Case Report

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Giant Pseudoangiomatous Stromal Hyperplasia of the Adolescent Breast: A Case Report with Emphasis on Image Findings and Literature Review

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign mesenchymal proliferation of the breast with unclear etiology. It is diagnosed in a wide spectrum from incidental microscopic findings to palpable breast mass. Diagnosis is generally made in the premenopausal age group. PASH cases seen in adolescents reported in the literature are very rare and none of them has mentioned the radiologic features in detail. In this case report, we present a 12-year-old patient with giant (150x100x127 mm in size) PASH in her right breast, giving emphasis to the findings of the ultrasonography and magnetic resonance imaging, including contrast-enhanced dynamic and diffusion-weighted imaging sequences. To the best of our knowledge, the present case is the second biggest giant PASH seen in the adolescent age.

Keywords: Pseudoangiomatous stromal hyperplasia, breast neoplasms, adolescent

Introduction

Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign mesenchymal proliferation of the breast that was first described by Vuitch et al. (1) in 1986. Even though the pathogenesis is unclear, either endocrine or exocrine stimulation may play a role in abnormal mesenchymal proliferation. PASH is diagnosed in a wide spectrum from incidental microscopic findings to palpable breast mass (2). It is most commonly seen in middle-aged premenopausal women (3). Only 25 of 200 cases reported as PASH in the literature were seen in the adolescent age group so far, with only 2 of them being more than 10 cm (4-6). In this case report, we presented a giant PASH case in an adolescent, giving emphasis to imaging findings. To the best of the authors' knowledge, this is the second biggest giant PASH seen in the adolescent age.

Case

Informed consent was obtained verbally from the family of the child. A 12-year-old girl was admitted to the hospital with a palpable mass in her right breast that had grown rapidly in the last 8 months. Her menarche age was 11 with normal menstrual cycles. The right breast was asymmetrically big and sensitive on physical examination with no accompanying erythema, temperature change, or nipple retraction. The family history was unremarkable regarding breast cancer. On ultrasonographic (US) examination, a giant hypoechoic heterogeneous giant solid mass with scattered cystic areas inside (Figure 1). In short-tau inversion recovery (STIR) images, the lesion was heterogeneous hyperintense (Figure 2), and in the obtained axial T1W, contrast-enhanced image with a maximum intensity projection, the feeding artery was obviously seen (Figure 3). On performed gadolinium-enhanced magnetic resonance imaging (MRI), a Ta1-weighted image (T1WI) isointense mass lesion with sharp and lobulated contours and 150 x 100 x 127 mm in size was observed. The lesion showed persistent enhancement beginning from the early arterial phase (Figure 4) and no diffusion restriction with an ADC value of 1,327x10^{-3} mm^2/s. MRI features were concordant with the BIRADS-3 lesion.

Due to the rapid increase in size, a tru-cut biopsy under US guidance was performed with a 14-gauge needle after obtaining a written consent form. In the pathology specimens, stromal proliferation with hyalinization around

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scattered sparse ducts was observed (Figure 5). Cells wrapping the pseudovascular spaces showed staining for CD34, vimentin, and alpha-smooth muscle actin and were negative for CD31 and Factor VIII. The pathology report revealed the diagnosis of PASH.

Discussion

PASH is generally seen in premenopausal or postmenopausal women who are under hormone replacement therapy (2). A positive response to anti-estrogen medication like tamoxifen supports the hormonal effect in the pathogenesis of PASH (6). The size of PASH lesions commonly varies between 0.6 and 12 cm (7). However, PASH can present with giant breast lesions in the child age group, as seen in this study. Juvenile fibroadenoma, hamartoma, phyllodes tumors, and less frequent PASH and angiosarcoma were considered in the initial diagnosis of our case. Differential diagnosis with angiosarcoma is critical because it affects treatment.

The diagnosis of PASH radiologically is difficult as imaging features are generally nonspecific. Only 40% of the cases were diagnosed incidentally by imaging (2,7). PASH is generally seen as a hypoechoic, well-circumscribed solid lesion in the US. Solid lesions may contain scattered cystic areas seen as lace-like reticular components (6). Mammographic features of PASH vary from a well-defined mass to an asymmetrical density. The most common mammographic appearance is a noncalcified solid mass or localized increase in stromal composition. However, PASH was identified in only 31% of the patients with palpable masses in their breasts (8). Due to the age of the patient, we did not perform mammography in this study.

In parallel to mammographic imaging, PASH is seen varying from focal and segmental enhancing non-mass areas to well-defined circumscribed masses on MRI. Non-specific T1, T2 weighted imaging contrast enhancement patterns have been described in the literature. Reticular lines seen inside the mass are relatively diagnostic for PASH. This imaging feature was first described by Teh et al. (5) as high signal slit-like spaces on T2-weighted imaging and STIR. In our opinion; this pattern, if evident, is useful for the diagnosis, as it matches with dense collagen deposits that surround the less dense ducts on pathologic examination. Regarding contrast kinetic curves, a study with the largest case series of PASH so far demonstrated persistent (Type I) in 65%, washout (Type III) in 25%, and plateau-type (Type II) enhancement in 6% (9). The mass in this study had a Type I kinetic curve. Diffusion-weighted imaging characteristics and clear diffusion coefficient (ADC) values of PASH have not been defined in the literature so far. In this study, the ADC value was $1{,}327 \times 10^{-3}\ mm^2/s$, which corresponded to benign lesions. This value may

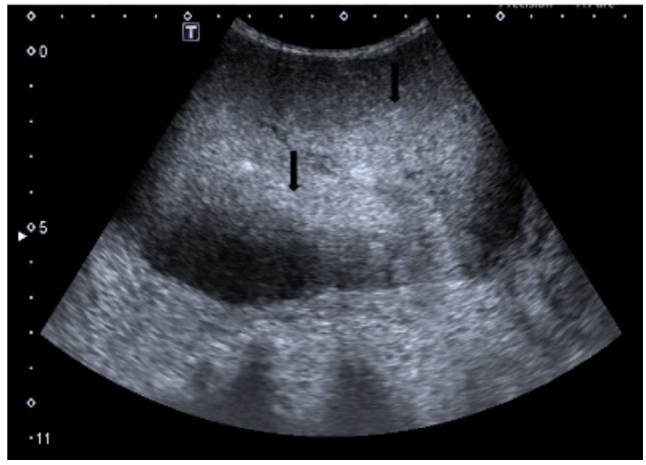

Figure 1. Ultrasonography shows lace like hyperechoic areas inside the mass lesion (black arrows)

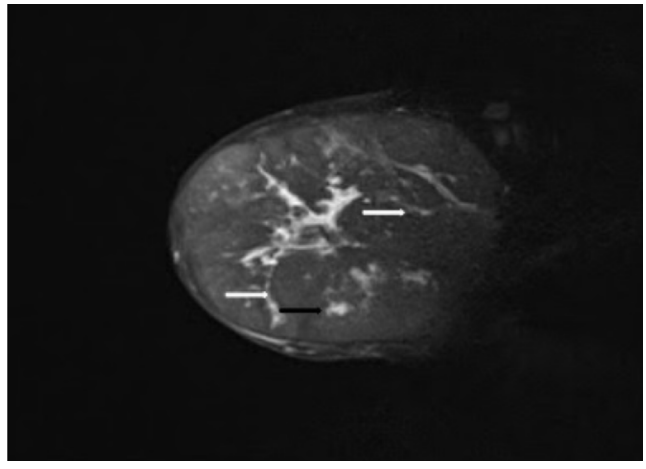

Figure 2. Sagittal T2WI of the right breast shows an abundant hyperintense linear lacelike network (white arrows) and cystic spaces (black arrow)

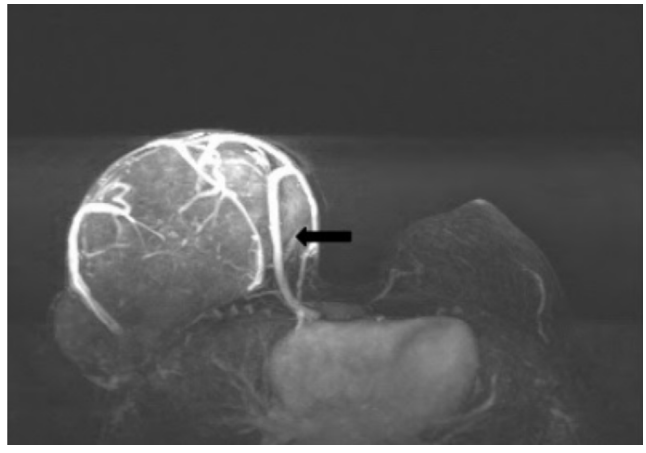

Figure 3. Axial MIP reveals a feeding vessel sign (black arrow)
MIP: Maximum intensity projection

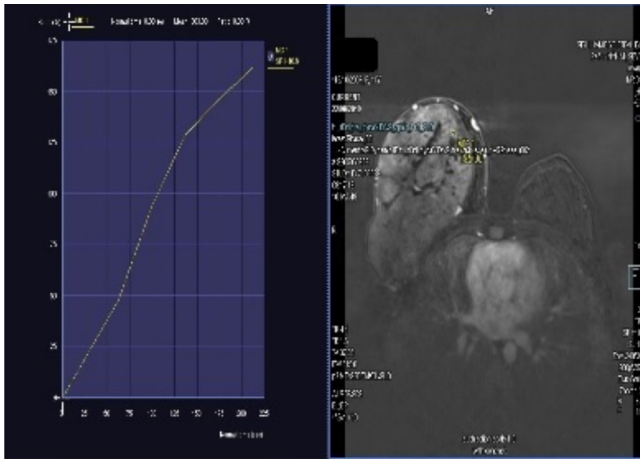

Figure 4. The kinetic curve shows persistent contrast enhancement

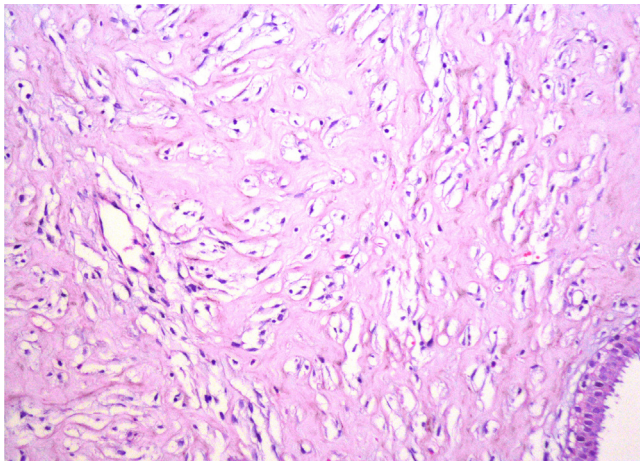

Figure 5. Anastomosing endothelial vascular like structures (pseudovascular spaces) scattered in hyalinised stroma (haematoxylin and eosin, magnification ×100)

exclude phyllodes tumors, angiosarcomas, or malignant mesenchymal tumors. Hamartomas and fibroadenomas can be included in the differential diagnosis. However, hamartomas typically contain intralesional fat and fibroadenomas have T2 hypointense septations inside.

This case highlights the radiological findings of a very rare and complex entity in the pediatric age group. Radiologists should be aware of the imaging findings of PASH. The differential diagnosis of PASH and angiosarcoma is crucial as these two lesions have similarities in clinical presentation, especially in giant sizes.

Ethics

Informed Consent: Informed consent was obtained verbally from the family of the child.

Authorship Contributions

Concept: B.D., B.A.U., Design: B.D., B.A.U., Data Collection or Processing: B.D., B.A.U., Analysis or Interpretation: B.D., B.A.U., Literature Research: B.A.U., Writing: B.A.U.

Conflict of Interest: No conflict of interest was declared by the authors.

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