Case Report

Pseudoangiomatous stromal hyperplasia of the breast: a case report of a 12-year-old girl

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH) of the breast is a benign lesion, characterized by a dense proliferation of stromal mesenchymal cells of myofibroblastic origin forming empty, slit-like channels. We report PASH in a 12-year-old girl with a huge rapidly enlarged right breast. Biopsy of the mass showed histopathologic features characteristic of PASH. Immunohistochemical studies revealed diffuse positive membranous immunoreactivity to CD34. Although it is a benign lesion, lumpectomy was performed to minimize the damage from developing breast tissue.

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Introduction

Pseudoangiomatous stromal hyperplasia (PASH) is an unusual benign breast condition that was first described in 1986 [1]. The reported age range of patients is 14-67 years, although the vast majority of PASH patients present in their late thirties and forties [2]. It is histologically defined as a complex network of slit-like spaces lined by endothelial-like spindle cells against a background of stromal hyperplasia [3]. On histologic analysis, PASH can be mistaken for a low-grade angiosarcoma and phyllodes tumor, but it is a benign condition. Correct identification of PASH of the breast has important treatment implications for patients [3-5].

Case report

A 12-year-old girl, complaining of an enlargement of her right breast, presented for diagnostic work-up. She mentioned that 2 months ago she had palpated a small nodule near the nipple, which thereafter rapidly increased in size. There was no nipple discharge. She had an unremarkable personal and family history, with reported menarche at 11 years and a normal menstrual cycle.

Physical examination revealed an overall enlarged right breast replaced with a huge lump occupying the whole breast, with diffuse tenderness and firmness on palpation. There was no evidence of thickening of the breast skin or nipple retraction. (Fig. 1).
The patient was initially examined with breast ultrasonography, with a high frequency (8-10 MHz) linear array head, which showed a huge right breast heterogeneous hypoechoic mass measuring 17.3 cm at maximum diameter with cystic channels noted within the mass. The ipsilateral axilla showed benign-looking lymph nodes (Fig. 2).

The differential diagnosis at this point included low-grade angiosarcoma, PASH, and phyllodes. Therefore, sampling was advised by the Breast Imaging-Reporting and Data System (BIRADS 4).

Under ultrasonography guidance, core needle biopsy was performed, which showed the presence of spindle cells in the fibrous tissue in a pseudoangiomatous pattern, fitting the diagnosis of PASH. A complex pattern of anastomosing slit-like spaces in a dense collagenous stroma was seen. These spaces are lined with myofibroblasts rather than endothelial cells and do not contain red blood cells. This was the same finding on the final histopathology after excision (Fig. 3-5).

Immunohistochemical studies revealed diffuse positive membranous immunoreactivity to CD34. In addition, negative staining was observed for CD31, a cellular adhesion marker highly restricted to endothelial neoplasms, and for desmin, a marker for cytoplasmic intermediate filaments found in the smooth muscle. The results of this panel of studies confirmed the characteristic immunophenotype of PASH while helping exclude lesions of vascular endothelial and smooth muscle origin such as angiosarcoma.

The patient was finally operated; lumpectomy with reduction mammoplasty was performed. The patient had excellent cosmetic results postoperatively. The patient has been followed up for 6 months now without evidence of recurrence (Fig. 6).

Fig 1 – Image showing the asymmetry and the large mass in the right breast on presentation.

Fig 2 – Ultrasonography showed a huge right breast with well-defined heterogeneous hypoechoic mass measuring 17.3 cm at maximum diameter, with cystic channels noted within the mass and no significant vascularity detected on Doppler imaging.
Discussion

PASH is a benign proliferation of the mammary stromal tissue. On histologic analysis, it shows complex interanastomosing slit-like spaces which appear to be lined by spindle cells in the breast parenchyma [1].

We present a case of a huge, rapidly growing PASH in a 12-year-old patient, which is not frequently seen at this age group; to our knowledge, the youngest female patient reported in literature with PASH is a 10-year-old girl [6]. This tumor mimicked a low-grade angiosarcoma on ultrasonography; true cut biopsy revealed PASH, and surgical excision was performed.

It is important to differentiate the lesion from angiosarcoma. It can be confused histologically with PASH. Angiosarcoma often shows a more aggressive infiltrative pattern into surrounding fibroadipose breast tissue and is highly vascular, lined by endothelial cells. It does not show a collagenous stroma. Angiosarcoma displays positive immunoreactivity for CD31 and, although less frequently and with less sensitivity, for the factor VIII–related or von Willebrand factor antigens [7].

PASH is associated with several benign entities including proliferative and nonproliferative fibrocystic changes, such as fibroadenomas, gynecomastia, normal breast tissue, and sclerosing lobular hyperplasias. The etiology and pathogenesis remain unknown.

Hormonal factors are thought to play a role in the etiology of PASH [8].

Microscopically, PASH consists of a network of slit-like spaces lined by myofibroblasts that resemble vascular spaces. On immunohistochemistry, PASH is positive for CD34 and vimentin and negative for factor VIII–related antigen and cytokeratins [9,10].

On radiologic examination, there are no specific or diagnostic features. Most of these tumors mimic fibroadenomas or hamartomas and angiosarcomas, and larger tumors can be mistaken for phyllodes [9,11,12].

Management with wide local excision is the treatment of choice for PASH owing to its uncertain natural history [13]. 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. The reason for recurrence could be attributed to the growth of a residual mass after incomplete excision [14,15].

Conclusion

PASH is a benign entity that is regarded as a myofibroblastic proliferation of the breast.
In summary, we present the case of a huge and rapidly growing PASH in a 12-year-old patient, which is an uncommonly seen lesion in this age group and is infrequently reported. The lesion reported is one of the largest in the literature and the largest in a teenage girl.

Local excision is curative in most cases. Although recurrence may occur, the overall rate is low. Follow-up after excision is recommended as local recurrence has been reported.

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