

Large tumor forming pseudoangiomatous stromal hyperplasia of the breast: a case report

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SUMMARY

Pseudoangiomatous stromal hyperplasia (PASH) is a benign mesenchymal proliferation with numerous slit-like spaces lined by spindle cells. This is not an uncommon finding showing small microscopic foci, but tumor-forming PASH is rare, especially in postmenopausal women. It has to be distinguished from other benign and malignant tumor, especially from low-grade angiosarcoma. The recommended treatment is wide local excision.

Key words: Breast Neoplasms; Breast Diseases; Angiomatosis; Hyperplasia: Diagnosis, Differential

INTRODUCTION

Pseudoangiomatous stromal hyperplasia (PASH) is a benign mesenchymal proliferation with numerous slit-like spaces lined by spindle cells, originally described by Vuitch *et al.* in 1986 (1-5). This is not an uncommon finding showing small microscopic foci, but tumor forming PASH is rare, especially in postmenopausal women (1, 3, 6, 7). The etiology and pathogenesis of this tumor-like lesion remains controversial, but it has been suggested that hormone-related proliferation of myofibroblasts plays a major role in the histogenesis of PASH (3, 5, 7, 8).

CASE REPORT

A 73-years old woman came to medical attention with a painless tumorous lump of the left breast. Physical examination revealed about 80 x 80 mm, round, easily movable mass of upper quadrants, with no palpable axillary lymph nodes. The skin overlying the mass, nipple and areola were normal except for the presence of thin surgical scar in the mass region. This was the fourth time the patient underwent a surgical procedure for a mass lesion in the left breast, the first three being performed outside our institution, but with no official medical record saved. However, the patient stated that the removed lesions proved to be benign. The interval between the two last operations was as long as 17 years. She had no history of hormone replacement therapy. Mammographic finding pointed to a well-circumscribed oval, non-calcified mass. Ultrasonography showed a well-defined hypoechoic mass. Surgical excision was done with pathologic frozen section examination, but the pathologist decided to defer the diagnosis to permanent sections. Gross examination revealed an 80 x 75 x 45 mm, well demarcated, rubbery-hard mass with fibrous grayish-white cut surface. There were a few small cysts filled with a tan fluid. No areas of necrosis and hemorrhage were present. Microscopic examination showed a non-encapsulated tumor mass consisting of anastomosing slit-like empty spaces, lined by spindle cells with bland nuclei. Neither mitotic figures nor nuclear atypia was seen (Figure 1). Immunohistochemically, the spindle cells were negative for CD31 and D2-40, thus indicating that they were not vascular or lymphatic endothelial cells. The cells were positive for vimentin, CD34 (Figure 2), calponin (Figure 3) and weakly for smooth muscle actin, and negative for cytokeratins, estrogen and progesterone receptors. This immunophenotype confirmed the myofibroblastic nature of the cells and the final diagnosis was tumor forming pseudoangiomatous stromal hyperplasia.

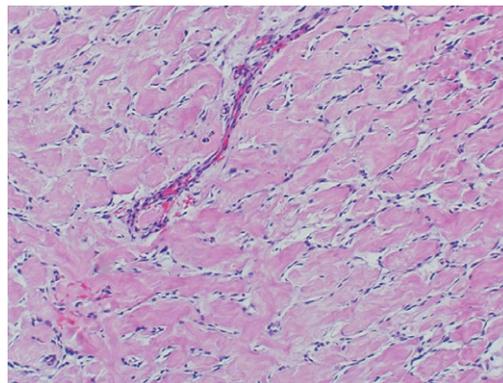


Figure 1. Empty slit-like spaces in PASH lined by cytologically bland spindle cells (H&E, 100x)

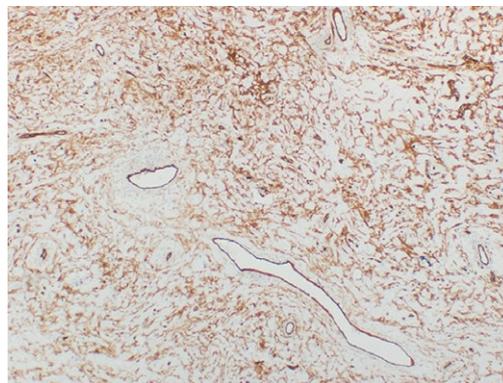


Figure 2. Strong immunoreactivity with CD34 (Immunoperoxidase, 40x)

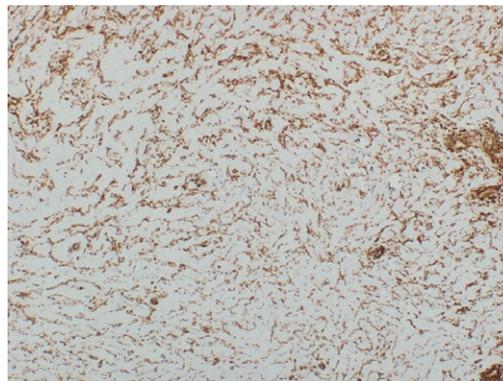


Figure 3. Strong immunoreactivity with calponin (Immunoperoxidase, 40x)

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DISCUSSION

The presentation of PASH ranges from a solitary microscopic finding to diffuse involvement of the breast or as a mammographically and clinically evident mass. Microscopic foci of PASH can be found in association with fibrocystic change, benign or malignant tumors and even with normal breast tissue in about 20% of breast specimens (1, 2, 3, 8). The tumor forming PASH is commonly seen in pre-menopausal women as a single, painless, circumscribed, fibroadenoma-like lesion (2, 3, 8). Herein we report a case of tumorous PASH in postmenopausal women. The etiology and pathogenesis of this tumor-like lesion remains controversial, but many authors have suggested a hormonal cause of this condition. Widely accepted opinion is that aberrant, progesterone-related reactivity of myofibroblasts is the cause of PASH (3, 7, 9). The positivity of progesterone receptor status of spindle cells supports this theory (3, 8), but in our case there was no positivity for estrogen and progesterone receptors. The patient has no history of hormonal replacement therapy. PASH has no specific or diagnostic features on imaging examinations so the diagnosis is based on microscopic examination considering the characteristic histomorphological features supported by immunohistochemical profile (3, 6, 8, 10, 11). Differential diagnosis of tumor-forming PASH includes many benign and malignant tumors. Because of its microscopic features mimicking a vasoformative tumor it has to be differentiated from low-grade angiosarcoma (1, 4, 5, 7). Pleomorphism and hyperchromasia of the stromal cell nuclei are minimal to absent in PASH and the slit-like spaces lack red blood cells in contrast to angiosarcoma. Immunohistochemistry markers can be helpful. Both PASH and angiosarcomas are positive for CD34, but the latter are also immunoreactive with CD31 and Factor VIII, which is not the case with PASH. The other tumors, which should be distinguished from PASH, include fibroadenoma, myofibroblastoma, mammary hamartoma, and phyllodes tumor (2, 3, 4, 12). The recommended treatment is wide local excision (1, 4, 8, 13). The use of tamoxifen in a patient with bilateral PASH, breast enlargement, and pain was reported (14). The prognosis for patient with PASH is good and there are no published cases of distant metastases or death related to PASH (10, 11).

Conflict of Interest

We declare no conflicts of interest.

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