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Case Report

Giant Pseudoangiomatous Hyperplasia of the Breast Stroma: About A Complicated Case of Gigantomastia

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Abstract

Pseudoangiomatous stromal hyperplasia (PASH) is a lesion that is often discovered incidentally in breast biopsies performed for benign or malignant lesions of the breast. More rarely, it may present as a palpable lesion. The term "Pseudoangiomatous" has been proposed to underline its particular histological aspect simulating a vascular proliferation. We report a case of PASH in a 42-year-old female patient presenting as a recurrent lesion that resulted in a considerable and rapid increase in breast volume (135 x 100 x 97mm) requiring a large lumpectomy with muscular-fat remodeling. Angiosarcoma and fibroadenoma remain the main differential diagnoses. Anatomopathological examination allows us to eliminate them, in particular through immunohistochemistry.

Keywords: breast biopsies, Pseudoangiomatous stromal hyperplasia (PASH), Pseudoangiomatous, proliferation.

BACKGROUND

Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign breast lesion, first described in 1968 by Vuich *et al*., [1]. It is defined as a mesenchymal proliferation of breast myofibroblasts characterized by anastomotic clefts lined with flattened cells giving a pseudovascular appearance posing diagnostic problems, which may be misdiagnosed as a low-grade angiosarcoma or phyllodes tumor [2]. PASH has a wide range of clinical presentation, from an incidental microscopic finding accompanying other breast lesions to a rapidly growing palpable mass [3]. It is found primarily in perimenopausal women [4].

The optimal treatment of PASH tumor is still controversial due to its rarity.

CASE PRESENTATION

The patient was 42 years old, with a gynecological history of a phyllodes tumor of the right breast treated by lumpectomy 4 years ago, G4P4 4 living children.

The onset of the symptomatology dates back to 04 months with the fortuitous discovery at autopalpation of a breast mass located in the right superior-internal quadrant, the evolution was marked by the rapid increase in volume of the mass (Figure 1).

The patient underwent a mammography with breast ultrasound, the mammography showed a retroareolar opacity without microcalcifications (Figure 2), the ultrasound showed an oval mass with well limited contours with a double cystic component (without partitions or vegetation's) and fleshy vascularized in places in the center and periphery measuring 135x100x97 (Figure 3 and 4).

A microbiopsy was performed in favor of a Pseudoangiomatous stromal hyperplasia.

We performed a lumpectomy with muscular-fat remodeling. The postoperative aesthetic result was excellent. The postoperative course was unremarkable (Figure 6).

Pathological examination of the surgical specimen (Figure 5) confirmed the diagnosis of pseudoangiomatous hyperplasia of the breast stroma.

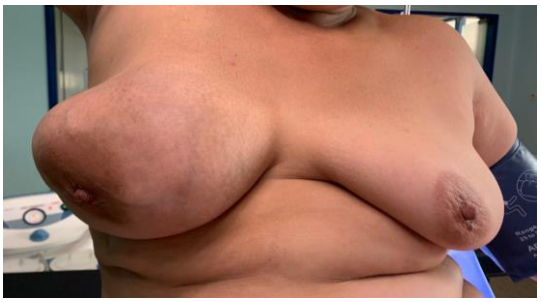

Figure 1: Giant mass of the right breast

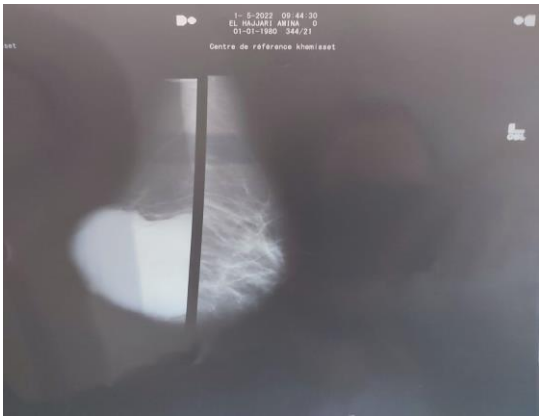

Figure 2: Isolated opacity without microcalcifications

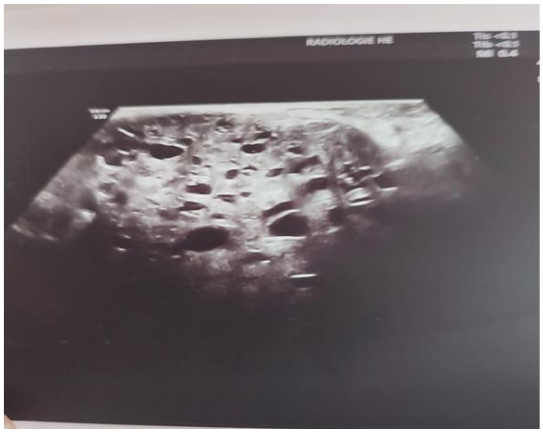

Figure 3: Image with a mixed tissue and fluid component

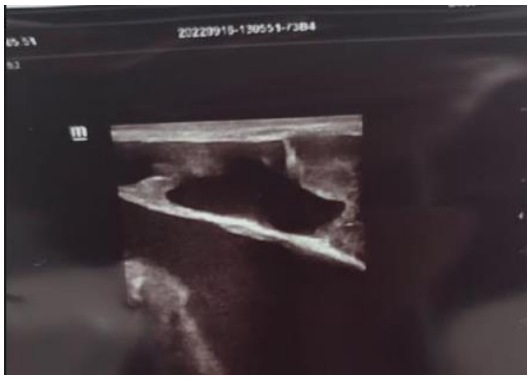

Figure 4: Associated cystic

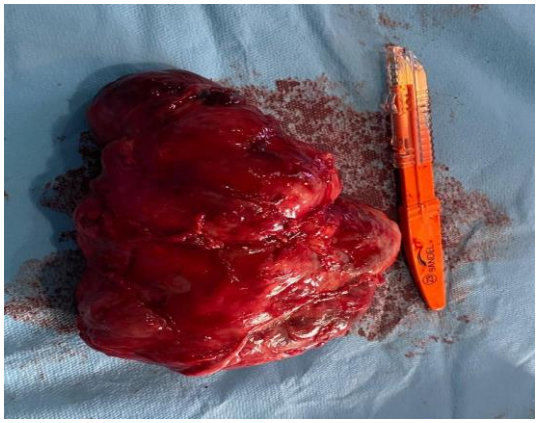

Figure 5: The surgical specimen

DISCUSSION

Pseudoangiomatous stromal hyperplasia is a rare benign breast tumor that is rarely reported in the medical literature with unspecified impact in the literature. It was first described by Vuitch, Erlandson and Rosen in 1986 [1]. They reported a series of nine cases of histologically quite similar breast tumors characterized by a network of anastomotic clefts lined with flattened cells of endothelial appearance, simulating vascular clefts. These empty spaces, devoid of red blood cells, are arranged in a characteristic densely collagenous stroma and may take on a concentric arrangement around the lobules.

This pathology is characterized by its great clinical diversity, ranging from a simple incidental finding on histological examination for non-tumoral forms to gigantomastia, Ibrahim et al subsequently demonstrated that PASH is a relatively frequent but often unrecognized lesion [5]. They observed aspects of PASH in 46 cases among 200 unselected biopsy and mastectomy cases recorded consecutively in their routine recruitment, wich is 23% of the cases studied. They reported cases of PASH in patients with normal physical breast examination, when at age it is mostly observed in perimenopausal women, but a case of giant tumor (35 cm) at the age of 13 years was reported by Abdelrahman *et al*., [6]. Also Almohawes reported a giant form at the age of 12 years [2], our patient was 42 years old and presented with a giant mass of 12 cm. This tumor resembled a low-grade angiosarcoma on ultrasound; a biopsy was performed which revealed a PASH, and surgical excision was performed.

The main differential diagnosis is angiosarcoma, the histological examination allows to make the diagnosis, Angiosarcoma often presents a more aggressive infiltration in the surrounding fibro-adipocytes breast tissue and is highly vascularized, lined with endothelial cells. It does not have a collagenous stroma. Angiosarcoma shows positive immunoreactivity for CD31 [7].

Radiological signs have been repeatedly reported to be non specific in differentiating PASH, in the study by Raj *et al*., and in some cases were similar to fibroadenoma [8].

PASH is more prevalent and more likely to occur in hormonally active women [4, 8].

The treatment of PASH tumor depends on the size and growth rate of it. Surgical and non-surgical treatments (anti-hormonal therapy can be a non-invasive treatment alternative [4] have been both successfully applied in previous studies. Management with wide local excision is the treatment of choice for PASH owing to its uncertain natural history [9].

CONCLUSION

PASH is a benign entity characterized by a myofibroblastic proliferation of the breast, Gigantomastia caused by PASH is rare, the lesion reported in this case is one of the largest in the literature. 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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