Pseudoangiomatous stromal hyperplasia (PASH) is a benign entity of the breast. It is a histological diagnosis and is characterised by a network of vascular-like clefts lined by endothelial-like spindle cells, against a background of stromal hyperplasia. Previously thought to be rare with only about a hundred cases described in literature, PASH is now diagnosed with increasing frequency. The non-mass-forming or diffuse form of PASH is believed to have an incidence of 23% in breast biopsies and has been reported in 25% of cases of gynecomastia (1). The clinical spectrum varies from incidental microscopic findings to a focal palpable or non-palpable mass. One case of PASH presenting as a rapidly growing mass has been described in literature (2). Histological and radiological, we can make a distinction between diffuse non-mass forming PASH and nodular PASH, which is less frequent. However, most case reports in radiological literature is of nodular PASH, which is indistinguishable from fibroadenoma (on mammography, ultrasound and MRI). Here, we wish to report a case of diffuse PASH with its mammographic, ultrasonographic and magnetic resonance imaging findings.

Case report

A 33-year-old woman presented at the gynaecologist with the chief complaint of a painful right breast and pain at the right axillary region. Clinical examination showed some mild redness and tenderness on the right breast. The patient was treated with antibiotics for suspected mastitis. She showed some improvement, but because of remaining tenderness additional tests were done. Blood results were completely normal. Mammography showed a larger amount of dense glandular tissue in the right breast (the mammography couldn’t be retrieved for publication). There was no architectural distortion, nor microcalcifications. Ultrasound confirmed a larger amount of glandular tissue in the right breast, compared with the left breast (Fig. 1). There were no signs of mastitis.

Because of these non-specific changes, MRI of the breast was performed. Precontrast axial view T2-weighted images showed a larger right breast with diffuse heterogeneous T2 hyperintense signal alterations (Fig. 2). On contrast-enhanced subtraction images there is a diffuse
or ILC (invasive lobular carcinoma) couldn’t be ruled out. According to the ACR BI-RADS grading system, the lesion was categorised as suspicious abnormality (category 4). Therefore, a vacuum-assisted biopsy with a 10 G needle was performed. This happened under the guidance of MRI, given the extent of the lesion and the fact it was best seen on MRI. Four biopsies of 1.5 cm length were taken in the retro-areolar region and six in the pre-pectoral space. This delivered the pathological evidence of diffuse PASH (Fig. 5).

Discussion

Pseudoangiomatous stromal hyperplasia (PASH) is a mesenchymal tumor of the breast with benign clinical outcome. It is histologically characterised by a network of slit-like spaces lined by endothelial-like spindle cells against a background of stromal hyperplasia. It is a histological diagnosis and must be differentiated from a low grade angiosarcoma, which has similar vascular clefts, but is lined by endothelial cells and contains blood cells. Also, pleomorphism, mitotic activity and necrosis is not present in PASH (3).

Because it is mainly diagnosed in premenopausal women or postmenopausal women under hormone replacement therapy, PASH is believed to be hormone-related (4).

The presentation of PASH is non-specific. It can be a clinical insignificant microscopic finding or a diffuse disease presenting as gynecomastia or detected coincidentally on screen-
ings mammography. In rare cases it can present as a firm, painless and mobile mass (nodular PASH) with no associated nipple or skin change. As stated in previous studies (5), the most patients with the clinical presentation of PASH had no mammographic abnormalities. The most frequent abnormalities on mammography were of a dense homogeneous mass with smooth to partially or even ill-defined borders, and second as a localised increased stroma or focal asymmetric density. The majority gets a score of three according to the BI-RADS scoring system: probably benign lesion.

Ultrasound is normal in most cases of mammographic detected PASH. The nodular form presents as a well-defined solid mass with hypoechoicinity or heterogeneous and with acoustic features ranging from posterior enhancement to mild posterior shadowing. Large lesions can contain lace-like reticula (echo-genic or hypoechogenic) and areas of scattered cystic changes or even central hyperechogenic areas (1, 3).

On MRI images nodular PASH has the same characteristics as a fibroadenoma, with rapid enhancement in the two minutes after contrast injection, followed by a slowly increasing enhancement during the rest of the examination (3). While diffuse PASH presents as a focal or segmental non-mass like clump of enhancement with a plateau or persistent phase (6). In our case the distribution pattern of non-mass like enhancement is diffuse and asymmetric, which haven’t been described before. While nodular PASH is mostly classified in the ACR BI-RADS system as category 2 (benign findings) or 3 (probably benign findings), diffuse PASH is more often classified as category 4 (suspicious abnormalities) (7). For detaining pathological evidence fine-needle aspiration cytology is frequently unsatisfactory because there is cytopathological overlap with fibroadenoma, there are a lot of dry samples (samples with no cells) and often could not be discriminated from alternative diagnosis (4). Often core needle biopsy is needed. In the late 1990s vacuum-assisted biopsy came available. This sampling technique enables more accurate and larger tissue volume sampling, what makes it most appropriate in case of microcalcifications and tissue distortion or asymmetry in density (8). Ultrasound is the first-line mode for guiding breast biopsy because of its availability, low cost and no need for ionizing radiation. For lesions that are only visible on mammography, like microcalcifications, stereotactic guidance is the choice. MRI is increasingly used because it gives a precise determination of the extent of disease, including the presence of multifocal or multicentric disease. If there are any suspicious features from sampling, further sampling is needed because PASH can coexist with malignant processes. When PASH is diagnosed, no further intervention is needed given the benign course of PASH. A large study by Celliers et al. (4) for example investigated 73 patients diagnosed with PASH with a follow-up period between one and eight years. This study showed that not one patient developed cancer. But in case of co-existent malign disease or if patient prefers, PASH can be excised. In the same study there was one case of recurrent disease at the same site. In the literature recurrence rate after excision range from 15 to 22% (4).

References