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Pseudoangiomatous Stromal Hyperplasia of the Breast: A Case Report of a 32-year-old Woman

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ABSTRACT

Pseudoangiomatous Stromal Hyperplasia (PASH) is a benign breast tumor with a histological pattern that mimics the appearance of angiomatous proliferation. The lesion appears as a complex tissue with slit-like spaces lined by spindle cells resembling endothelial cells with a background of stromal hyperplasia. PASH manifests as a palpable mass, multifocal nodules, or a large diffuse mass that makes the breast asymmetrical. We report a case of breast lump in a 32-year-old woman caused by PASH. Biopsy of the mass showed histopathologic features of PASH. This patient then received tumor excision to remove the mass. Treatment of PASH tumors depends on the size and growth rate of the tumor. Anti-hormonal therapy such as tamoxifen may be an option for non-invasive therapy, but after the diagnosis has been confirmed through a core biopsy and the tumor size is no more than 2 cm. Surgical excision of the tumor by minimizing damage to the surrounding breast tissue is the most recommended treatment.

Keywords: pseudoangiomatous stromal hyperplasia; breast tumor; benign tumor

INTRODUCTION

Pseudoangiomatous Stromal Hyperplasia (PASH) is a rare condition that was first reported in 1986 by Vuitch et al. The term pseudoangiomatous refers to a histologic pattern that mimics the appearance of angiomatous proliferation [1]. PASH is a benign breast tumor that can develop at the age of 14 - 67 years with the majority of patients aged 30 - 40 years [2]. The exact cause of PASH is unknown, but it is hypothesized to be due to hormonal factors. This condition clinically manifests as a palpable mass, multifocal nodules, or a large diffuse mass that makes the breast asymmetrical. Ultrasound findings often resemble fibroadenoma because of the appearance of an oval, hypoechoic mass with regular margins. Histologically, PASH consists of a complex network of silt-like spaces lined by spindle cell endothelial-like with a background of stromal hyperplasia. This condition is similar to low-grade angiosarcoma and phyllodes tumor [3]. The treatment is usually surgical excision, although in some cases, "watch and wait" can be applied depending on the size of the tumor [4].

We report a case of breast lump in a 32-year-old woman caused by PASH. This patient then received tumor excision and there was no recurrence until this study was made.

CASE REPORT

A 32-year-old woman, complaining of a lump in her left breast, that she has been experiencing for three years. The lump appears to be growing in size, not painful and feels hard. The patient is married and has three children who are normally breastfed. She denied having similar complaints before or a family history of the disease.

On physical examination, the general condition was good, compos mentis, and vital signs normal. Breast examination showed the right breast within normal limits. On the left breast, a palpable mass was discovered measuring approximately 6 x 5 cm with a hard consistency, flat surface, well-circumscribed, and painless. On the local status of the left axilla region, enlarged lymph nodes are palpable, supple, fixed, and nontender. Ultrasound examination revealed an abnormality in the left breast: a solid hypoechoic nodule that was well demarcated, inhomogeneous with hyperechoic sections, accompanied by an increased vascular pattern, at 12 o'clock to the retro-nipple of the left breast, measuring 7.51 x 3.4 cm, which leads to an angiofibrolipoma. While in the left axilla, there were enlarged lymph nodes that were well-defined, oval, and hyperechoic in the central hilum, measuring 10.6-11.4 mm, giving the impression of multiple lymphadenopathies.

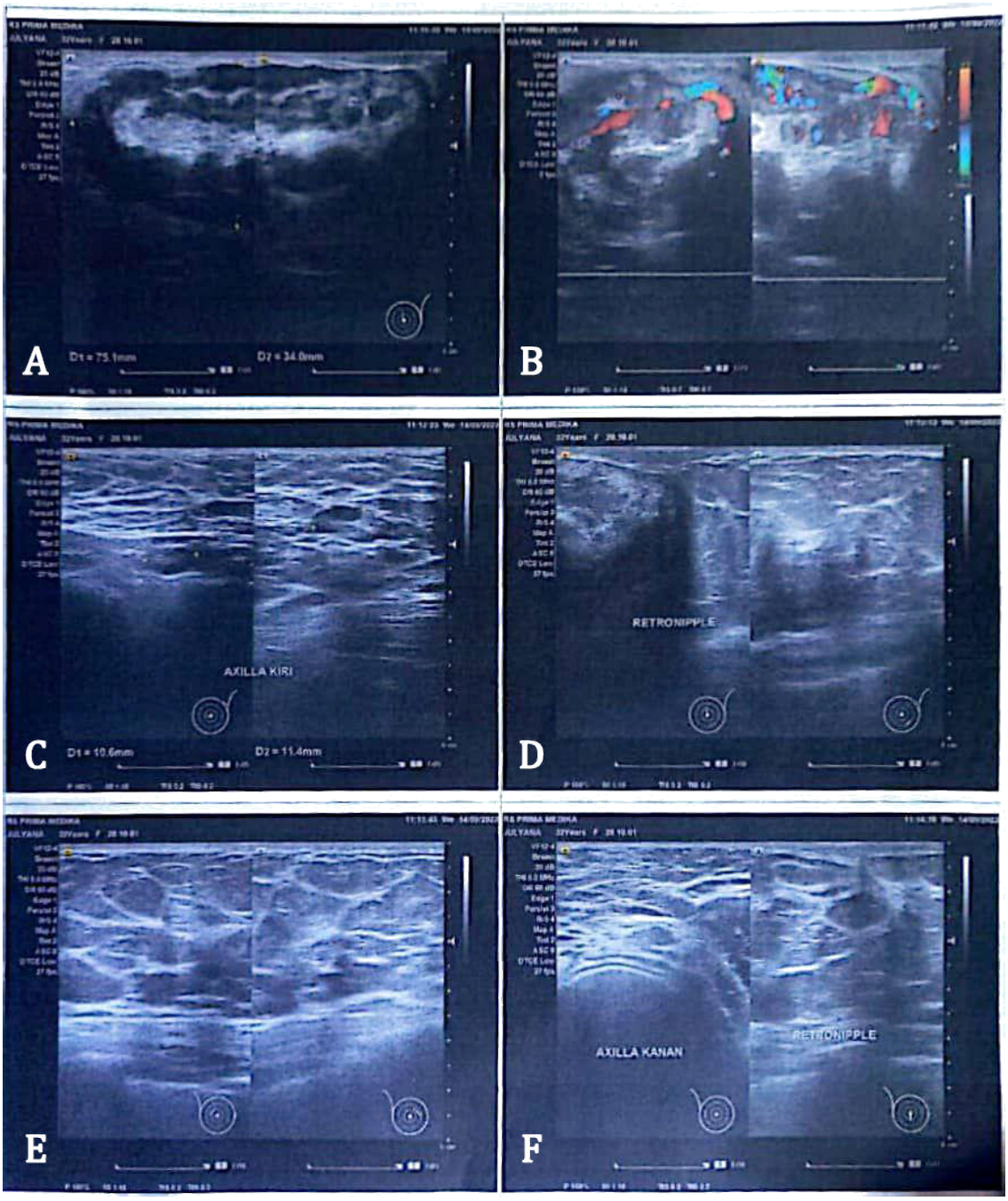

FIGURE 1: Ultrasound examination revealed in the retronipple region (D) there is a well-circumscribed (A), inhomogeneous, hypoechoic solid nodule with hyperechoic patches (E), with an increased vascular pattern (B). In the left axilla, lymph node enlargement, well defined, oval, hyperechoic hilar center was found (C). The right axilla is normal (F).

A core needle biopsy examination was then performed. Histopathological results provide a distorted picture of the terminal duct lobular units (TDLU), which consisted of stromal and ductuli components. Ductuli showed intracanalicular, pericanalicular, some part had cystic dilatation. The stroma is made up of fibromyxomatous tissue with myofibroblastic cell hyperplasia that formed empty spaces between the hyaline stroma. Based on the morphological depiction, it gives the impression of pseudoangiomatous stromal hyperplasia. These findings were similar to the final histopathological result after tumor excision. Surgical excision is performed with an inferior circumareolar incision. The mass is excised through a small incision, then simple glandular remodeling is performed to get a good cosmetic result on the breast. The pathology results later confirmed the diagnosis of PASH.

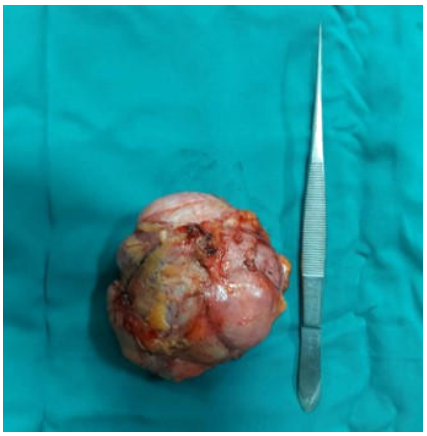

FIGURE 2: Tumor mass after excision.

Then, a follow-up was carried out within 6 months after surgery, and the patient had no complaints. On ultrasound examination, there were no nodules, tissue distortions, or calcifications in the right and left breast. In the axilla, no enlargement of the right or left axillary lymph nodes was found. The patient was also screened for mammography in both breasts; the cutis and subcutis were normal, and there was no microcalcification, lactiferous duct dilatation, or lymph node enlargement.

DISCUSSION

Pseudoangiomatous stromal hyperplasia is a rare benign tumor that is usually found in premenopausal women [5]. PASH can present with a wide clinical spectrum, from incidental histological findings to clinically palpable breast masses [3]. PASH is a slow-growing breast tumor [6]. We also found this in our patient with a slow-growing tumor in the last three years. On physical examination, PASH is a solitary, hard, painless, well-circumscribed, and mobile tumor that mimics a fibroadenoma [6]. However, in some cases, the tumor can grow quickly or diffusely, giving the appearance of malignancy. On this patient we discovered solitary mass, hard, well-circumscribed, and nontender. Thus, giving an initial impression as a benign breast tumor.

Radiologic findings have reportedly been found to be non-specific in identifying PASH on numerous cases [7]. On mammography, usually a clear, dense, homogeneous and non-calcified mass is found. From ultrasound often shows a solid hypoechoic mass with or without cystic spaces and well-circumscribed [8]. Rarely, a diffuse and hyperechoic mass can also be found. In our case, ultrasound showed a well-circumscribed inhomogeneous solid hypoechoic nodule with some hyperechoic parts. In retronipple area of the breast also found an increased vascular pattern, which gave the impression of angiofibolipoma. Because the radiological examination result is not specific, a histopathological examination was performed.

Macroscopically, PASH tumor is round to oval, well-circumscribed with smooth and unencapsulated surfaces. A homogenous solid lesion with a gray-white color and infrequently, cysts is visible on the incision site [6]. PASH can appear as a single mass or coexisting with any breast lesion such as fibroadenoma, phyllodes tumor, and even breast carcinoma. From histological examination, the lesion appears as a complex tissue with slit-like spaces lined by spindle cells resembling endothelial cells with a background of stromal hyperplasia that mimics blood vessels [9]. These slit-like spaces are seen in intralobular as well as interlobular stroma. Because these slit-like spaces resemble blood vessels, they are called “angiomatous” [5]. However, they are lined by myofibroblasts rather than endothelial cells, which distinguish PASH from low-grade angiosarcoma. Immunohistochemistry was positive for CD34 and SMA, while negative for CD31 and ERG, which confirmed that these endothelial-like cells were actually myofibroblasts rather than true endothelial cells [2,5,10]. On our case, from the initial core biopsy, the stromal component was found to be composed of fibromyxomatous tissue with hyperplasia of myofibroblastic cells that formed empty spaces between the hyaline stroma. This is suitable with the histopathological characteristics of PASH which helps to establish the diagnosis. At the final pathological examination after the excision, a dilated ductuli filled with cysts was also found. Then there was no atypia or increased mitotic activity in the stroma, which ruled out the differential diagnosis of a breast malignancy.

The exact etiology and pathogenesis of PASH are still unknown. According to many studies, hormonal factors are the most influential factor in the development of PASH [11].

Stromal cell nuclei of PASH express progesterone receptor with high density. Whereas normally, the mammary stroma does not show progesterone expression on staining [4]. Currently the most widely accepted hypothesis in the literature is that stromal hyperplasia results from an exaggerated and aberrant mammary myofibroblast response to hormonal stimuli [5]. This hypothesis is also supported by the majority of patients who are premenopausal women, or postmenopausal women who receive hormonal replacement therapy [12]. Some reports also mention a man with gynecomastia and a male transgender woman experienced PASH during hormone therapy [11,13]. Unfortunately in our case, the patient's progesterone or estrogen receptor levels were not tested. Further studies are needed to support the hypothesis of this hormone-induced etiopathogenesis.

Treatment of PASH tumors depends on the size and growth rate of the tumor. Previous research has found that both surgical and non-surgical procedures are effective. Considering the role of hormones as the etiology of PASH, anti-hormonal therapy may be an option for non-invasive therapy such as tamoxifen, but after the diagnosis has been confirmed through a core biopsy and the tumor size is no more than 2 cm [4,7]. Surgical excision of the tumor by minimizing damage to the surrounding breast tissue is the most recommended action in the literature [7]. In our patient, surgical excision of the tumor was performed due to its large size.

After surgical excision, there is a 22% chance of recurrence [10]. There is insufficient evidence whether PASH tumors after excision can develop into a malignancy. In a study conducted by Degnim et al., PASH patients had a lower rate of malignancy than other benign breast lesions within 18.5 years of their onset [14]. Then, when compared with the general population, PASH tumors do not show an increased risk of breast cancer [7]. In our patient, within 6 months after surgery, and the patient had a follow up examination with no recurrence. On ultrasound, there were no nodules, tissue distortions, or calcifications in the right and left breast. The patient was also screened for mammography in both breasts; the cutis and subcutis were normal, and there was no microcalcification, lactiferous duct dilatation, or lymph node enlargement.

CONCLUSION

Pseudoangiomatous stromal hyperplasia is a rare benign breast tumor with a broad clinical presentation. Radiological examination is not sufficient to diagnose PASH, and histological examination is necessary to obtain a certain diagnosis. Treatment of PASH tumors depends on the size and growth rate of the tumor. If the diagnosis has been confirmed as PASH and the size of the tumor is less than 2 cm, anti-hormonal therapy such as tamoxifen may be an option. If the tumor is more than 2 cm, then surgical excision is recommended.

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