

PSEUDOANGIOMATOUS STROMAL HYPERPLASIA IN A YOUNG GIRL: A CASE REPORT

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ABSTRACT

Vuitch et al first reported Pseudoangiomatous stromal hyperplasia (PASH) as a benign proliferative breast disease. It is thought to be hormone sensitive and represents a hyper-response to estrogen and progesterone resulting in myofibroblast proliferation. It is commonly reported histologically in breast biopsy specimen. It usually occurs in peri and premenopausal women, but may also occur in adolescent, postmenopausal women on hormone replacement therapy and rarely in men. Definitive diagnosis is challenging as it mimics with other benign breast lesions. Less than 20 cases of diffuse and less than 200 cases of tumorous PASH has been reported so far. Its association with malignancy is very rare. Here we report a case of giant pseudoangiomatous stromal hyperplasia in a young girl presented with asymmetrical enlargement of her right breast.

Introduction

Vuitch et al first reported Pseudoangiomatous stromal hyperplasia (PASH) as a benign proliferative breast disease.¹ It is thought to be hormone sensitive and represents a hyper-response to estrogen and progesterone resulting in myofibroblast proliferation. It is commonly reported histologically in breast biopsy specimen. It usually occurs in peri and premenopausal women, but may also occur in adolescent, postmenopausal women on hormone replacement therapy and rarely in men. Definitive diagnosis is challenging as it mimics with other benign breast lesions. Clinically the main differential would be fibroadenoma and phyllodes tumor and low grade angiosarcoma on histology. Less than 20 cases of diffuse and less than 200 cases of tumorous PASH has been reported so far.² Its association with malignancy is very rare. Here we report a case of giant pseudoangiomatous stromal hyperplasia in a young girl presented with asymmetrical enlargement of her right breast.

Case Summary

A 17 year old girl came to our department for breast ultrasound. She presented with complain of asymmetric enlargement of her left breast for last 3 months. She has no family history of breast cancer and any other comorbid.

On ultrasound a large hypoechoic solid lesion was present occupying almost entire left breast parenchyma. Mild vascularity also seen on Doppler examination. Imaging features favor the differentials of giant fibroadenoma / phyllodes tumor. (Fig.1) Mammogram was also performed on clinician's request that showed a large high density mass lesion compressing the breast parenchyma. There were no calcifications. Axilla was unremarkable. (Fig.2a,2b) Computed Tomography (CT) showed a large well defined enhancing mass lesion posteriorly closely abutting the chest wall. (Fig.3a,3b) Rest of the scan was unremarkable and no deposits were present.

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Ultrasound guided core needle biopsy of the lesion was performed and histopathology results showed breast tissue exhibiting proliferation of interconnecting narrow spaces in dense collagenous stroma with stromal cells highlighted on CD34. (Fig.4) The diagnosis was Pseudoangiomatous stromal hyperplasia (PASH).

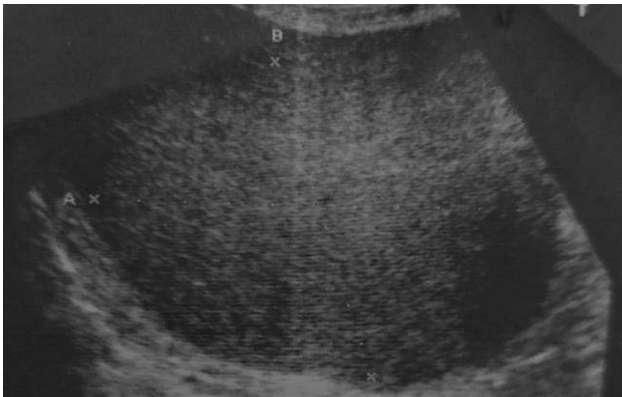


Figure 1: Ultrasonographic image showing a large hypoechoic solid lesion

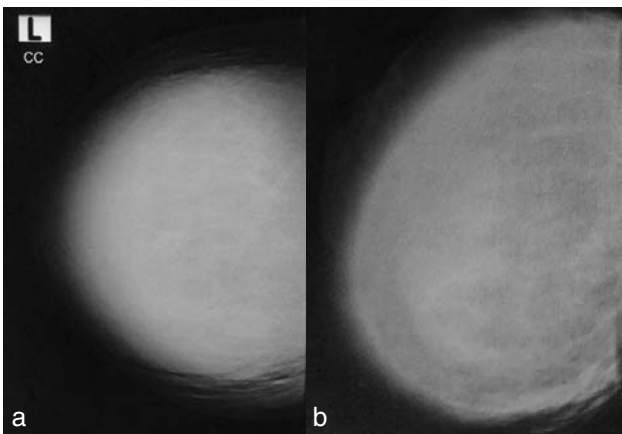


Figure 2a,b: Craniocaudal and mediolateral oblique views showing a large high density mass lesion occupying left breast

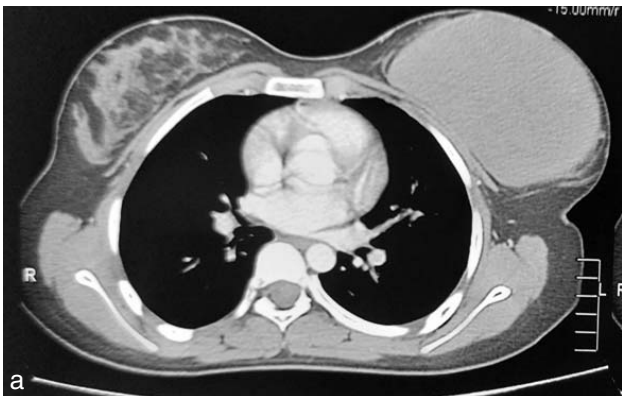


Figure 3a,b: Axial and sagittal images of CT Scan showing a large enhancing lesion in left breast

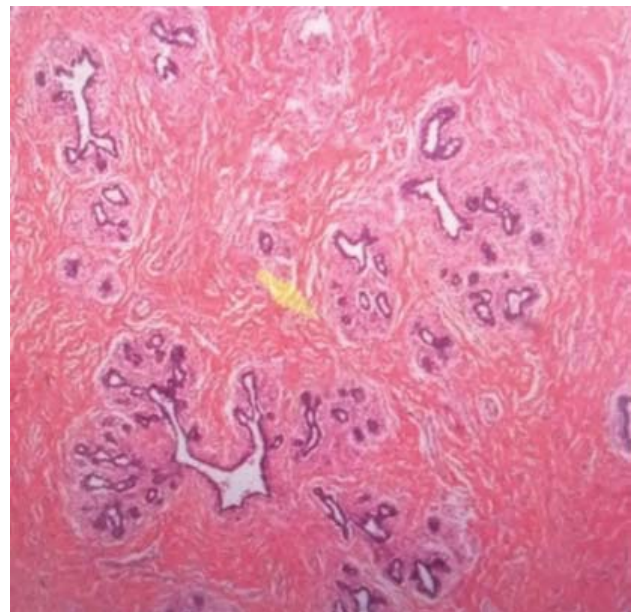


Figure 4: Histopathological image showing proliferation of interconnecting narrow spaces in dense collagenous stroma with stromal cells highlighted on CD34

Discussion

Pseudoangiomatous stromal hyperplasia (PASH) is commonly found incidentally on biopsy and rarely present as a symptomatic lesion. When symptomatic

it may present as a firm mobile lump or asymmetric enlargement of breast. It may present as multiple nodules and can occur bilaterally.³

On mammography, its features are non specific and according to one study, in approximately 68% of the cases no definite mammographic abnormality was noted.⁴ It may present as a well circumscribed mass or an asymmetric density. Calcifications are rare. In this case a large high density mass was seen almost completely occupying entire breast.

On ultrasound, it presents as an oval or rounded hypoechoic mass like fibroadenomas or heterogeneous mass with cystic areas mimicking phyllodes. Like in this case, ultrasound features were in favor of giant fibroadenoma / phyllodes. Only few cases of PASH have been reported on MRI. High signal slit like spaces may be seen on T2 weighted images and it usually exhibits Type I-time signal intensity curve.⁵

Definitive diagnosis can only be made on histopathology. Histologically, it mimics low grade angiosarcoma but lacks invasive features. Multiple slits like spaces with stromal proliferation is pathognomonic of PASH. On immunohistochemical stain, CD34 is usually positive.⁶ In our case, histopathology showed breast tissue exhibiting proliferation of interconnecting narrow spaces in dense collagen stroma with positive CD34 expression.

Management of PASH depends upon the clinical presentation. If asymptomatic, it can be followed on mammography and ultrasound yearly for up to 36 months. If symptomatic, surgical removal is recommended. Surgery may include lumpectomy or skin sparing mastectomy. Recurrence after complete surgical removal has also been reported.⁷ As a noninvasive management, anti-hormonal therapy can also be given as it is hormone sensitive. In this case surgical excision was done because of its massive enlargement and pain.

Case was approved by ERC of DUHS.

Conclusion

PASH is a benign breast entity often diagnosed incidentally. Less commonly it presents as a palpable mass with nonspecific features radiologically. Histopathological correlation is necessary for definitive

diagnosis. Both surgical and conservative treatment can be advised. Recognition and reporting of this entity is important for proper management and further surveillance.

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