Tumour forming Pseudoangiomatous stromal hyperplasia: a case report

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ABSTRACT

Tumor-forming Pseudoangiomatous stromal hyperplasia (PASH) is a rather uncommon breast lesion and only very few cases of tumor-forming PASH were reported from 1986, since the lesion was originally described. In contrast, focal, non-tumor forming PASH may be an incidental microscopic finding in up to 23% of breast biopsies. PASH is a benign proliferative lesion of the breast stroma that is characterized by slit-like pseudovascular spaces lined by endothelial-like spindle cells. It is indeed rare for a discrete breast mass to have PASH as the main pathological feature on histopathology. The breast mass, typically unilateral, was usually diagnosed clinically as a fibroadenoma. When found in tumour form, PASH most commonly manifests as a single, circumscribed, palpable mass in a premenopausal female. The main differential diagnosis histologically are fibroadenoma, phyllodes tumor and angiosarcoma. However, PASH is a benign condition with very good prognosis. Hence possibility of PASH in nodular or tumour form has to be considered when approaching a case of breast lump whose management is different.

We report here a case of PASH in tumour form from our Institute.

Keywords: Breast lump, Core needle biopsy, Hyperplasia, Mammary hamartoma, Myofibroblast, Pseudoangiomatous stromal, Pseudovascular

INTRODUCTION

It is rare for a discrete breast mass to have PASH as the main pathological feature on histopathology even though it can be present along with other disease entities as an incidental finding.1,2 Thus tumour or nodule forming PASH is very rare indeed with not more than few hundreds of cases reported since it was first described in 1986. PASH is a benign, localized, mesenchymal proliferative lesion with possible hormonal etiology.2 It is characterized by the presence of slit like spaces in dense collagenous stroma. The spaces are lined by benign spindle cells, which show myofibroblastic differentiation and express progesterone receptors.4,5 It can be confused with vascular tumors such as low grade angiosarcoma and cystosarcoma phyllodes, and may require immunohistochemistry studies when in doubt. Local surgical excision with adequate margins is the recommended treatment for tumorous PASH.4,6,7 The recurrence rates of PASH after the excision ranges from 15 to 22%.7 Mastectomy is not generally recommended. It is not considered a premalignant condition and prognosis is excellent.14

CASE REPORT

40-year-old housewife presented with complaints of lump on the right breast, which was slowly growing. There was pain associated with the swelling since last 2 months which was causing discomfort to the patient. But there
was no history of any symptoms of inflammation. There was no nipple discharge or skin changes. She has two children and underwent sterilization after her second delivery. Her menstrual history was regular with mild dysmenorrhea during the initial couple of days. Clinically her general and systemic examination was within normal limits. On breast local examination, her right breast was larger than the left and revealed a 6×4 cm lump centrally occupying all the quadrants but more towards the lower quadrants. There was no attachment to skin or underlying fascia. No axillary lymph node enlargement. A provisional diagnosis of fibro adenoma and a differential of Phyllloides tumour was made.

**Investigations**

Routine investigations were within normal limits.

US of breast showed a discrete oval homogenous hypoechoic mass measuring 5.1×2.1 cm in the retro areolar region occupying all quadrants and was reported as BIRADS III. MMG (Mammogram) showed a radioopacity on the right breast which was discrete, well defined and retro areolar consistent with BIRADS III. An FNAC was done to arrive at tissue diagnosis. Cellular smear showed uniform cohesive duct epithelial cells in mono layered sheets clustered and grouped along with bare nuclei and occasional stromal fragment in an eosinophilical back ground suggestive of Fibroadenoma.

**Figure 1:** US finding of a discrete homogenous hypoechoic mass lesion.

It was decided to do a CNB (core needle biopsy) on this patient. The core biopsy was reported as breast parenchyma containing occasional ducts lined by epithelial and myoepithelial cells surrounded by proliferation of stromal cells forming clefts lined by spindle cells seen scattered in a dense hyalinized stroma. There was no atypia. Adjoining fibro-fatty area showed focal haemorrhage. The CNB report was compatible with PASH.

**Figure 2:** MMG showing discrete radiopacity on the right side.

**Management**

Being a relatively large size of the mass and patient was developing continuous pain which was causing discomfort, she was keen on removing the mass. A wide local excision was planned. After obtaining consent and discussing various related factors with the patient, she was posted for surgery. The patient underwent wide local excision of the mass under GA.

The HPE (Histopathology) showed lesions composed of stromal and epithelial structures with increased amount of stroma seen surrounding lobules and duct structures. Collagenization of interlobal stroma with few ducts showing epitheliosis noted. The report was consistent with PASH.

**Figure 3:** The swelling after wide local excision. Specimen cut through the center to show the homogeneity of the mass lesion.
Patient remained symptom free and was found comfortable during the first follow up after one month. Patient was reassured and asked for follow up after a year.

**DISCUSSION**

Tumor-forming Pseudoangiomatous stromal hyperplasia (PASH) is a rather uncommon breast lesion and only very few hundreds of cases of nodule/tumor-forming PASH reported from 1986, since the lesion was originally described by Vuitch et al.\(^1\) In contrast, focal, non-tumor-forming PASH may be an incidental microscopic finding in up to 23% of breast biopsies.\(^2\) Pseudoangiomatous stromal hyperplasia (PASH), is a type of benign proliferative lesion of the breast stroma that is characterized by slit-like pseudo vascular spaces lined by endothelial-like spindle cells.\(^3\) It is also known as nodular myofibroblastic stromal hyperplasia of the mammary gland.\(^3\)

The breast mass, typically unilateral, was usually diagnosed clinically as a fibroadenoma.\(^4\) The tumoral form of PASH most commonly manifests as a single, circumscribed, palpable mass in a premenopausal female.\(^5\) The mass is usually large which may vary from 2-12 cm. The presenting age ranges from 14-65 years. But in most cases, it is premenopausal or perimenopausal.\(^6\) Clinically It is hard to differentiate a PASH from a fibroadenoma.

The etiology of mass-forming PASH is not clearly known. Vuitch and colleagues and Rosen et al 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.\(^1,7\) It is generally considered that hormones, especially progesterone, can contribute to the formation of PASH. The differential diagnoses of PASH include low-grade angiosarcoma, myofibroblastoma, fibroadenoma, and mammary hamartoma. Of these, angiosarcoma is a malignant condition which has to be ruled out.

Unfortunately, imaging features of PASH are non-specific. On ultrasound, PASH tends to be an oval, round hypoechoic mass or can presents as a heterogeneous mass with cystic areas.\(^8,12\) According to Cohen et al, when a focal lesion with well-defined borders, containing no calcifications on mammography or a well-defined hypoechoic mass on ultrasound is seen, PASH can be considered and included in the differential diagnosis.\(^8\) On mammography, the most common appearance described is a well-defined, uncalcified mass, with regular borders. Spiculated borders, suspicious borders, and architectural distortion can also be seen but are uncommon.\(^9,12\) Fine-needle aspiration and cytologic examination is not very helpful in making definitive diagnosis. Tumorous PASH does not have any unique features on cytology that can help in making the accurate diagnosis. Differential diagnoses on cytologic examination include fibroadenoma, phylloides tumor, or fibrocystic change due to the overlapping cytologic features. The major utility of cytologic examination lies in ruling out malignant lesion rather than in providing the definitive diagnosis.\(^13\) The definitive diagnosis is only through histopathological examination along with immunohistochemical analysis if HPE is inconclusive. The most important differential diagnosis is angiosarcoma and it must be distinguished from PASH.

Because of the rarity of pseudoangiomatous stromal hyperplasia tumors and uncertainty about their clinical behavior, surgical excision has been the recommended treatment.\(^1,4,6\) However in a recently published study showed that non-surgical management strategies can be considered for patients who refuse a surgical procedure and options may be acceptable, especially when the lesion is small and triple assessment has been performed to exclude a malignancy.\(^14\) Some reports document an impressive response to tamoxifen in a patient presenting with breast enlargement, pain, and breast masses.\(^15\) Tumorous PASH is slow growing and there is a chance to recur after excision. The recurrence rates of PASH after the excision ranges from 15 to 22%.\(^1,5\) Overall, the prognosis for PASH is excellent. Pseudoangiomatous stromal hyperplasia is not considered a premalignant lesion or a risk factor for malignancy.\(^14\) Only 1 single instance has been reported in which invasive ductal carcinoma was present within tumorous PASH, and the authors considered that to be an incidental finding rather than true malignant transformation.\(^14\)

**CONCLUSION**

PASH is a benign, localized, mesenchymal proliferative lesion with possible hormonal etiology. It is characterized by the presence of slit like spaces in dense collagenous stroma.

The spaces are lined by benign spindle cells, which show myofibroblastic differentiation and express progesterone receptors. It can be confused with vascular tumors such as low grade angiosarcoma and cystosarcoma phylloides, and may require immunohistochemistry studies when in doubt. Local surgical excision with adequate margins is the recommended treatment for tumorous PASH. Mastectomy is not generally recommended. It has an excellent prognosis, with very low risk of recurrence.

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**REFERENCES**
