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LETTERS TO THE EDITOR

Lymphangioma Circumscriptum of the Vulva of Late Onset

Sir,

Lymphangioma circumscriptum (LC) is a benign disorder of the lymphatic system that only rarely affects the vulva.

We report a case of vulval LC that developed in an elderly patient, without evidence of secondary lymphatic damage.

CASE REPORT

A 76-year-old woman presented at our department with a 2-year history of recurrent, clear fluid oozing from vesicular lesions of the vulva.

There was no history of any significant genital or extragenital disorders. Physical examination revealed multiple, persistent, translucent, thick-walled vesicles, filled with a clear fluid, scattered over the labia majora, which appeared swollen, thickened and partially hyperpigmented (Fig. 1).

A biopsy specimen from the lesion showed dilated lymph vessels, lined by a single layer of endothelial cells, in the upper dermis, with epidermal acanthosis and hyperkeratosis (Fig. 2). A diagnosis of LC was made.

No gynaecological disease was apparent and further investigations, particularly computed tomography scan of the abdomen and pelvis, revealed no abnormality.

A magnetic resonance imaging (MRI) scan was performed, but no alteration was evidenced in the vulval area.

Topical treatments to prevent irritation or infection, as well as periodic follow-ups, were recommended, but a more aggressive approach was refused by the patient.

DISCUSSION

Cutaneous lymphangiomas may arise secondary to the obstruction of the lymphatic flow, in a previously normal lymphatic system, and thus they are better defined as lymphangiectasias or acquired lymphangiomas.

To our knowledge, 12 cases of vulval lymphangiectasias have been reported in the literature; 8 cases were associated with surgery and/or radiotherapy for cervical tumours (1–7), 2 with inguinal lymph node tuberculosis (8, 9) and 2 with Crohn's disease with perineal involvement (2).

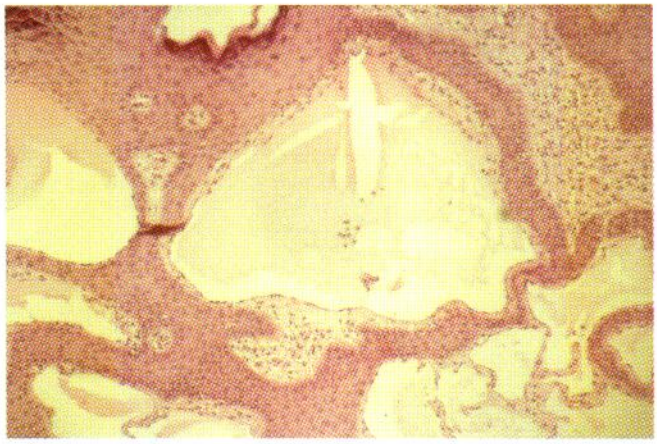

Fig. 1. Multiple, thin-walled, translucent vesicles on the labia majora.

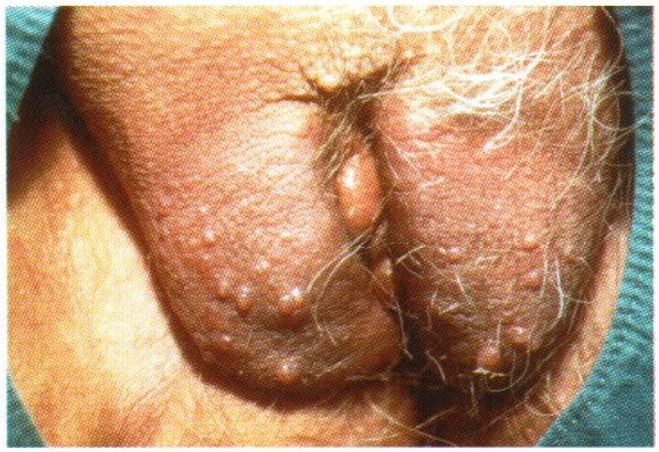

Fig. 2. Dilated lymph vessels in superficial dermis, and epidermal acanthosis and hyperkeratosis (haematoxylin and eosin, ×100).

Classic LC is considered to be a naevoid malformation of dermal and subcutaneous lymph vessels. On the vulva, it has been up to now reported in only 4 cases (10–12), and the oldest reported patient with this condition was a 42-year-old woman (10).

In spite of the extraordinarily late onset, the present case appears to be a classic form of LC, since no obvious cause of acquired lymphatic damage at the vulval level could be determined.

In particular, in our patient there were no history or signs of surgery or radiotherapy, Crohn's disease, tuberculosis or other inflammatory diseases, i.e. recurrent cellulitis, potentially responsible for a vulval lymphatic obstruction.

Whimster (13) postulated that LC resulted from a collection of deep lymphatic cisterns with pulsating muscular coats, connected through dilated channels with the superficial vesicles.

Recently, an MRI study supported this hypothesis, emphasizing the utility of this technique in the assessment of lesion extent, both therapeutically and prognostically (14).

As previously reported (11), in our case the MRI scan showed that the lesion was confined to the vulval skin, without evidence of dilated lymphatic channels in depth.

However, the treatment of cutaneous lymphangiomas is often difficult and recurrences are common.

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