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Pseudoangiomatous Stromal Hyperplasia of the Breast: Multimodality Review With Pathologic Correlation

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Pseudoangiomatous stromal hyperplasia (PASH) is a rare benign breast condition. PASH is thought to be hormonally responsive, and it is usually identified in premenopausal and perimenopausal women. PASH may also be seen in postmenopausal woman on hormone replacement therapy (HRT). Approximately 53% of patients with PASH present with abnormalities on screening mammography, and 44% of patients with PASH present with palpable abnormalities. On imaging studies, PASH appears similar to fibroadenomas. On mammography, PASH is usually seen as a noncalcified, circumscribed mass. On ultrasound, PASH often appears as an oval, circumscribed, hypoechoic mass. On magnetic resonance imaging, PASH usually has progressive (Type 1) enhancement, and high-signal slit-like spaces may be seen on T2-weighted and short tau inversion recovery (STIR) images. The slit-like spaces correspond to empty clefts within acellular hyalinized stroma on histopathology. PASH may be mistaken for a low-grade angiosarcoma on pathologic examination. While angiosarcoma has true vascular spaces, PASH has a network of pseudoangiomatous slit-like clefts. Women with biopsy-proven PASH usually undergo follow-up imaging. Surgical excision may be considered for larger lesions and in women at an increased risk for developing breast cancer. In the future, additional studies are needed to provide definitive data regarding appropriate management and long-term outcomes for women with PASH. PASH has become increasingly recognized, but the literature regarding the imaging features of PASH is scarce. This paper reviews the imaging and pathologic features of PASH and some processes that may simulate PASH are discussed. Features of PASH on mammography, ultrasound, MRI, and nuclear medicine studies are discussed with pathologic correlation.

Introduction

Pseudoangiomatous stromal hyperplasia (PASH) was first described in 1986, and it is an uncommon benign breast condition with a broad spectrum, ranging from an incidental microscopic finding to a palpable mass (Fig 1).[1,2] PASH is usually a focal process, but diffuse disease and multifocal nodules have been described.[3] PASH is known to resemble other lesions, and histopathologic correlation is usually needed to confirm the diagnosis.[4] PASH has been associated with benign and malignant breast lesions in up to 23% of cases.[5] PASH is thought to be hormonally responsive and it is typically seen in premenopausal and perimenopausal women.[6] The clinical management of PASH remains a controversial issue.

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Histopathology

PASH is classified as simple (Fig 2) or fascicular or proliferative (Fig 3), with some series describing a 2:1 predilection for the simple form.[6,7] PASH is typically characterized by a proliferation of fibrous stroma, with fibroblasts and myofibroblasts, and a network of slit-like empty clefts within acellular hyalinized stroma. The simple type displays predominantly anastomosing channels (Fig 2F), whereas the fascicular or proliferative type is characterized by areas of spindle cell proliferation (Fig 3D and E). Typical PASH lesions are progesterone, estrogen, and androgen receptor, CD34, and vimentin positive.[8] PASH lesions are negative for endothelial markers (CD31 and factor VIII) and cytokeratin.[8,9]

Angiosarcoma: Pathologic Mimicker

PASH can be mistaken for low-grade angiosarcoma (Fig 4), which has vascular channels containing erythrocytes.[10] The term pseudoangiomatous highlights the difference between the pseudovascular spaces of PASH (which rarely contain a

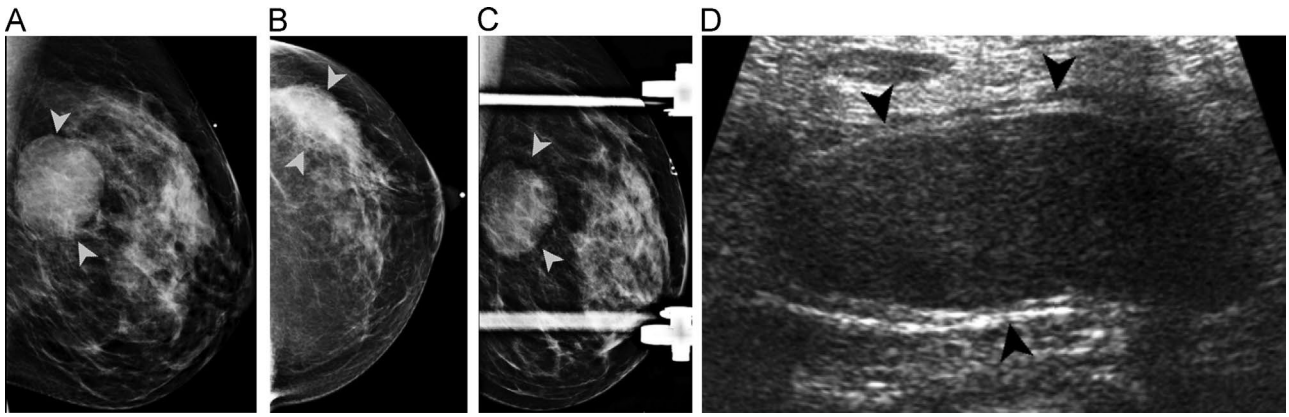

Fig. 1. PASH in a 37-year-old woman with a firm, mobile, palpable left breast mass. (A) Left mediolateral oblique and (B) left craniocaudal (CC) mammograms show a 5 cm noncalcified oval mass (arrowheads) correlating with the palpable abnormality. (C) Left exaggerated CC spot compression mammogram demonstrates circumscribed margins (arrowheads). (D) Ultrasound shows a hypoechoic, oval, well-circumscribed mass (arrowheads).

few erythrocytes) and the true vascular spaces of angiosarcoma (Fig 4C). Pathologic examination should include endothelial markers (CD31 and factor VIII) if an angiosarcoma is suspected, and atypical endothelial cells are noted in angiosarcomas.[5] PASH is not associated with the malignant cytologic features found in angiosarcoma, such as atypia, mitoses, or pleomorphism.

Clinical Manifestations

PASH is a rare entity with a wide spectrum of clinical presentations. A recent study of 57 patients with PASH by Jones et al[4] found that 44% of patients presented with palpable abnormalities.[11] PASH has not been reported with skin or nipple abnormalities. In the study by Jones et al,[4] 53% of patients with PASH presented with

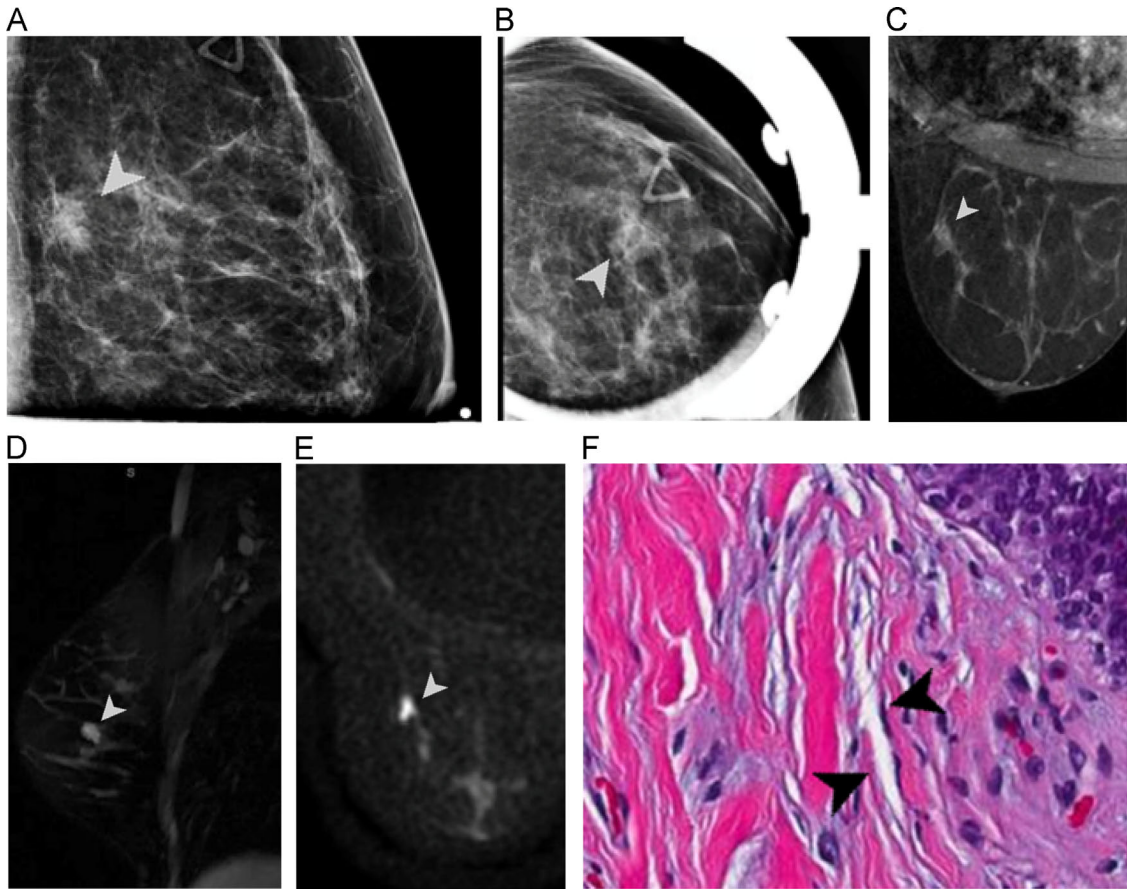

Fig. 2. PASH in a 63-year-old woman with left breast pain associated with a palpable abnormality. (A) Left exaggerated craniocaudal mammogram shows a focal asymmetry (arrowhead) that is seen on the (B) left lateromedial view with spot compression (arrowhead in the region of the palpable marker [triangle]). (C) Axial T1-weighted fat saturated postcontrast MRI shows a lobular mass with heterogeneous enhancement (arrowhead). The mass is hyperintense on (D) sagittal T2-weighted MRI series (arrowhead) and on (E) axial diffusion-weighted MRI series (arrowhead). Photomicrograph (F) shows the simple type of PASH with predominantly anastomosing channels (arrowheads) lined by myofibroblasts (original magnification, ×200; hematoxylin-eosin stain). MRI, magnetic resonance imaging.