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Caso Clínico / Radiological Case Report

PSEUDOANGIOMATOUS STROMAL HYPERPLASIA IN PEDIATRIC AGE: A CASE REPORT AND REVIEW OF LITERATURE

HIPERPLASIA ESTROMAL PSEUDOANGIOMATOSA EM IDADE PEDIÁTRICA: CASO CLINICO E REVISÃO DA LITERATURA

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign disease, characterized by abnormal proliferation of fibroglandular stroma. It was first described in 1986. The authors present a case of a twelve year-old girl with a history of kidney transplantation due to nephrotic syndrome with rapidly progressive and painful breast asymmetry with approximately six months duration. No lymphadenopathy or other signs or symptoms were associated. Ultrasound didn't reveal specific findings. Breast magnetic resonance (MR) showed a massive heterogeneous nodular mass with regular contours and contrast enhancement. Given the degree of breast asymmetry as well as the patient's symptoms, surgical excision of the tumor was preferred over core biopsy. Histopathological and immunohistochemical examination showed pseudoangiomatous stromal hyperplasia. The authors describe the clinical presentation, imaging and histological features as well as therapeutic approach in these patients

Key-words

Pseudoangiomatous stromal hyperplasia; Pediatric; Radiology.

Resumo

A hiperplasia estromal pseudoangiomatosa é uma patologia mamária benigna rara, caracterizada pela proliferação anormal do estroma mamário, descrita pela primeira vez em 1986. Os autores descrevem o caso de uma adolescente de 12 anos, transplantada renal por síndrome nefrótico, que apresentava uma volumosa assimetria mamária, rapidamente progressiva e dolorosa, com aproximadamente 6 meses de evolução. Sem adenopatias ou outros sinais ou sintomas associados. O estudo ecográfico foi inespecífico e a ressonância magnética mamária revelou volumosa formação nodular heterogénea, captante e de contornos regulares. Dado o grau da assimetria mamária, assim como a sintomatologia da examinada não foi realizada core biopsia, tendo-se optado pela cirurgia com excisão do tumor. O exame histopatológico e imuno-histoquímico mostraram tratar-se de hiperplasia estromal pseudoangiomatosa. Os autores descrevem a apresentação clínica, imagiológica e histológica assim como a orientação terapêutica nestes pacientes.

Palavras-chave

Hiperplasia estromal pseudoangiomatosa; Criança; Radiologia.

Case Report

A twelve year-old girl with a previous history of renal transplant due to nephrotic syndrome, presented at our institution with breast pain. At physical examination marked breast asymmetry was noticed, with diffuse enlargement of the right breast presenting with hyperemia and tenderness (fig. 1).

The patient underwent breast ultrasound that did not reveal specific findings (fig. 2). On US the breast parenchyma showed diffuse inhomogeneity and no distinct focal lesions were found. Ultrasound evaluation was limited due to the large volume of the breast, allowing only the most superficial areas of the breast to be reached so BI-RADs classification 0 was attributed.

Due to the incongruity between sonographic findings, patient´s symptoms and ultrasound limitations MR was done. MR showed a large heterogeneous tumor with areas of hyperintensity in T2-weigthed sequences and hypointensity in T1-weigthed sequences (fig. 3). The tumor enhancement was progressive but avid and heterogeneous (fig. 4). These imaging finding were classified as BI-RADS 4a. The patient was very symptomatic and the mass continued to grow so it was decided to perform surgery with tumorectomy leading to the histological diagnosis of PASH (fig. 5).

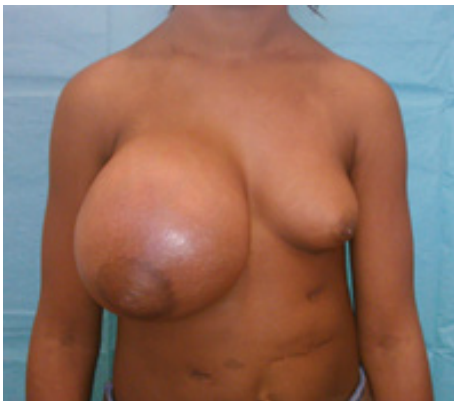

Figure 1 – The right breast is markedly enlarged with overlying skin stretched out and reactive hyperemia.

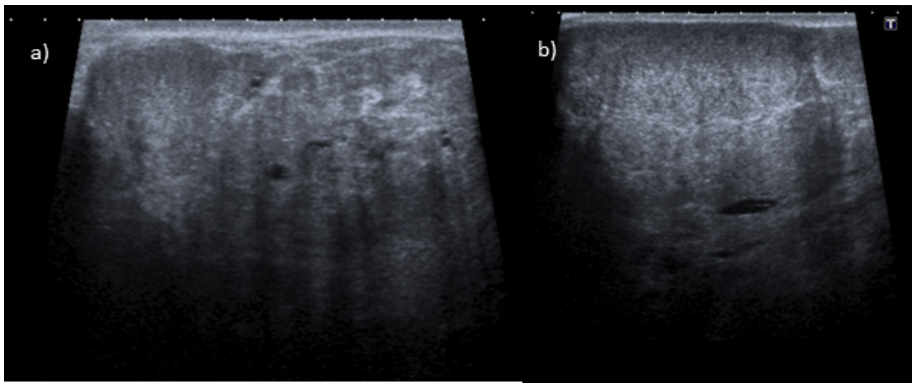

Figure 2 – Ultrassonography images (a and b) showing a diffuse increase of the fibroglandular stroma but no evidence of a defined tumor was found.

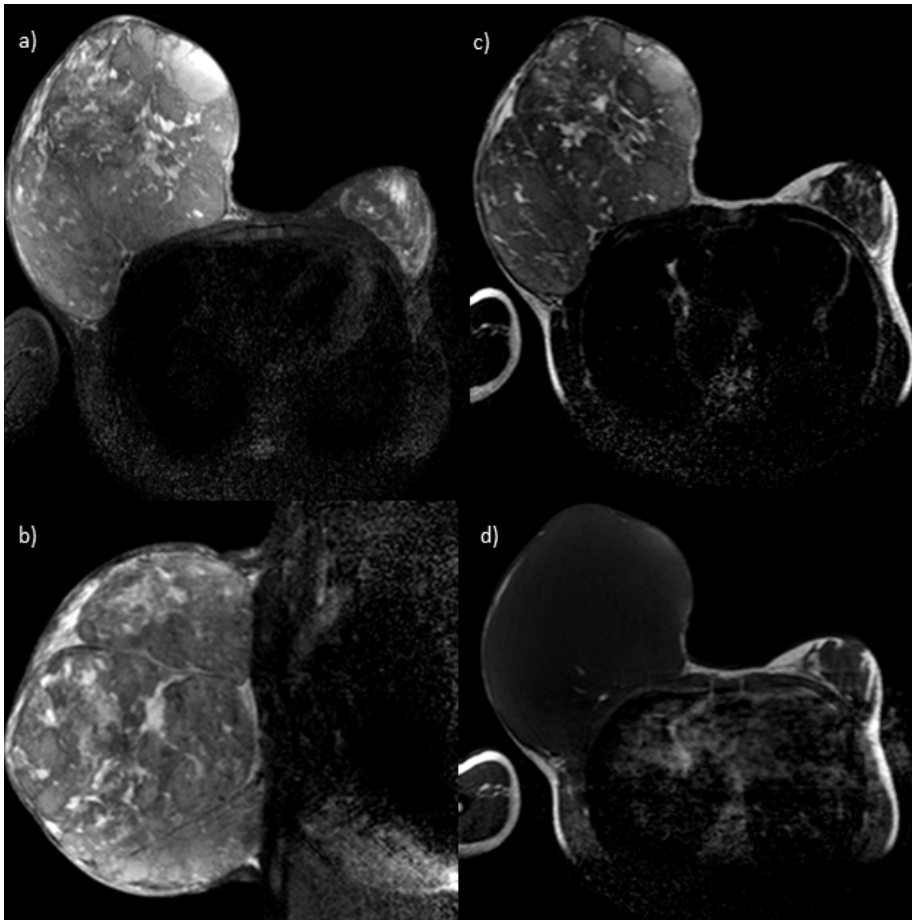

Figure 3 – a) and b) Fat supressed T2 weighted sequences in axial and sagital views, respectively, demonstrating a heterogeneous tumor with regular and lobulated contours and hyperintense focus inside. c) and d) The mass is hyperintense and heterogeous in T2-weighted sequences and hypointense in T1-weighted sequences.

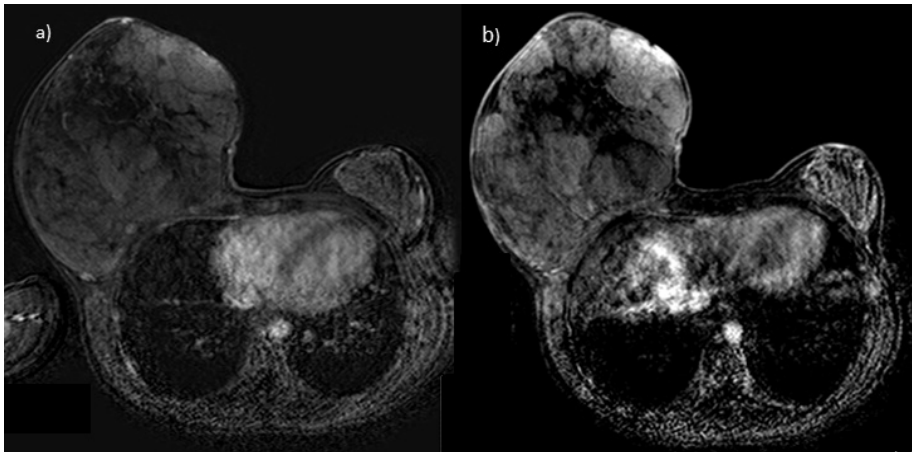

Figure 4 – Post-contrast breast MR (a) – early phase – and b) late phase, showing progressive and heterogenous enhancement of the tumor (right breast).

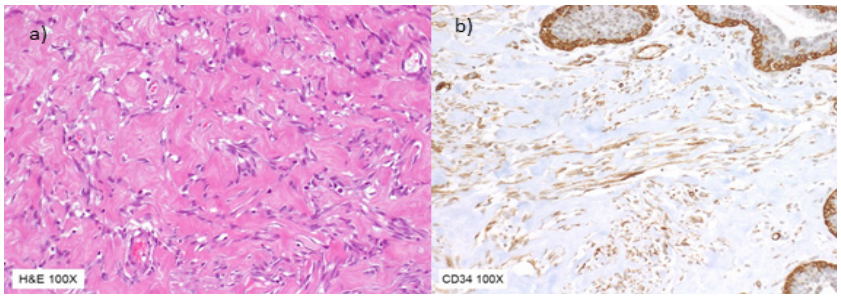

Figure 5 – a) - HE, 100X - Histopathologic examination showed a myofibroblastic proliferation intermixed with epithelial elements. The stromal cells form a complex pattern of empty, often interanastomosing spaces in a densely collagenous stroma. No necrosis, mitotic figures, atypia or destruction of normal breast tissue were identified. The histologic aspects are typical of PASH. b) Immunohistochemistry for CD34, 100X - Immunohistochemical study shows CD34-positive myofibroblasts lining the slit-like spaces.

Discussion

Breast masses are uncommon in the pediatric age and most of them are benign. PASH is a benign proliferation of breast stromal cells, classified as a mesenchymal tumor. This entity was first described in 1986 by Vuitch et al. It is characterized by dense myofibroblastic proliferation of breast stroma associated with interanastomosing capillary-like spaces, hence the name "pseudoangiomatous" and the need of differentiation from angiosarcoma[1,2].

Most authors believe that this pathology may be caused by an exaggerated response of estrogen breast receptors to progesterone stimulation[3]. Thus it is most commonly seen in premenopausal women or women receiving hormonal therapy, and rarely male or elderly females not undergoing hormonal therapy[4].

PASH can be divided in three types varying from insignificant incidental microscopic changes to focal mass-like nodules and diffuse involvement. Focal areas of histological changes typical of PASH are commonly found in biopsy and mastectomy specimens, but tumors composed solely or predominantly of PASH as in our case are rare[4]. Such cases in children are usually described isolated in literature, and making its true incidence in the general population difficult to ascertain without further studies.

The prevalence of breast cancer in the pediatric age is extremely low compared with that in the adult population, thus a conservative approach of clinical and sonographic follow-up is more commonly adopted in children[4,5].

The preferable initial breast imaging study performed in children is ultrasonography (US), with mammography being reserved for selected cases. Mammography plays a role in the evaluation of microcalcifications and in suspicious masses in older adolescents. US has several advantages over mammography such as the lack of ionizing radiation and greater sensitivity in dense fibroglandular tissue of young girls. There is still little experience with magnetic resonance (MR) although some authors describe the increasing MR value in preoperatory evaluation[5,6].

At US, tumor-like PASH is most often solid and hypoechoic, oval in shape, and oriented parallel to the chest wall with or without posterior acoustic enhancement. Its appearance resembles that of a fibroadenoma, its main differential diagnosis. It can be multiple and in a minority of cases small anechoic spaces may be evident[5].

MR characteristics of PASH in T1-weighted and T2-weighted images signal vary widely. The lesions usually present inhomogeneous hiperintensity in T2-weighted images and isointensity to the surrounding parenchyma on T1-weighted images. Persistent, progressive and avid contrast enhancement may be present due to its vascular component, as seen in our case[6,7].

Although these tumors are benign, surgery is indicated for symptomatic or growing masses. Some lesions diagnosed with image-guided core needle biopsy have shown clinical and radiologic stability and cases of spontaneous regression have been described. Recurrence has been found in up to 26% therefore close imaging follow-up is recommended[6]. Our patient underwent surgery and has been asymptomatic without signs of recurrence since.

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