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Pseudoangiomatous stromal hyperplasia (PASH) of the breast – report of 2 cases

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Summary

Background:

Pseudoangiomatous stromal hyperplasia (PASH) of the breast is a rare benign proliferation of breast stroma usually diagnosed accidentally during pathological examination of other lesions, but in some cases presenting as a rapidly growing tumor. On clinical examination and imaging it may be mistaken for phyllodes tumor or fibroadenoma and on pathological examination for a low-grade angiosarcoma. The treatment of choice for PASH is surgical removal with wide margins, because of the recurrence possibility. However, the lesion is neither associated with malignancy nor considered as a premalignant one.

Case Report:

A 28-year-old female presented with a large, painless, right breast tumor. In US an extensive, well circumscribed hypoechoic mass with small cysts inside was found. In MR a large, well circumscribed lesion with heterogeneous signal was found, in dynamic examination with enhancement curve type Ib, suggesting benign dysplasia. The second patient, a 22-year-old female presented with a fast growing, small, painless right breast tumor. In US a well circumscribed hypoechoic area behind the nipple was found. In MR no well circumscribed area behind the nipple was found, but scattered lesions, in dynamic examination with enhancement curve type Ib, suggesting benign dysplasia. In both patients on pathological examination of material obtained in US guided mammotomic biopsy PASH was diagnosed. Both patients refused lesion excision, so have been followed-up with US every 6 months and MR every year, with no progression found in the course of 4 years in the first and 6 years in the second patient.

Conclusions:

Tumoral form of PASH is a rare pathology, but it should be known by radiologists, because of features overlapping with other pathological lesions, particularly low-grade angiosarcoma.

Key words:

breast • PASH • magnetic resonance

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Background

Pseudoangiomatous stromal hyperplasia (PASH) of the breast is a rare benign proliferation of breast stroma usually revealed accidentally during pathological examination of other lesions, but in some cases presenting as a rapidly growing tumor [1–3]. On clinical examination and imaging it may be mistaken for phyllodes tumor or fibroadenoma and on pathological examination for a low-grade angiosarcoma [1,2]. The treatment of choice for PASH is surgical removal with wide margins, because of the recurrence

possibility. However, the lesion is neither associated with malignancy nor considered as a premalignant one.

Cases Report

A 28-year-old patient, E-B, presented with a large, painless tumor of the right breast. On US (Figure 1), an extensive (3/4 of the breast), well circumscribed, hypoechoic mass lesion with small cysts inside was found. On MRI (Figure 2–4), a large, well circumscribed lesion with heterogeneous signal was found, in dynamic examination

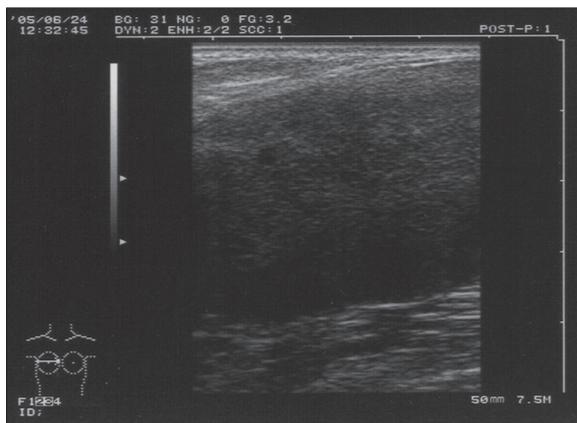


Figure 1. Patient E-B, US of the right breast – large well circumscribed hypoechoic lesion with small cysts.

with enhancement in phase I of up to 145% and enhancement curve type Ib (Figure 3), suggesting benign dysplasia. Histopathological examination of the sample material obtained in US-guided mammotomic biopsy revealed pseudoangiomatous stromal hyperplasia (PASH).

The second patient, a 22-year-old female, A-K, presented with a fast growing, small, painless right breast nodule.

- US showed two independent structures in the right breast:
- in upper, external quadrant: lesion suggestive of fibroadenoma,
 - behind the nipple: quite well circumscribed hypoechoic area of approx. 43×20 mm (Figure 5).

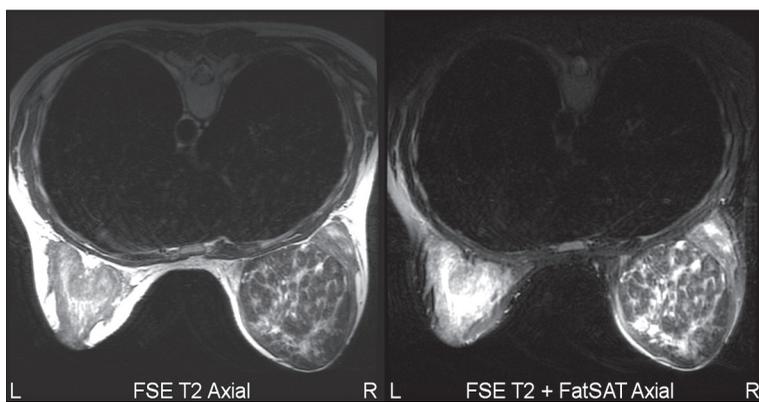


Figure 2. Patient E-B, MR of both breasts, FSE T2-weighted and FSE T2-weighted with fat saturation sequences, axial slices - large well circumscribed tumor of the right breast with inhomogeneous signal.



Figure 3. Patient E-B, MR of both breasts, SE T1-weighted and 3D T1-weighted after i.v. contrast medium administration (Multihance 0.1 ml/kg) dynamic (6 times repeated with 56s interval) sequences, axial slices – large well circumscribed tumor of the right breast with inhomogeneous signal, enhancement curve type Ib in marked point.

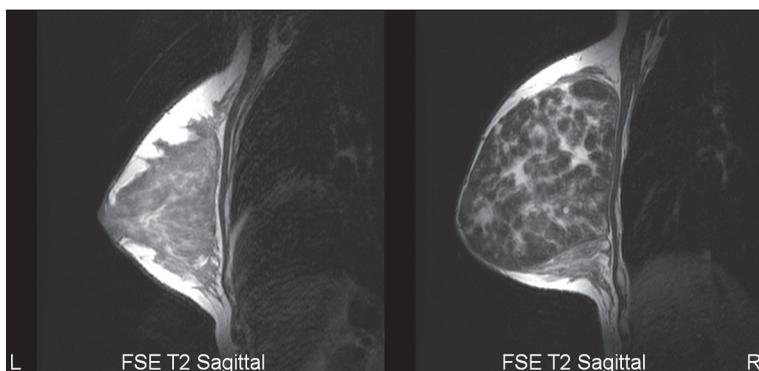


Figure 4. Patient E-B, MR of both breasts, FSE T2-weighted sequence, sagittal slices – large well circumscribed tumor of the right breast with inhomogeneous signal.



Figure 5. Patient A-K, US of the right breast – well circumscribed hypoechoic area behind the nipple.

MR (Figures 6–8) showed:

- in upper external quadrant: lesion suggestive of fibroadenoma,
- behind the nipple: no well circumscribed area, as on USG, but only scattered lesions, in dynamic examination with enhancement in phase I of up to 109% and enhancement curve type Ib (Figure 7), suggesting benign dysplasia.

Pathological examination:

- the excised nodular lesion of the upper, external quadrant confirmed fibroadenoma nipple,
- the material sampled from the lesion located behind, obtained in the course of the US-guided mammotomic biopsy, revealed pseudoangiomatous stromal hyperplasia (PASH).

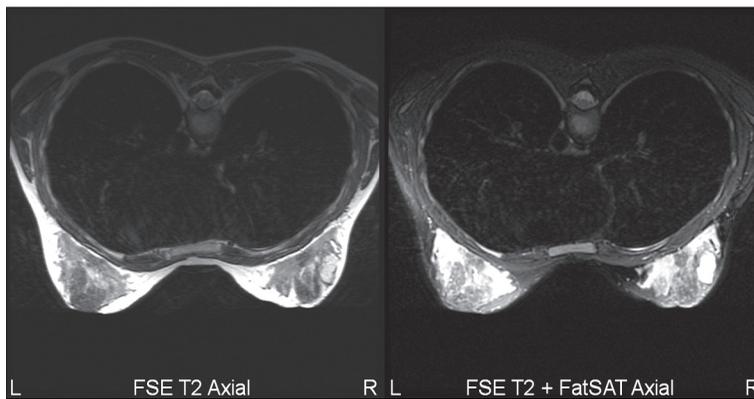


Figure 6. Patient A-K, MR of both breasts, FSE T2-weighted and FSE T2-weighted with fat saturation sequences, axial slices – fibroadenoma in the upper external quadrant of right breast, behind the nipple no equivalent of well circumscribed area visible in US, only scattered lesions.

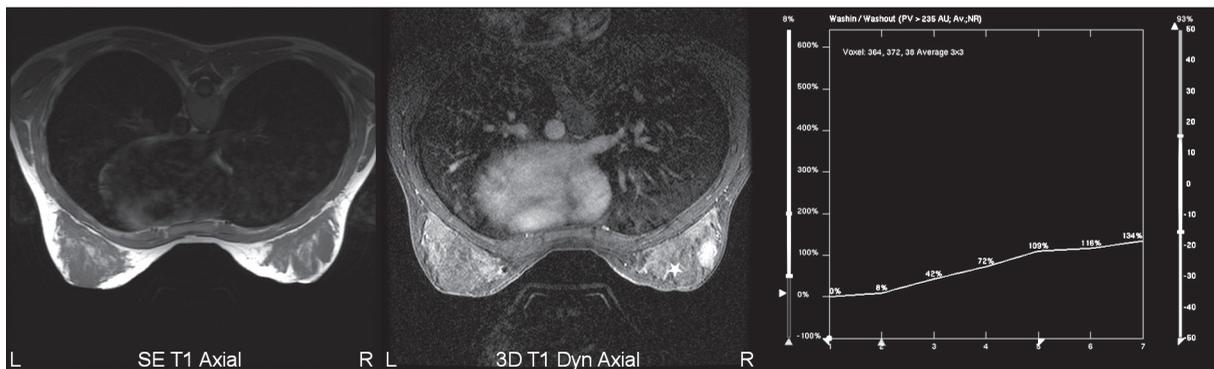


Figure 7. Patient A-K, MR of both breasts, SE T1-weighted and 3D T1-weighted after i.v. contrast medium administration (Multihance 0.1 ml/kg) dynamic (6 times repeated with 56s intervals) sequences, axial slices – fibroadenoma in the upper external quadrant of right breast, behind the nipple no equivalent of well circumscribed area visible in US, only scattered lesions. Enhancement curve type Ib in marked point.

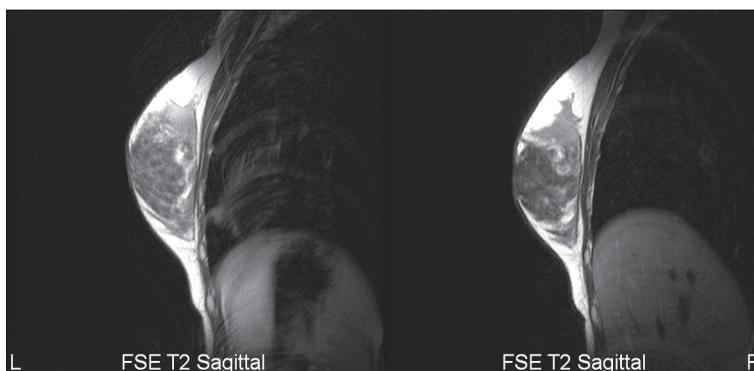


Figure 8. Patient A-K, MR of both breasts, FSE T2-weighted sequence, sagittal slices – behind the nipple no equivalent of well circumscribed area visible in US, only scattered lesions.

Both patients refused the excision of the lesions verified as PASH (the second patient had only one nodule of fibroadenoma juvenile type excised). Follow-up US every 6 months and MR every year did not show any progression in the course of 4 years in the first and 6 years in the second patient.

Discussion

Pseudoangiomatous stromal hyperplasia is a benign proliferative process of the breast stroma which, histopathologically, reveals complex, fused areas, lined with flattened myofibroblasts [4]. It is said that the lesion is a result of an incorrect hormonal reactivity of some myofibroblasts, especially to the changes of progesterone concentration [4,5].

Clinical presentation of PASH involves a wide range of lesions, from fine foci revealed microscopically by chance, to extensive nodular breast lesions. However, while the accidentally revealed clinically silent lesions can be quite popular, (Ibrahim [3] found PASH foci in 23% of the excised samples obtained in the diagnostic process of other breast pathologies), the nodular PASH is a very rare disease [1,2]. PASH characteristics found on US and mammography were presented several times in the literature already [1,2] – especially in the paper by Hargaden [1], basing on a large material of 149 patients. And so, the mammographic examination reveals most often a well circumscribed mass lesion or an asymmetrical breast parenchyma density, without calcifications (if present, then due to some other comorbidity).

Mammography was not performed in the presented cases due to the young age of the patients.

US examination reveals typically an oval, well circumscribed hypoechoic lesion, less frequently isoechoic, with heterogeneous echogram and the presence of fluid foci; while behind the lesion there is an enhancement, less frequently the decreased echogenicity.

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In both presented cases, the lesion on US was well circumscribed, hypoechoic and including small cysts in the first case.

Articles concerning PASH on MRI have been much less numerous and presenting some single (or a few casuistic) cases, not the analysis of some larger groups of patients [6–12].

A small, well circumscribed nodular lesion looked exactly like fibroadenoma, and the dynamic examination revealed a fast-increasing contrast enhancement in the first 2 minutes, which then slowed down a little [6].

The larger nodular lesion showed a well circumscribed polycyclic area with inhomogeneous, mostly decreased, signal intensity in T2-weighted sequences, and in the dynamic examination: a gradually and continuous increase of contrast enhancement (gently increasing enhancement curve) [7].

In case of a diffuse lesion resulting in an asymmetrical enlargement of the breast, an extensive oedema of the stroma and a higher volume of the glandular and fibrous tissue was observed in T2-weighted sequences, while the dynamic examination showed a fast increase of contrast enhancement in the first 2 minutes, and then a slowdown, until the 7th minute after IV contrast medium administration [8].

The above mentioned, observed by other authors, features of PASH found on MRI are similar to the ones revealed in the images obtained in the presented cases. This particularly concerns the dynamic of enhancement, typical for benign lesions.

Conclusions

Although the nodular type of PASH is a rare pathology, it should be known to the radiologists, as its features are partially overlapping with the ones of lesions of other type, including the fibroadenoma, phyllodes tumor or a low-grade angiosarcoma.