Pseudo-Angiomatous Stromal Hyperplasia (PASH) of the Breast: A Rare Entity.

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ABSTRACT

Pseudoangiomatous stromal hyperplasia (PASH) is an extremely rare benign mesenchymal proliferative lesion of the breast. Its significance lies mainly in differentiating it from an angiosarcoma of the breast. We present here two cases of PASH, both cases having different clinical presentations, different biologic characteristics and consequently different lines of management.

Introduction

Pseudoangiomatous stromal hyperplasia (PASH) is a benign mesenchymal proliferative lesion of the breast. Till 2005, only 109 cases had been reported since its initial description in 1986 by Vuitch et al.2,3 It is primarily a diagnosis of pre- and perimenopausal women.2 PASH is frequently a microscopic incidental finding in breast biopsies performed for benign or malignant disease. However, it may also produce a mass lesion.1 Essentially a benign entity, PASH is extremely rare and its significance lies mainly in differentiating it from an angiosarcoma of the breast3. Here, we present two cases of PASH with similar presentations, but differing lines of management.

Case 1

A 56 year old lady presented with the complaints of a lump in the right breast and mastalgia following a fall during which she sustained blunt trauma to the right breast. There was no prior history of similar lumps, no significant family history or history of any other medical or surgical illness. On examination, an ill-defined lump, approximately 1 x 1cm in size was palpated in the lower outer quadrant of the right breast approximately 2cm from the nipple-areola complex. There was localized tenderness over the lump, but no evidence of inflammatory changes or fixation to surrounding structures. No lymph nodes were palpable in the axilla or in the supraclavicular fossae. The opposite breast was normal. Based on these findings, a provisional diagnosis of traumatic fat necrosis was made.

USG of the right breast revealed a 0.5 x 0.5cm hypo-echoic lesion with irregular margins and calcific foci in the lower outer quadrant, 2cm from the nipple areola complex.

Mammography showed a BIRADS class IV lesion in the right breast.

USG guided FNA showed atypical duct epithelial cells.

An ultrasound guided core biopsy of the lesion was inconclusive.

The patient was therefore posted for an excisional biopsy of the mass. A wide local excision of the mass was performed under local anaesthesia and sedation.

Histopathology report of the specimen showed sclerotic breast parenchyma with normal duct epithelial cells, foci of fibrotic changes with apocrine metaplasia and a few slit like spaces. Features were seen to be consistent with Pseudoangiomatous Stromal Hyperplasia (PASH).

IHC showed the tumour to be hormone receptor (ER- PR) negative.

Fig. 1: Case 1; Actual slide photograph showing anastomosing slit like spaces, on H&E staining.

Fig. 2: Case 1; Immunohistochemistry showing ER/PR negative status.
Case 2

A 27 year old female presented with a lump in the left breast since 2 months with minimal discomfort, not associated with fever or constitutional symptoms.

On examination she had a 2cm lump in the left axillary tail of Spence, which was firm in consistency, nontender, freely mobile and not involving the overlying skin. Opposite breast and both axillae were normal.

An ultrasonography showed a hypoechoic conglomerate appearing like a lymph node mass with altered hilum in left axilla of size 2.4 x 2.2 x 1 cm with echogenic surrounding parenchyma. Sub-centimeter lymph nodes with intact hilum were also seen in left axilla.

A core biopsy of the breast lump was done which showed minimal epithelial component with an occasional compressed and few dilated ducts. Some of the latter showed usual type of ductal hyperplasia as well. The stroma was abundant and contains proliferating myofibroblasts, vessels and collagen fibres suggestive of PASH.

Discussion

The patient in Case 1 was a 51 year old, peri-menopausal lady who presented with a breast lump, which on imaging was shown to be a BI_RADS IV lesion. Since the FNAC showed atypical duct epithelial cells and the core needle biopsy was inconclusive, the suspicion for malignant disease was high and hence an excisional biopsy was performed. It was only after the histopathological analysis of the tumour that it was diagnosed as PASH. In the largest case series of PASH till date, only 42% cases were diagnosed with core needle biopsy whereas 58% cases required an excisional biopsy for the diagnosis. The recurrence rates of PASH after excision are reported to range from 15 to 22% although longer follow-up studies are needed to evaluate the recurrence rate. Since the tumour was ER/PR negative further treatment was necessary for this patient apart from wide local excision, although regular follow-up has been maintained to watch for recurrence.

The patient in Case 2 was a young female, and since a confirmatory diagnosis was achieved on core biopsy, and the patient had a very low risk of malignancy, she was offered watchful waiting with regular follow-up.

PASH usually presents as a clinically palpable breast lump in women with the mean age at presentation being 37 years. The mass, typically unilateral, is usually diagnosed clinically as a fibroadenoma. Sonography demonstrates most lesions to be hypoechoic in echotexture. Mammography findings are indistinguishable from those of the more common type of fibroadenoma, and they are categorised as BI_RADS type 3 lesions.

Grossly it appears as a well-circumscribed tumor with a firm white-gray cut surface. Microscopically, classical PASH appears as anastomosing slit-shaped spaces outlined by flat, bland spindle cells. The spindle cells are vimentin and CD34 positive and factor VIII negative. In more cellular fascicular variants, the stromal cells are found to be desmin and actin positive. Reactivity for progesterone receptor (PR) typically exceeds that of estrogen receptor (ER) in the nuclei of stromal and glandular cells. One case series reported 95% of the tumours to be ER or PR positive.

The pathogenesis of PASH remains uncertain, but aberrant reactivity of myofibroblasts to endogenous or exogenous hormones is likely to be an important factor. Recurrences are infrequent and simple excision is adequate as initial treatment. The importance of this benign lesion, lies in its distinction from angiosarcoma.

Conclusion

PASH is a rare entity which presents as a mass lesion in the majority of cases. In spite of being a benign lesion, it’s similarity to an angiosarcoma warrants a wide local excision.

Ethical Approval and Consent

All procedures performed in this study were in accordance with the ethical standards of the institutional research ethics committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from all individual participants included in the study.

Disclosure

The authors report no conflict of interest.

References