

Case 13684

Uncommon case of mammary Paget's disease (MPD) after ductal carcinoma in situ (DCIS) treated with breast-sparing surgery and brachytherapy

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Section: Breast Imaging

Published: 2016, Jul. 18

Patient: 68 year(s), female

Clinical History

68-year-old woman, status after left breast DCIS, breast-sparing surgery (histology: healthy margins) and brachytherapy 3 years before [Fig. 1, 2], was referred for MRI because of a 2-year history of sporadic nipple discharge, eczema and itching without palpable mass. At admission no nipple discharge was detectable. Previous imaging studies were normal [Fig. 3, 4].

Imaging Findings

Ultrasound (Fig. 5): In lower-inner quadrant at 8:00, in the scar area two contiguous hypoechoic sharply-defined focal lesions of 16x7 mm and 9x8 mm and at 9:00, in the scar area one hypoechoic not well-defined lesion of 15x13x12 mm, without increased perfusion. Several round lymph nodes could be detected in the ipsilateral axilla with normal structure (<1 cm). Result: BIRADS IV".

MRI (Fig. 6): Volume reduction of the left breast after lumpectomy. New retraction of the nipple with new-onset enhancement lesion sub areolar of about 10x16 mm (Gottingen score of 5: strong initial signal increasing over 100%, plateau, inhomogeneous enhancement, unclear boundary). Signs of diffusion restriction in the diffusion-weighted imaging. Result: BIRADS IVB".

A punch biopsy of the nipple at 8:00 revealed chronic dermal inflammation with focal atypical intraepidermal cells. Paget disease of the nipple, immunohistochemically positive for CDK7, EMA and HER2, while P63 was negative.

Discussion

MPD is an uncommon malignant disease of the nipple-areolar complex (NAC) and represents 0.5-4.3% of breast cancers (BC) [1]. Usually it occurs in postmenopausal women: the mean age at diagnosis of MPD in association with DCIS is 63.8 years [2]. In more than 90% MPD is accompanied by underlying BC [3] (with or without palpable mass). If no mass is detectable, MPD is usually associated with DCIS [3, 4].

Clinical findings are unilateral eczema, itching, erythema, erosion or ulceration of the NAC with scaly, bloody or clear discharge or an oozy lesion. Therefore, MPD may be misdiagnosed as benign dermatological disease [4]. Because the presented case neither had a palpable mass nor showed a clear clinical progression, a benign process was suspected.

The features in mammography (asymmetry, architectural distortion, malignant calcifications or lump) are unspecific. Thickening of the NAC is a highly suspected malignant sign; however, it should be considered that mammography is normal in 22-50% of BC patients [3]. Ultrasound (US) features (e.g. ductal ectasy, flattening or retraction of the NAC) are also not pathognomonic [5]. MRI shows enhancement or thickening of the NAC, with or without lump. MRI can display unsuspected breast lesions even in the absence of clinical findings, and is more sensitive than other imaging techniques [3, 6, 7]. Radiologists should be aware: normal nipples may demonstrate enhancement on MRI, usually bilaterally.

Subareolar malignancies are difficult to diagnose because they can either be unspecific or can mimic normal structures in mammography and US. In cases with previous treatment (like in the presented one) the image-guided diagnosis might be a challenge due to scars and post-irradiation changes [8, 9, 10]. Therefore, we recommend to use multimodal imaging to further improve the diagnostic sensitivity [11]. Additionally, clinical visible pathologies in the nipple after conservative therapy required a biopsy.

Final Diagnosis

Biopsy: DCIS (high-grade malignancy) close to the nipple, and MPD.

Differential Diagnosis List

Eczema / lichen simplex chronicus, Psoriasis, Atopic or contact dermatitis, Florid papillomatosis of the nipple, Paget's disease, Pagetoid Bowen's disease (squamous cell carcinoma in situ), Pagetoid Spitz nevus or malignant melanoma, Other malignancies (Merkel cell carcinoma, mycosis fungoides, Langerhans histiocytosis, nipple adenoma and ductal exocrine carcinoma)

Figures

Figure 1 Mammography 2009

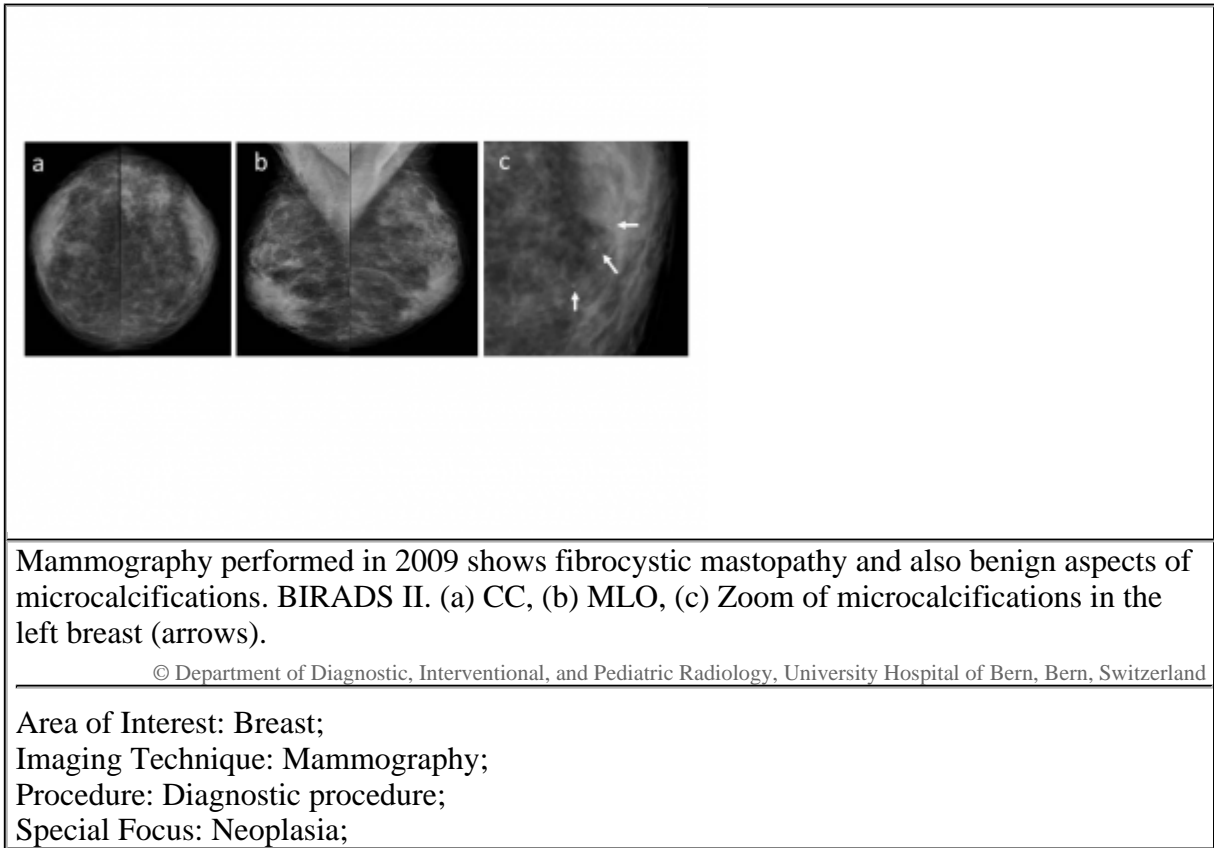


Figure 2 Summary of the clinical history

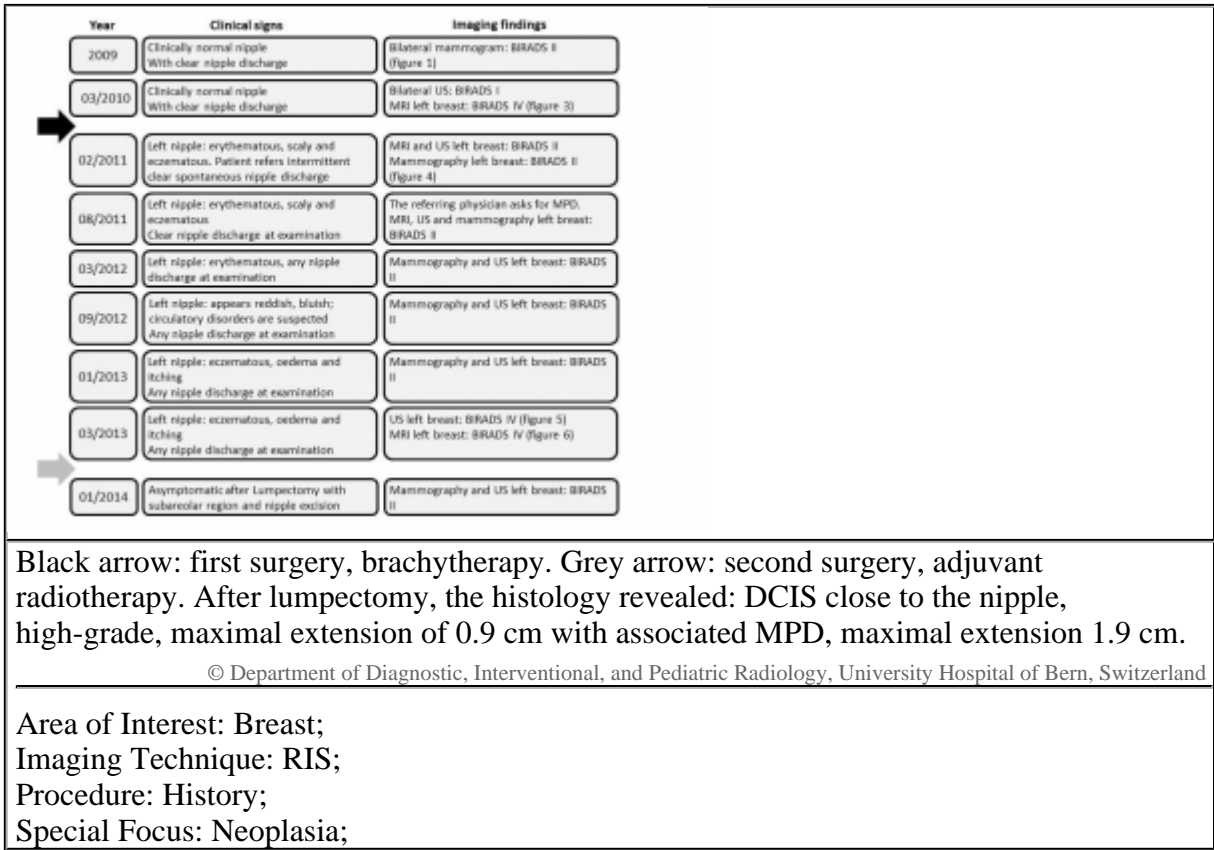
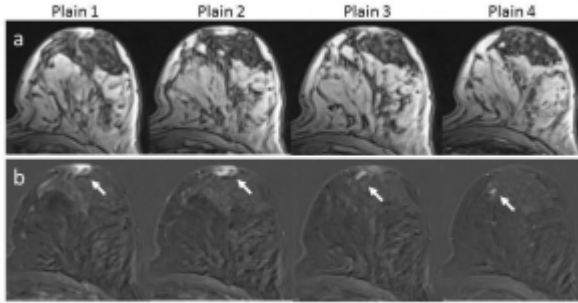


Figure 3 MRI 03/2010

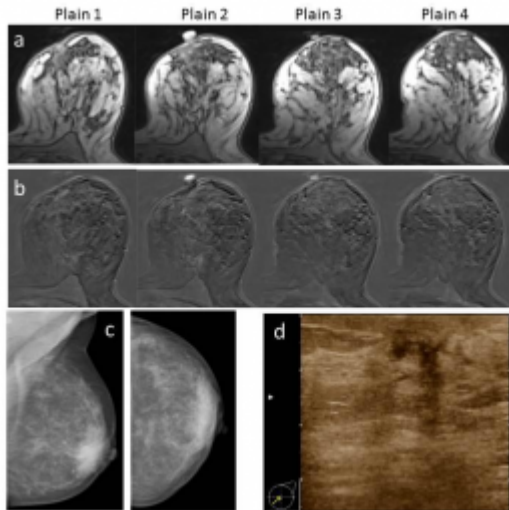


MRI 03/2010: occasional spontaneous nipple discharge. Extended duct with protracted enhancement in the left-upper quadrant, up to the nipple, small adjacent foci. DD: intraductal papilloma, intraductal malignancy. BIRADS IV (a) Dynamic-MRI (b) subtracted-MRI 4-minutes-delay.

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Area of Interest: Breast;
Imaging Technique: MR;
Procedure: Diagnostic procedure;
Special Focus: Neoplasia;

Figure 4 Multimodal imaging 02/2011

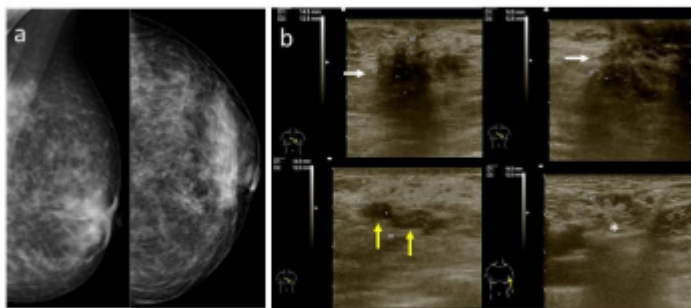


02/2011 Multimodal imaging: history of spontaneous discharge, slightly retractile scar tissue. MRI: postoperative changes: BIRADS II. (a) Dynamic-MRI 4-minutes-delay, (b) subtracted-MRI after 4 minutes, (c) mammography BIRADS II, (d) US scar. BIRADS II.

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Figure 5 Mammography 01/2013 & US 03/2013

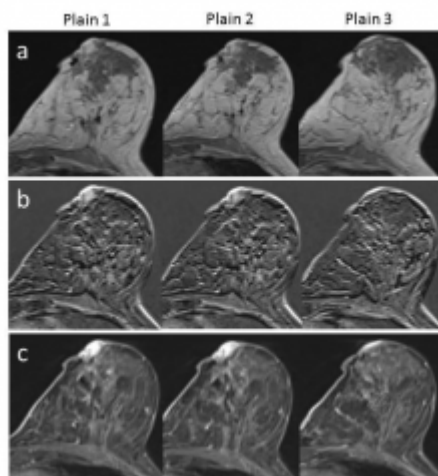


(a) Mammography 01/2013 BIRADS II. (b) US 03/2013: top left and right, white arrows: scar. bottom left, yellow arrows: two contiguous hypoechoic, well-defined focal lesions. Histology: MPD. Axillary lymph node (bottom right, asterisk): normal. BIRADS IV

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Figure 6 MRI 03/2013



MRI 03/2013: shows a new onset suspicious lesion in the left subareolar region with retraction of the nipple. BIRADS IV. (a) Dynamic and (b) subtracted after 2 minutes. (c) Fat saturated sequence after 4 minutes.

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Citation

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