Unusual presentation of more common disease/injury

Diffuse primary large B cell lymphoma of leg type presenting on the breast: a rare case of surgical excision

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Primary breast lymphoma is a rare subtype of breast malignancy whose diagnosis relies on histopathological characteristics and a high index of suspicion in clinically unusual presentations of breast pathology. The authors report a rare case of diffuse primary large B cell lymphoma of leg type presenting as a single lesion confined to the breast. Unusually for this subtype of lymphoma, the lesion was surgically excised and the patient, to date, has made an uneventful recovery.

BACKGROUND

This is a rare case of diffuse primary large B cell lymphoma of leg type presenting as a single lesion confined to the breast. This subtype of lymphoma usually presents in the leg and if it is seen on the trunk, it usually presents with synchronous leg lesions. Diagnosis relies on histopathological characteristics and a high index of suspicion in clinically unusual presentations of breast pathology.¹ The case highlighted to our team the importance of considering unusual pathological diagnoses when presented with atypical breast lesions. Diffuse large B cell lymphoma of leg type is usually associated with a poor prognosis and the fact that our patient underwent surgical excision and has made a good recovery to date, provides an important lesson about possible patient prognosis in such cases.

CASE PRESENTATION

A 90-year-old Caucasian lady presented to the breast outpatient clinic in April 2010 with a right areolar lump. She was wheelchair-bound and co-morbidities included

hypertension, previous stroke and osteoarthritis but her health was otherwise commensurate with her age. There was no family history of breast cancer or other malignant disease. On examination, she had an unusual lesion covering the lower half of the areola. Clinically, it appeared to be almost like a blister but was hard on palpation with no evidence of a lump beneath it (figures 1 and 2). The lesion was painful and she was keen to have it excised. Systemic examination was normal with no evidence of lymphadenopathy and she had no history of lymphoma.

INVESTIGATIONS

Mammography revealed thickening around the right nipple with some fine microcalcifications deep to the lesion (figure 3). There was no convincing evidence of any masses or ductal carcinoma in situ within the breasts. Ultrasound of the right breast confirmed a 15×21×7 mm area of thickening in the right areola with no evidence of infiltration into the breast and no duct dilatation behind the nipple.

Figure 1 Right breast lump covering lower half of areola.

Figure 2 Close up of areolar lump showing blister-like appearance.

Figure 3 Mammography showing fine microcalcifications but no evidence of breast mass or ductal carcinoma in situ.

TREATMENT

We chose to perform an excision rather than a biopsy because of the unusual clinical picture and the patient's strong wish to have the lesion removed.

She underwent wide local excision and histology revealed a diffuse large B cell lymphoma of non-germinal centre phenotype. Sections showed a diffuse infiltration of large atypical lymphoid cells that expanded from the dermis into the subcutaneous tissue (figure 4). Immunohistochemistry showed that the large atypical lymphoid cells were CD20, CD79a, BC1-2, interferon regulatory factor-4/multiple myeloma-1 (IRF-4/MUM-1) and IgM positive (figure 5). CD10, CD23, CD30, Cyclin D-1, CD5 and CD3 were negative (figure 6). The diagnosis of a lymphoma of the category diffuse large B cell lymphomas of leg type was made, given the localised nature of the lesion.

OUTCOME AND FOLLOW-UP

Surgical excision was complete. The haemato-oncological opinion was that any adjuvant treatment (systemic or local) was not necessary. She went on to make an uneventful postoperative recovery. At 10 months' follow-up, she is well and free of symptoms.

DISCUSSION

Primary cutaneous diffuse large B cell lymphoma, leg type (PCLBCL-LT) is characterised by a predilection for the lower limb (72%), a high proportion of Bcl-2 expression, an

Figure 4 HE stain showing skin with lymphoid infiltrate and large atypical cells.

Figure 5 Immunohistochemistry stain showing positive B cell marker CD20.

advanced age at onset and frequent relapses and extracutaneous dissemination.² Aside from the lower limb, cases have been reported occurring in the head, arms and trunk. To our knowledge, there has only been one other reported case of diffuse large B cell lymphoma of leg type presenting on the breast, which presented with synchronous leg lesions.³ Indeed Patrizi *et al* reported that in all the existing reports of large series of patients since the new WHO-European Organisation for Research and Treatment of Cancer (EORTC) classification (derived by consensus from the EORTC classification and the WHO classification) for PCLBCLs, only 25 patients with a solitary trunk/dorsum involvement have until now been mentioned out of 209 patients with a diagnosis of PCLBCL-LT.⁴ Our case appears to be unique in that the lesion was confined to the breast and, contrary to the poor prognosis usually associated with diffuse large B cell lymphoma of leg type, our patient has made an excellent recovery. As there has been no relapse, locally or systemically at 10 months, the excision appears to have been adequate treatment. She will undoubtedly need to remain under close follow-up to monitor for disease recurrence given the tendency of this disease to disseminate. This type of lymphoma is distinguished from

Figure 6 B cells are negative for CD10.

primary cutaneous follicle centre lymphoma by being positive for Bcl-2 protein and IRF-4/MUM-1,⁵ as was the case with our patient.

Primary breast lymphoma should be remembered as a differential diagnosis of a breast lump especially in those with unusual clinical or radiological presentations. Ideally, a core biopsy should be performed to come to a preoperative diagnosis, but the unusual superficial nature of this lesion and patient's wishes precluded this in our case. Ultimately diagnosis relies on histopathological analysis, with immunohistochemistry playing a vital diagnostic role in distinguishing between the subtypes of lymphoma.

Learning points

- ▶ Primary breast lymphoma should be remembered as a differential diagnosis of a breast lump.
- ▶ Unusual clinical or radiological presentations should prompt consideration of rarer forms of breast disease such as lymphoma.
- ▶ Ultimately diagnosis relies on routine histopathological analysis of all potentially malignant breast lesions.
- ▶ Diffuse primary large B cell lymphoma of leg type may present as a single lesion on the trunk without associated leg lesions and effective surgical excision is possible.

Competing interests None.

Patient consent Obtained.

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