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 ecstasy, 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

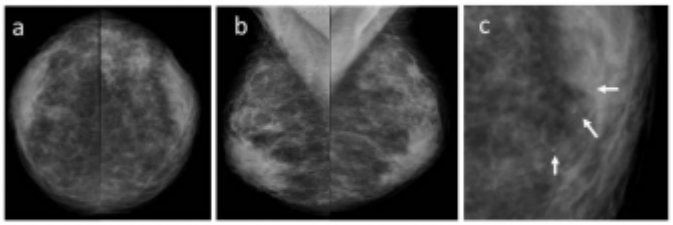
Biopsy: DCIS (high-grade malignancy) close to the nipple, and MPD.

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

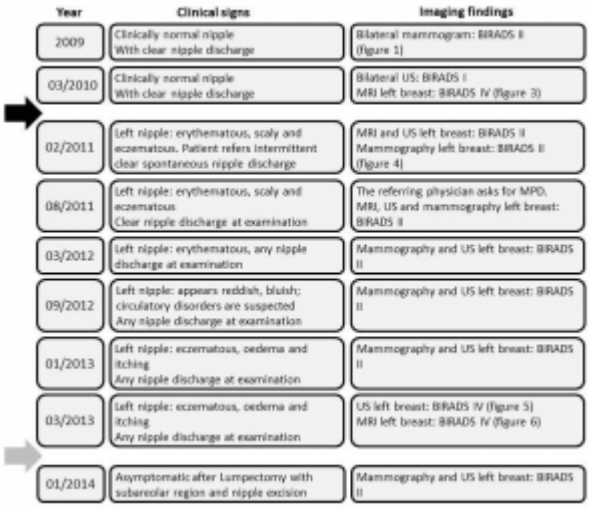
Figure 1 Mammography 2009



Mammography performed in 2009 shows fibrocystic mastopathy and also benign aspects of microcalcifications. BIRADS II. (a) CC, (b) MLO, (c) Zoom of microcalcifications in the left breast (arrows).

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

Figure 2 Summary of the clinical history

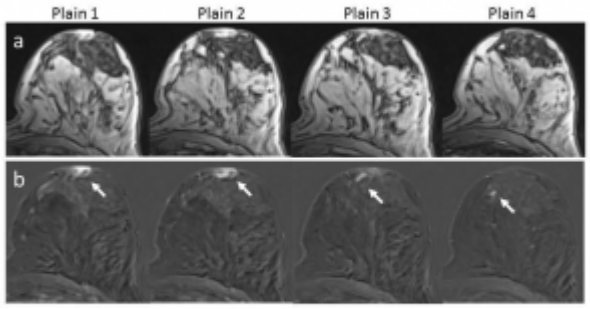


Year	Clinical signs	Imaging findings
2009	Clinically normal nipple With clear nipple discharge	Bilateral mammogram: BIRADS II (Figure 1)
03/2010	Clinically normal nipple With clear nipple discharge	Bilateral US: BIRADS I MRI left breast: BIRADS IV (Figure 3)
02/2011	Left nipple: erythematous, scaly and eczematous. Patient refers intermittent clear spontaneous nipple discharge	MRI and US left breast: BIRADS II Mammography left breast: BIRADS II (Figure 4)
08/2011	Left nipple: erythematous, scaly and eczematous Clear nipple discharge at examination	The referring physician asks for MPD. MRI, US and mammography left breast: BIRADS II
03/2012	Left nipple: erythematous, any nipple discharge at examination	Mammography and US left breast: BIRADS II
09/2012	Left nipple: appears reddish, bluish; circulatory disorders are suspected Any nipple discharge at examination	Mammography and US left breast: BIRADS II
01/2013	Left nipple: eczematous, oedema and itching Any nipple discharge at examination	Mammography and US left breast: BIRADS II
03/2013	Left nipple: eczematous, oedema and itching Any nipple discharge at examination	US left breast: BIRADS IV (Figure 5) MRI left breast: BIRADS IV (Figure 6)
01/2014	Asymptomatic after Lumpectomy with subareolar region and nipple excision	Mammography and US left breast: BIRADS II

Black arrow: first surgery, brachytherapy. Grey arrow: second surgery, adjuvant radiotherapy. After lumpectomy, the histology revealed: DCIS close to the nipple, high-grade, maximal extension of 0.9 cm with associated MPD, maximal extension 1.9 cm.

Area of Interest: Breast;
Imaging Technique: RIS;
Procedure: History;
Special Focus: Neoplasia;

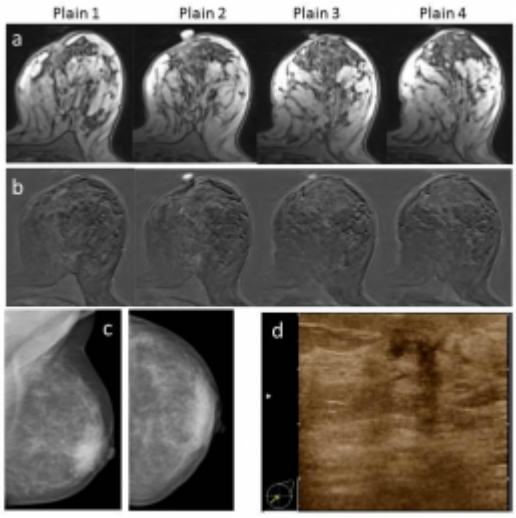
Figure 3 MRI 03/2010



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.

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.

Area of Interest: Breast;
Imaging Technique: Mammography;
Procedure: Diagnostic procedure;
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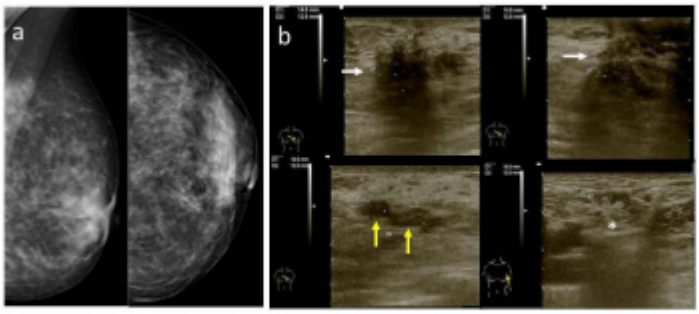
Figure 5 Mammography 01/2013 & US 03/2013



(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

Area of Interest: Breast;
Imaging Technique: Mammography;
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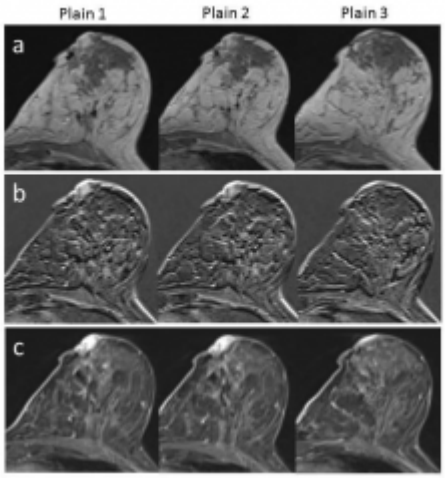
Figure 6 MRI 03/2013



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.

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

References

[1] Morrogh M, Morris EA, Liberman L, Van Zee K, Cody HS3rd, King TA (2008) MRI identifies otherwise occult disease in select patients with Paget disease of the nipple J Am Coll Surg 206(2):316-21

[2] Chen CY, Sun LM, Anderson BO (2006) Paget disease of the breast: changing patterns of incidence, clinical presentation, and treatment in the US Cancer 107(7):1448-58

[3] Lim HS, Jeong SJ, Lee JS, Park MH, Kim JW, Shin SS, Park JG, Kang HK (2011) Paget disease of the breast: mammographic, US, and MR Imaging findings with pathologic correlation Radiographics 31(7):1973-87

[4] Sandoval-Leon AC, Drews-Elger K, Gomez-Fernandez CR, Yepes MM, Lippman ME (2013) Paget's disease of the nipple Breast Cancer Res Treat 141(1):1-12

[5] Gaspari E, Ricci A, Liberto V, Scarano AL, Fornari M, Simonetti G (2013) Case report: An unusual case of mammary Paget's disease diagnosed using dynamic-enhanced MRI Case reports in radiology 206235. Doi:10.1155/2013/206235

[6] Malich A, Fischer DR, Wurdinger S, Boettcher J, Marx C, Facius M, Keiser WA (2005) Potential MRI interpretation model: differentiation of benign from malignant breast masses AJR Am J Roentgenol. 185(4):964-70

[7] Sripathi S, Ayachit A, Kadavigere R, Kumar S, Eleti A, Sraj A (2015) Spectrum of Imaging Findings in Paget's Disease of the Breast-A Pictorial Review Insights Imaging 6(4):419-29

[8] Moon JY, Chang YW, Lee EH, Seo DY (2013) Malignant invasion of the nipple-areolar complex of the breast: usefulness of breast MRI AJR Am J Roentgenol 201(2):448-55

[9] Shah SK, Shah SK, Greatrex KV (2005) Current role of magnetic resonance imaging in breast imaging: a primer for the primary care physician J Am Board Fam Pract 18(6):478-90

[10] Hassett MJ, Jiang W, Habel LA, Nekhlyudov L, Achacoso N, Acton L, Schnitt SJ, Schrag D, Punglia RS (2016) Characteristics of second breast events among women treated with breast-conserving surgery for DCIS in the community Breast Cancer Res Treat 155(3):541-9

[11] Shao H, Li B, Zhang X, Xiong Z, Liu Y, Tang G (2013) Comparison of the diagnostic efficiency for breast cancer in Chinese women using mammography, ultrasound, MRI, and different combinations of these imaging modalities J Xray Sci Technol 21(2):283-92

Citation

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URL: http://www.eurorad.org/case.php?id=13684