



A Rare Case Presenting with Breast Mass Pseudoangiomatous Stromal Hyperplasia

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Abstract

Pseudoangiomatous Stromal Hyperplasia (PSAH) is a disease resulting from benign proliferation of the breast stroma. It is characterized histomorphologically by fibroblasts and myofibroblasts forming cleavage-like spaces in the mammary stroma. Clinically, it is rarely diagnosed by palpable mass and different diseases such as fibroadenoma, phyllodes tumor, low-grade adenocarcinoma and angiosarcoma may be suspected. Here we present a case of a 45-year-old woman who presented with right-sided pain and mass complaints and was diagnosed as PSAH by surgical excision.

Keywords: Pseudoangiomatous stromal hyperplasia; Breast mass; Benign breast neoplasms

Introduction

Pseudoangiomatous Stromal Hyperplasia (PSAH) is a benign disease of the breast that was first described in 1986 [1]. It is caused by excessive proliferation of the breast stroma. Although the cause of this proliferation is thought to be hormonal stimulation, the definitive etiology and pathogenesis is still unknown. PSAH, which is rarely seen in the cases examined due to mass in the breast, is macroscopically detected as a well defined, pale, homogeneous and fibrous lesion; and as a result of its well demarcation from surrounding parenchyma, it can be confused with fibroadenoma and phyllodes tumor in imaging examinations [2]. Histopathologic correlations confirm the diagnosis. Surgical treatment is in the form of removal of mass with a 2 cm border, in patients with pain complaints and cosmetic concerns. Radiologically and pathologically confirmed small lesions can be followed.

Case Presentation

A 45-year-old female patient was admitted to our clinic with the complaint of pain in the right breast and a palpable mass. It was learned from her medical history that she had pain in the right breast for the last 3 months, and the mass was palpable for the last 1 month. There was no history of previous operation and there was no chronic disease present. The menstrual cycle was normal. Physical examination showed breast asymmetry in the inspection and the right breast was bigger than the left breast. There was no difference in the breast skin and areola compared to the other breast. Hard, mobile smooth, limited mass in the right breast with 5 cm diameter at 11-12 o'clock position was palpated. There were no palpable axillary lymph nodes. By ultrasonography: a fusiform, hypo-isoechoic formation of 50 mm × 43 mm, whose longitudinal axis was extending parallel to the skin, with linear echogenicity and millimetric cystic areas, was observed in the right breast at 11-12 o'clock position, adjacent to the areola. Mammography detected a relatively smooth calcified formation on the upper half-median line at the retroareolar level with approximately 5 cm × 4.5 cm in dimension. No pathological lymph nodes were detected in the axilla (Figure 1). Fine needle aspiration biopsy was reported as negative for malignancy and consistent with fibrosis. Mass was removed surgically due to pain and cosmetic complaints. On pathological examination, a white circumscribed rubbery mass of 5 cm × 4 cm × 2.5 cm was detected. Histopathologically, stromal slit like clefts and rare poorly formed fascicles of CD 34 positive, CD31, ETS-Related Gene (ERG) and cytokeratin negative spindle cells were observed and reported as PSAH. Cells lining the slits and spindle cells were also positive for Smooth Muscle Actin (SMA), desmin, caldesmon ve Estrogen Receptor (ER) (Figures 2-4). Postoperative recurrence was not observed in 3 and 6-month controls.

Discussion

PSAH, one of the benign proliferative breast diseases, was described in 1986 by Vuitch et al.

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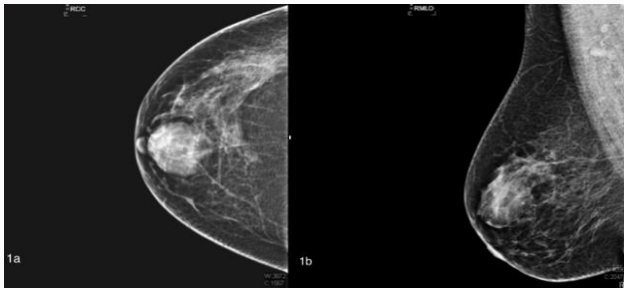


Figure 1: The Axilla.

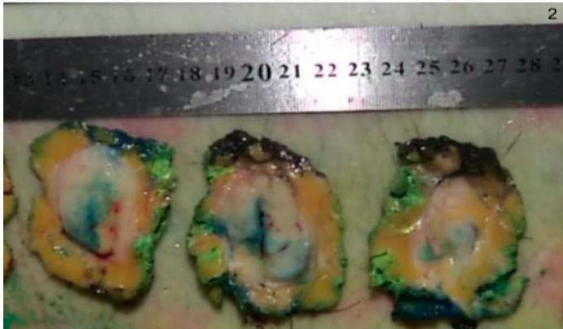


Figure 2: A white circumscribed rubbery mass of 5 cm x 4 cm x 2.5 cm.

[1]. It is characterized by dense proliferation in which breast stroma, fibroblasts, and myofibroblasts form fissura-like gaps [3]. Hormonal factors are thought to be involved in its etiology and formed as a result of an excessive response to estrogen and progesterone. PSAH shows positivity for estrogen, progesterone, androgen gene receptors; CD34, and vimentin. Cytokeratin and vascular markers such as CD31 and factor 8 are negative [4].

It was incidentally reported to be detected at the rate between 0.4% and 23% in breast biopsies by histopathologists. Clinical finding is a single or multifocal, palpable, regular, rigid mobile mass. Because of these properties, it is confused with fibroadenoma in physical examination [5].

PSAH is more common in women of young reproductive age. The observed age range is 14 to 67, and the frequency is mostly the beginning of the thirties and the end of the forties. It may be observed in postmenopausal women as unrelated to hormone therapy.

Findings in imaging studies are not specific. Sonographic findings are generally similar to fibroadenoma and phyllodes tumors, i.e. they are seen as heterogeneous masses with round, hypoechoic, uniformly restricted or having cystic areas. Nevertheless, less frequently, the sonographic findings of some lesions are similar to those of malignant lesions such as heterogeneous tissue density, increased echogenicity and irregular confinement.

In mammography, it is seen as a well-defined mass without calcification. Average diameters are between 0.3 and 11 cm. In rare cases, irregularly restricted areas can be observed. It may create focal asymmetry areas. Approximately 22% of pathologically diagnosed patients have no mammographic findings [6].

Definitive diagnosis is made by histopathologic examination. PSAH may be confused with benign and malignant tumors such as fibroadenoma, phyllodes tumor and angiosarcoma [7]. Although stroma is paucicellular in most cases, cellular variant of PASH must

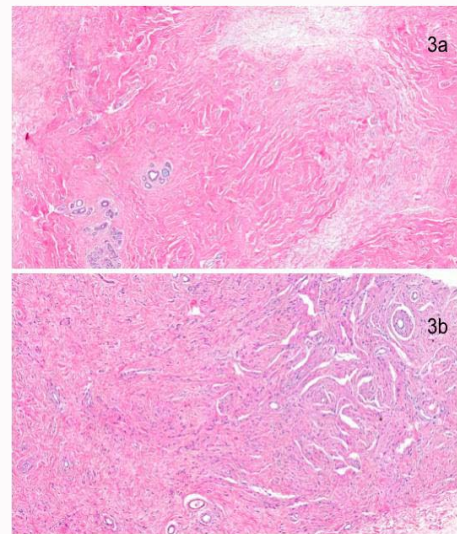


Figure 3: Stromal slit like clefts (3a) and poorly formed fascicles of spindle cells (3b). (3a: HE x 3.3, 3b: HE x 7.5).

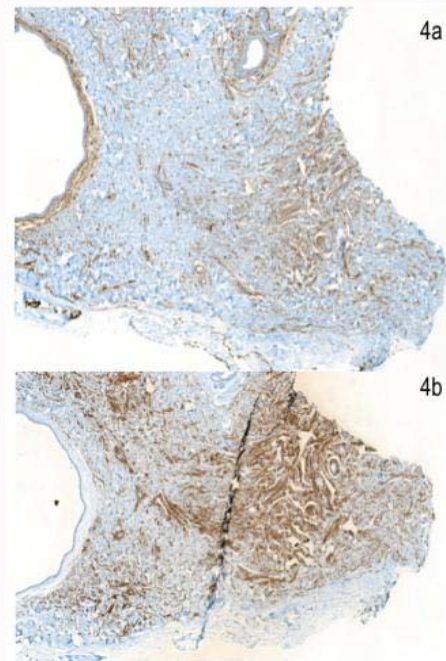


Figure 4: Spindle cells were positive for CD34 (4a) and SMA (4b). (4a: CD34 x 3.4, SMA x 4.4).

be included in the differential diagnosis of all spindle cell lesions of the breast. Coexisting neoplasms (two cases of invasive carcinoma, three cases of non-Hodgkin's lymphoma), although rare, were reported [8]. There is a study in women with PSAH showing no increased risk of breast cancer compared to the normal population [9]. Treatment varies according to the patient's clinic. In patients with pain and cosmetic concerns, treatment is surgical excision. Excision is recommended with a limit of more than 2 cm [10]. Clinical follow-up is possible in pathologically and radiologically benign cases and small lesions.

The recurrence rate after surgery varies between 0% and 22%. One case of bilateral mastectomy due to recurrent lesions was reported. There are insufficient data regarding medical follow-up

and treatment. A bilateral progressive proliferating estrogen receptor positive PSAH case was reported by Pruthi et al., [11] that was treated with tamoxifen for 6 months; and pain and the size of the mass was reported to decrease.

Conclusion

PSAH is a rarely diagnosed benign condition among the patients admitted to our clinic with a complaint of a palpable mass. Pathological confirmation is important because clinical and radiological findings are nonspecific. Treatment in patients with complaints such as chronic pain and cosmetic problems is surgical excision.

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