

Pseudoangiomatous stromal hyperplasia causing severe breast enlargement in a 15-year-old girl: A case report

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH), composed of proliferated stromal mesenchymal cells of myofibroblastic origin, is a benign lesion of the breast. A few cases associated with pseudoangiomatous stromal hyperplasia of the breast have been reported. We report this case of a 15-year-old girl with PASH accompanied by severe enlargement and painful mass in the right breast. There were no other palpable masses or lymph nodes. Biopsy of the mass showed histopathologic features characteristic of fibroadenoma. The palpable mass was around 6x5cm in diameter, while clinical manifestations aroused suspicion of malignancy mimicking sarcoma of the breast. Although, biopsy of the mass showed benign histopathologic features; surgical excision was performed because of the damage caused by enlarging breast tissue and clinical suspicion of malignancy.

Keywords: Pseudoangiomatous, Stromal hyperplasia, Breast tumour.

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Introduction

The definition of Pseudoangiomatous Stromal Hyperplasia (PASH) was originally proposed by Vuitch et al in 1986.¹ PASH has a histological structure showing areas such as complex inter-anastomosing slit-like empty spaces surrounded by isolated spindle cells.¹ It is often confused with various benign conditions and rapidly growing sarcomas of the breast due to both clinical manifestations and radiological and pathological findings. The definition criteria of PASH is still unclear. It is important to determine the pathology of the breast lesion before surgical excision because it changes the surgical approach.² Usually a definitive diagnosis is made after surgical excision.

Case Report

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A 15-year-old girl presented to the department of general surgery, at Sisli Hamidiye Etfal Medical Practice and Research Centre, Istanbul in February 2018 with a six-months history of progressive painful enlargement of the right breast. She reported menarche at the age of 11 and had a normal menstrual cycles since. There was no personal or family history of breast disease or breast and ovarian cancer.

On clinical examination, the girl showed unilateral and asymmetrical right breast enlargement and a palpable mass around 6x5cm in diameter in the middle of the lower quadrant of the right breast while no palpable mass was found in the left breast. There were no evidence of breast swelling, change in skin colour and pitting or thickening of the skin.

As a part of examination, breast ultrasonography with a high frequency (6-11 MHz) linear array head was performed. It showed normal breast skin thickness and subcutaneous fatty tissue, while bilateral breast parenchyma structure was sclerosing. Hypoechoic mass measuring 5x4 cm in diameter was observed. Both axilla showed lymph nodes which appeared normal in size and looked benign.

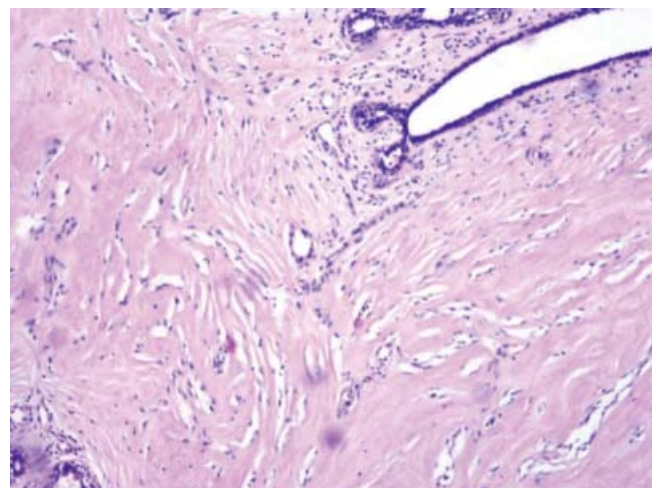


Figure-1: Pseudoangiomatous stromal hyperplasia, anastomosing endothelial vascular-like structures irregularly dispersed in hyalinized fibrotic interlobular stroma (haematoxylin and eosin, magnification $\times 100$).

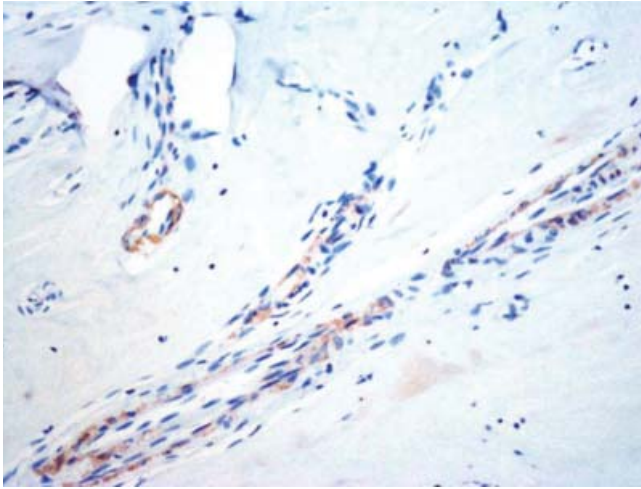


Figure-2: Pseudoangiomatic stromal hyperplasia, anastomosing endothelial vascular-like structures irregularly dispersed in hyalinized fibrotic interlobular stroma (haematoxylin and eosin, magnification $\times 200$).

Phyllodes tumour and angiosarcoma were considered as differential diagnoses. Thus sampling was advised by the Breast Imaging-Reporting and Data System (BIRADS 3).

Tru-cut biopsy, performed under ultrasound guidance, revealed fibroadenoma. Due to clinical suspicion and damage of the breast tissue, the patient was subjected to a surgical excision procedure. Surgical excision was done using an inferior circumareolar incision. Postoperatively, the patient had excellent cosmetic outcome and breast symmetry.

Macroscopic examination of the excised material showed well-demarcated, yellow-white external surfaces and

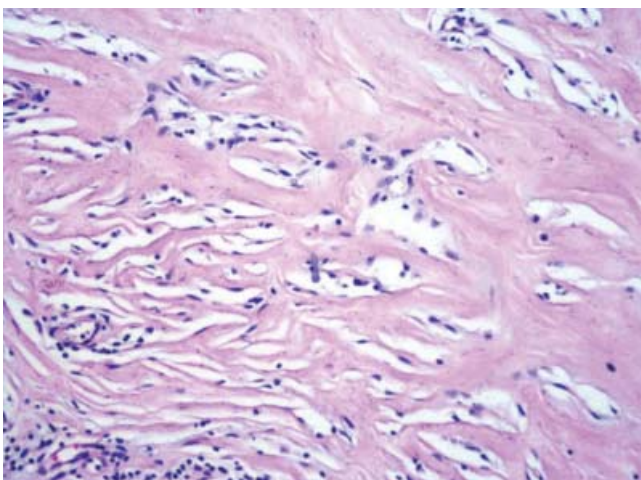


Figure-3: Anastomosing slit-like spaces and stromal cells are strongly positive for endothelial marker CD34 diagnosing PASH (CD34 immunohistochemical staining $\times 10$).

nodular structure. The final histopathology after excision was in correlation with the diagnosis of PASH (Figure-1,2). Immunohistochemistry was positive for CD34, while factor VIII, desmin, calponin, CD31, panck, somatomedin A(SMA), Multiplication-stimulating activity (MSA) and progesterone and oestrogen receptors were negative (Figure-3). There was no evidence of atypia or pleomorphism.

Discussion

Pseudoangiomatic stromal hyperplasia (PASH) is a benign proliferation of the breast stromal tissue. It is usually found incidentally during routine breast biopsies. Diagnostic rates vary from institution to institution due to the knowledge and abilities of the pathologists. The reported age for patients with PASH ranges between 14-67 years but most patients are between the ages of 30 to 40 years.³ Current case involved a large PASH that had grown rapidly over a period of six months in a 15-year-old patient; this is rarely seen in this age group. Most cases of PASH occur as painless, stiff, palpable masses and usually do not increase significantly in size. While, as in our case, many cases of PASH that mimicked local advanced malignancies have been reported.⁴

On ultrasonography, PASH is seen as well-defined hypoechoic or isoechoic masses.⁵ These tumours cannot be distinguished from fibroadenomas by radiological imaging methods such as mammography and ultrasonography. In our case, ultrasonography showed a well-circumscribed hypoechoic mass that could not be identified from fibroadenoma. Only 40% of the cases were diagnosed incidentally by radiological imaging because PASH is a rare disease and does not show significant imaging features.⁶ Thus, histological studies are required for definitive diagnosis. Fine needle aspiration cytology (FNAC) is often inefficient, and core needle biopsy is necessary for differential diagnosis, although in some cases, definitive diagnosis can be reached after surgical excision.

PASH is usually diagnosed as incidental microscopic foci during the diagnosis of benign or malignant breast diseases.⁷ The most important differential diagnosis on histopathological examination is angiosarcoma of the breast. Unlike angiosarcoma of the breast, PASH has good prognosis with low risk of recurrence and no need for additional treatment. Histologically, PASH is composed of complex inter-anastomosing slit-like spaces within the parenchyma of the breast surrounded by spindle cells. PASH does not contain blood cells within these structures and lacks cellular atypia or mitotic activity.⁸ Immunohistochemically, PASH is positive for CD34 and

vimentin and negative for factor VIII-related antigen and cytokeratin. The malignant cytological features and positive immunohistochemical staining, including CD31 and factor VIII related antigen helps distinguish low-grade angiosarcomas from PASH.⁹ Our patient had atypical clinical presentation of PASH. Initially, growth rate indicated malignancy and imaging findings imitated a fibroadenoma and tru-cut biopsy was performed. The possibility of malignancy could not be excluded by imaging findings and tru-cut biopsy. After surgical excision histopathology and immunohistochemical findings confirmed the diagnosis of PASH which is an extremely rare diagnosis.

PASH occurs mostly in hormonally active premenopausal women. Most of the reported cases in literature demonstrate the presence of progesterone receptor activity and weak or no oestrogen receptor activity.¹⁰ Our case was negative for both oestrogen and progesterone receptors. We reported positivity for CD34 in our case which is consistent with the findings in literature and supports the myofibroblastic stromal origin of these lesions.

Conclusion

In summary, we present the case of a painful and rapidly growing mass in the right breast, was diagnosed with PASH in consideration of its clinical features and histopathological findings. After physical examination, sonography and fine-needle aspiration or core biopsy are used for differential diagnosis. Our case implies that imaging studies are not required for definitive diagnosis in adolescent females because the final diagnosis is determined by surgical approaches. If PASH is proven by biopsy, it can be followed regularly. If there is a large mass or clinical suspicion and if the histology does not confirm the diagnosis preoperatively, surgical approaches are recommended.

Disclaimer: None to declare.

Conflict of Interest: None to declare.

Funding Sources: None to declare.

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