CASE REPORT

Acquired progressive lymphangioma of the nipple

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SUMMARY

A 47-year-old woman presented with left nipple itch and discomfort. On physical examination she was found to have a 7 mm lesion. She underwent bilateral mammography and bilateral breast ultrasound which were normal. A punch biopsy of the lesion was performed in the office and the specimen submitted to pathology. Histopathological examination showed ectatic vascular spaces lined by flattened, cytologically bland endothelial cells dissecting the dermal collagen. Evident lymphatic valves were present within the vascular spaces confirming that the vessels were lymphatic in nature. The diagnosis of acquired progressive lymphangioma (benign lymphangioendothelioma) was rendered.

BACKGROUND

Acquired progressive lymphangioma (benign lymphangioendothelioma) is a rare benign locally-infiltrative vascular tumour with only 44 cases reported in the literature to date.

CASE PRESENTATION

A 47-year-old woman presented to the breast clinic with left nipple itch and discomfort of 6 months duration. She did not notice any palpable masses or skin rash. Her medical history was significant for hypogammaglobulinaemia. Her family history was negative for breast or ovarian cancer. Physical examination showed a 7 mm asymmetrical outgrowth of the left nipple. She did not have any palpable masses or skin rash. Axillary examination revealed no lymphadenopathy. She underwent bilateral mammograms and breast ultrasound which were normal.

INVESTIGATIONS

The patient underwent bilateral mammography and bilateral breast ultrasound which were normal.

DIFFERENTIAL DIAGNOSIS

- Paget's disease
- Breast cancer
- Florid papillomatosis of the nipple
- Radial scar
- Vascular tumour

TREATMENT

A punch biopsy of the lesion was performed in the office and histopathological examination showed ectatic vascular spaces lined by flattened, cytologically bland endothelial cells dissecting through the dermis. Evident lymphatic valves were present within the vascular spaces confirming that the vessels were lymphatic in nature (figures 1 and 2). The diagnosis of acquired progressive

Figure 1 Histopathological examination showing ectatic vascular spaces (white arrow) lined by flattened, cytologically bland endothelial cells dissecting through the dermis (black arrow).

lymphangioma (benign lymphangioendothelioma) was rendered.

OUTCOME AND FOLLOW-UP

At her 2-week follow-up visit in clinic the patient symptoms has resolved and the biopsy site was healing well with no evidence of wound infection.

DISCUSSION

Benign lymphangioendothelioma (acquired progressive lymphangioma) is a rare slow-growing and locally infiltrative lymphatic vascular tumour that was first described by Jones *et al*¹ in 1990. It presents clinically as a haematoma-like patch or an erythematous plaque most commonly affecting the legs, head and neck.^{2 3}

Histopathologically, benign lymphangioendothelioma is characterised by proliferation of thin-walled single and discontinuous layer of flat endothelial cells interspersed between layers of collagen. Immunohistochemistry shows lymphatic endothelial cells that are CD31, CD34, podoplanin, LYVE-1 and PORX-1 positive.^{2 3} Immunostaining is negative for human herpes virus 8 (HHV-8) excluding the diagnosis of Kaposi's sarcoma, a disease that can be confused with benign lymphangioendothelioma.⁴ Forty-four cases have been described in the literature with three reported cases involving the breast.⁴⁻⁶ Rosso *et al*⁶ reported a case in which the disease involved the mastectomy scar in a patient who underwent radiotherapy for breast cancer. Paik *et al*⁷ reported a case that was in association with HIV infection. To the best of our knowledge, this is the first case of benign lymphangioendothelioma that involves the nipple and in association with immunosuppression secondary to hypogammaglobulinaemia.

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Figure 2 Histopathological examination showing ectatic vascular spaces (white arrow) lined by flattened, cytologically bland endothelial cells dissecting through the dermis (black arrow).

Treatment is needed in symptomatic cases only and consists of surgical excision and is curative although few cases of local recurrence have been described.⁴

Learning points

- ▶ Benign lymphangioendothelioma is a rare benign, locally infiltrative lymphatic vascular tumour that can involve any part of the body.
- ▶ It is characterised by proliferation of thin-walled lymphatic endothelial cells.
- ▶ It is very important to differentiate this disease entity from Kaposi's sarcoma or angiosarcoma.
- ▶ Treatment in symptomatic cases consist of surgical excision.

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Competing interests None.

Patient consent Obtained.

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