her right breast. She had noticed the lump incidentally while taking a bath. There were no other associated symptoms of pain or nipple discharge and she had no other known medical problems. Her age at menarche was 17 years, never used oral contraceptives and had no family history of breast cancer. On physical examination, there was a 7cm x 7cm firm, non tender, mobile lump at 12 clock position in her right breast. The overlying skin including nipple and areola were normal. There were small, non tender mobile lymph nodes in her right axilla. Her left breast and axilla were normal.

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Ultrasonography of the right breast showed benign heterogeneous hypoechoic tissue at 12 o'clock position corresponding to the lump (Fig.1). The rest of the breast parenchyma was unremarkable and there were no other solid or cystic masses. The skin including nipple and underlying ducts were normal. There were multiple subcentimeteric right axillary lymph nodes which were morphologically normal.

Fine needle aspiration cytology (FNAC) was performed and this showed sheets of benign ductal epithelial cells some with apocrine change. Myoepithelial cells were present in the background in keeping with a benign lesion.

She subsequently had a core biopsy which showed markedly sclerotic intralobular and interlobular breast stroma with widely spaced breast ducts and lobular units. The stroma was cellular and contained complex anastomosing slit like spaces imparting a pseudoangiomatous appearance (Fig.2A). The spaces had a discontinuous lining of spindle cells having elongated nuclei with indistinct nucleoli. There was no cytological atypia or mitotic activity (Fig.2B). Scattered thin walled blood vessels were also present. The breast ducts showed columnar cell change with apical snouts. There was no atypical hyperplasia, DCIS or invasive malignancy.

With immunohistochemistry, the stromal spindle cells were CD-34 and SMA positive (Fig.2C). Occasional stromal cells were positive for ER and PR with more intense staining with the latter (Fig.2D). The vascular endothelial marker CD-31 was negative. The appearances were consistent with pseudoangiomatous stromal hyperplasia (PASH) with columnar cell change.

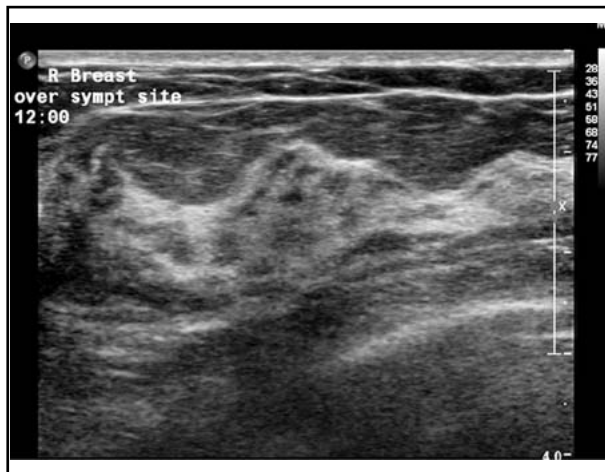


Fig.1: Ultrasound over the symptomatic site shows benign appearing heterogeneous hypoechoic tissue.

DISCUSSION

Pseudoangiomatous stromal hyperplasia is a rare, benign, breast stromal lesion that is classified under benign mesenchymal breast neoplasms in the WHO classification of breast disease.² It is characterized by proliferation of stromal spindle cells with an intervening network of complex anastomosing channels lined by slender spindle cells. It was first described in 1986 by Vuitch et al.¹ The term pseudoangiomatous was proposed due to the resemblance of PASH to low-grade angiosarcoma on low power examination. In view of the benign nature of this lesion, the alternative name 'nodular myofibroblastic stromal hyperplasia' has also been proposed by Leon et al.⁴ PASH usually presents as painless, single or multiple, palpable breast masses, or as continuous breast enlargement.³ Some cases are detected incidentally on imaging.

It presents across a wide age range (from 12 to 65 yrs) but is most common among premenopausal women and therefore tends to be confused with fibroadenoma on clinical examination.⁵ It also occurs in postmenopausal women and is particularly associated with the use of oral contraceptive pills. The size of the lump may vary from 2-15 cms with an average of 5 cm. A few cases have also been reported in males with gynecomastia.⁶ Ultrasonography often shows a well-circumscribed solid mass, with hypoechoic texture with or without heterogeneity, and parallel orientation. Variety of isoechoic and high-echoic cases which indicate small internal cysts may also present.⁷

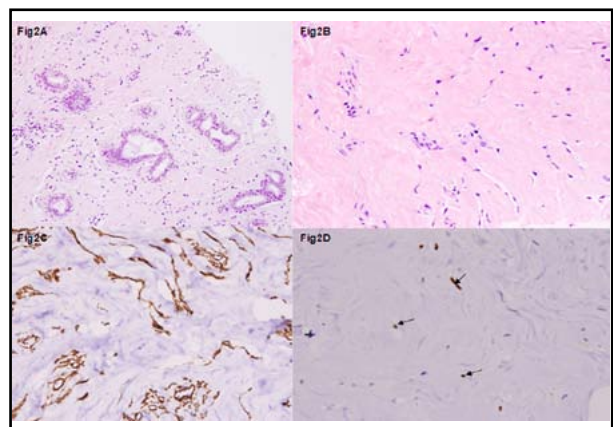


Fig.2A: Low power view showing terminal ducts with columnar cell change. Surrounding stroma appears cellular. Fig 2B: High power view shows anastomosing slit-like spaces lined by spindle cells. These cells are positive for SMA (Fig 2C) and focally positive for PR (Fig 2D, arrows).

Microscopically, there is a spectrum of features that may be seen. The stromal changes range from classical PASH, as in our case, to more proliferative lesions where the spindle cells may even feature a fascicular arrangement. Classical PASH findings are composed of intermixed stromal and epithelial elements, and the lobular and duct structures of the breast parenchyma are separated by an increased amount of cellular fibrous stroma. In the stromal areas, randomly distributed vessel-like slits surrounded by spindle cells are seen. The cells lining these spaces are typically positive for SMA and CD34 in keeping with their myofibroblastic origin. These lesions are thought to arise due to the influence of hormones. This hypothesis is supported by the fact that these cells express hormone receptors particularly PR. The main differential diagnosis is with a phyllodes tumour and angiosarcoma. PASH like changes is also incidentally detected with other breast lesions such as fibrocystic disease, fibroadenomas, columnar cell lesions, lobular neoplasia and in situ and invasive ductal carcinoma. In one of the largest case series on PASH by Gresik et al, out of 80 cases of biopsy proven PASH, 30% also had an associated in situ or invasive carcinoma.⁶

The standard treatment of tumorous PASH is wide local excision. Incomplete excision may result in local recurrence⁵, with the need for subsequent re-excision or even mastectomy in cases where there is breast deformity. A careful observation with breast ultrasound is an option for patients not at high risk for breast cancer.⁸

In our case, the patient refused to undergo surgery and opted for close regular surveillance with breast ultrasound every three months.

CONCLUSION

A case of PASH presented with the classical presentation of a painless unilateral palpable breast lump. She refused to have wide local excision and opted for regular surveillance. Since PASH may co-exist with other lesions, including malignant ones, it is important to ensure that the histopathological features correlate with clinical and radiological findings and that the core biopsy is representative of the lesion.

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