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Case Report

Pseudo-Angiomatous Stromal Tumor in A 12-Year-Old Female

A Case Report and Literature Review

Marcia Chung M.D*[1]., Albert Wright MD[2], Portia Siwawa MD[3]

1. *Attending general surgeon, Interfaith Medical Center.*
2. *Chairman of Surgery Interfaith Medical Center.*
3. *Attending general surgeon Interfaith Medical Center*

***Correspondence to:** Marcia Chung, 1. Attending general surgeon, Interfaith Medical Center.

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Introduction

Pseudo angiomatous Stromal Hyperplasia is an important entity in children with implications related to treatment modalities and prognosis. It is a benign disease, with no known direct correlation with malignancy. Described initially by Vuitech et al. in 1986, PASH is known as a benign breast mass consisting of mammary stromal proliferation with anastomosing slits which resemble vascular spaces. On sonogram, it is noted to be a slightly heterogeneous mass.

The etiology of PASH has been thought to be a consequence of an aberrant response of a high density of hormonal receptors associated with breast myofibroblasts to endogenous and exogenous hormones.

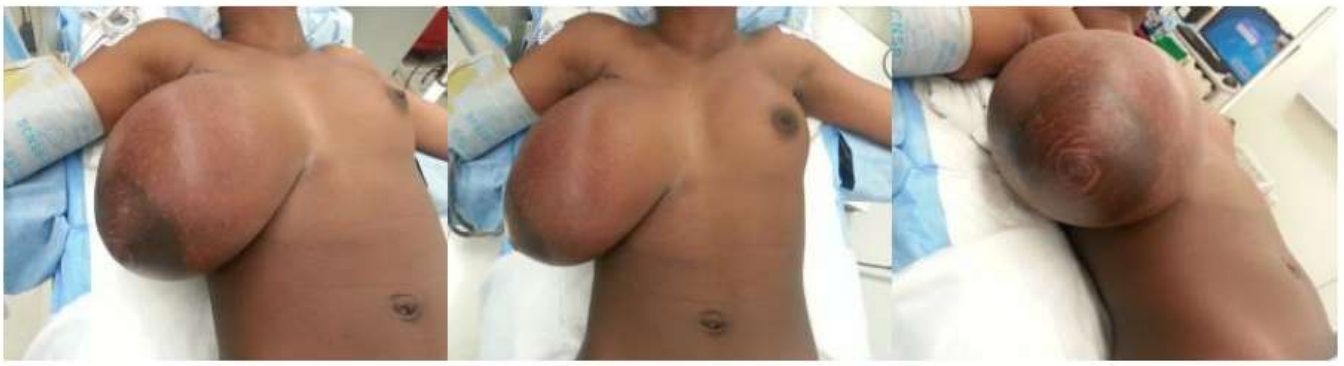
Recurrence has been described with incomplete excision, and its diagnosis therefore requires close follow up especially after surgical intervention.

Histopathological studies have encountered a stronger presence of progesterone receptors in the nuclei of myofibroblasts in PASH, with a more variable presence of estrogen receptors. These findings were supported by the histological changes seen in PASH and the normal breast during the luteal phase of the menstrual cycle. More than 50% of the patient population manifesting tumorous PASH were postmenopausal women receiving HRT. In addition, many premenopausal females diagnosed with the disease were actively taking oral contraceptives, while young male patients had gynaecomastia.

Noteworthy differential diagnoses are: Giant fibroadenoma, Low grade angiosarcoma, phyllodes neoplasm, mammary hamartoma.

Case Presentation

A 12-year-old girl accompanied by her mother, presented to the Interfaith Medical Center breast clinic complaining of increasing heaviness and progressive painless enlargement of the right breast over a three-month period. The patient had enjoyed prior good health with no known medical issues. Menarche had occurred at age 9 years and had been regular from onset. Patient denied prior use of oral contraceptives, or other hormonal medication and denied any sexual activity. No known family history of breast disease; neither benign nor malignant.



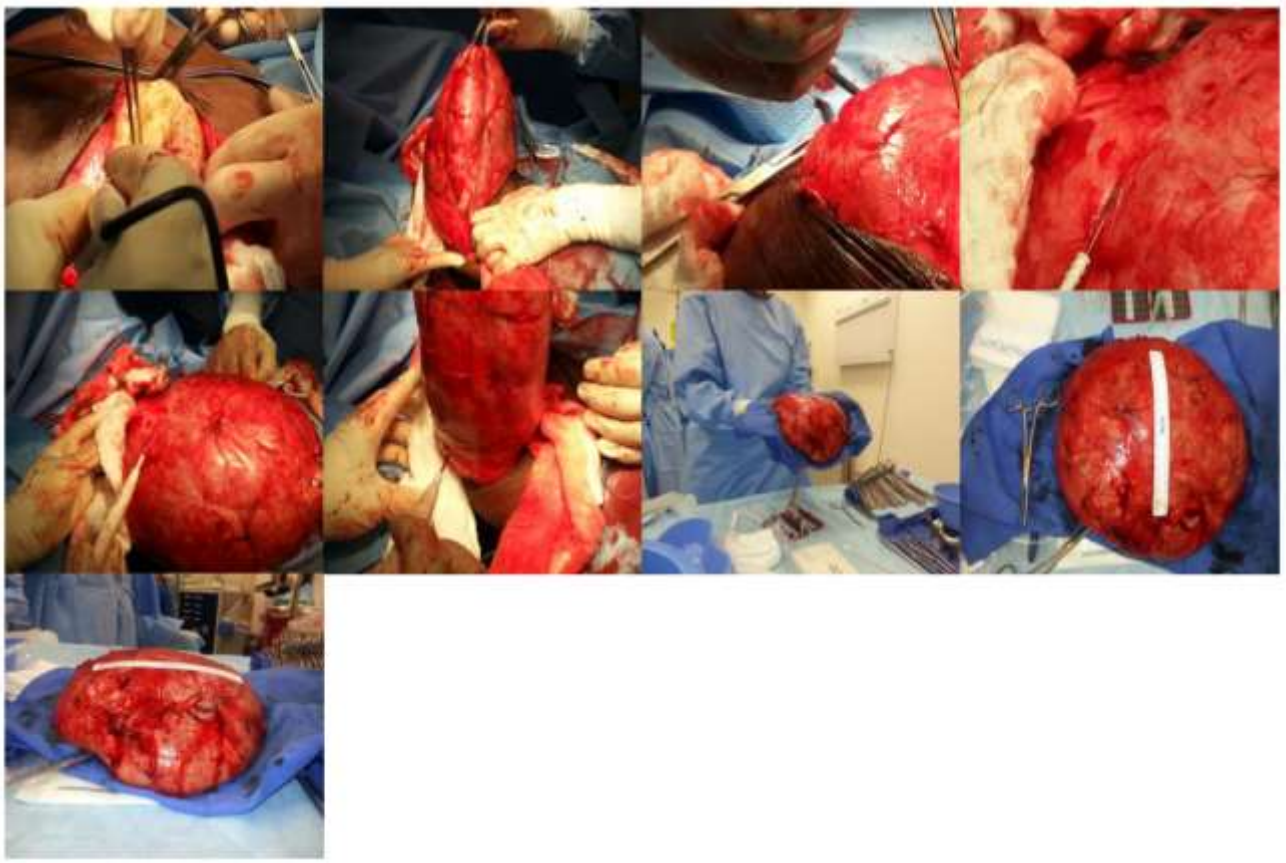
Physical Exam revealed a massively enlarged right breast which was pendulous in comparison to the left which had attained Tanner Stage 4 features. The mass measured approximately 20 to 30cm in size and was heavy. The skin overlying the breast was stretched and the mass grossly appeared well defined. Engorged veins depicting increased vascularity was noted overlying the surface of the breast. Breast was non tender to palpation. There was no skin dimpling and the areola was prominent with no nipple discharge. Palpation of the axillae did not reveal any lymphadenopathy.

Ultrasound of the breast was significant for: No obvious localized mass, diffuse enlargement with several areas of prominent ducts.

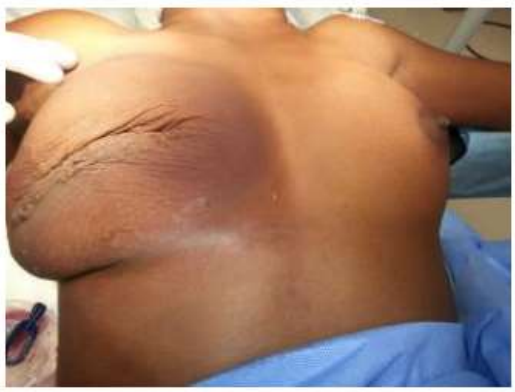
Excisional biopsy was indicated. She and her parents were counseled about the need for tissue diagnosis while maximizing the retention of developing adjacent breast tissue in the affected breast.

Surgery

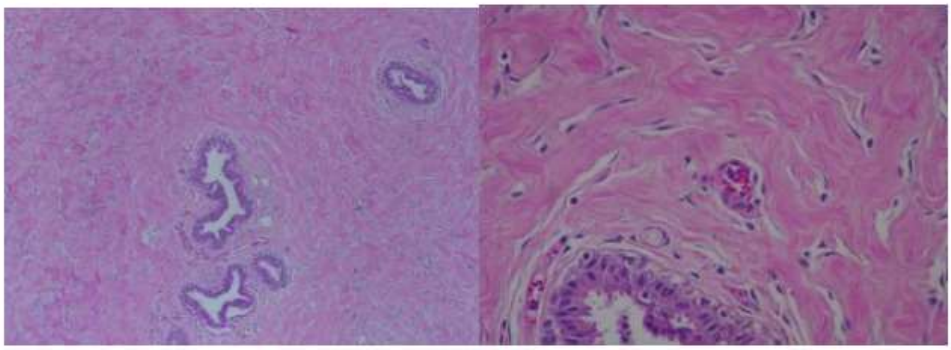
The procedure was performed under general anesthesia via an infra-areolar incision. The solid mass was firm and greyish white in color and appeared well encapsulated. The mass was readily dissectible from the rest of the breast substance and was easily freed completely from it. Posterior to the mass, normal breast tissue was noted, and had been compressed by it.



Wound closure was performed in the standard fashion and a suction drain was left in place. The patient was seen a week later, and the drain was removed. The wound was clean and dry.



Pathology



Marked expansion of interlobular stroma by collagenous fibrotic tissue with numerous slit-like pseudo-angiomatous spaces. H&E stains (x 40.). Spaces outlined by bland spindle myofibroblast cells with uniform small, flat nuclei (x400)

Discussion

Although treatment of PASH may vary, in this patient, excision was indicated. Acknowledging the fact that she was a preteen, care was taken to preserve uninvolved breast tissue posterior to the mass. There was no known family history of breast malignancy and less likely possible at her age, but the rapidly enlarging nature of the lesion warranted operative management while attempting to preserve as much as possible, the developing breast tissue to decrease the likelihood of hypoplasia or aplasia. At 12 month follow up she underwent reconstruction of the breast with native tissue.

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