LYMPHANGIOMA CIRCUMSCRIPTUM OF LABIA MINORA- A RARE CASE REPORT

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ABSTRACT

BACKGROUND

Lymphangioma circumscriptum is a rare, uncommon, hamartomatous and benign lesion of no specific aetiology involving the lymphatic channels in the deep dermal and subcutaneous layers.(1) It can occur either as a congenital abnormality or as an acquired damage to previously normal lymphatic channels.(2) Lymphangiomas can occur anywhere on the skin and the mucous membranes. The most common sites are the head and the neck followed by the proximal extremities, the buttocks and the trunk. However, sometimes they can be found on the intestines, pancreas and the mesentery. Deeper cystic lesions usually occur in areas of loose areolar tissue; typically, the neck, the axilla and the groin. We report a rare case of congenital lymphangioma circumscriptum of left labia minora in an 18-year-old female.

KEYWORDS

Hamartomas, Labia Minora, Lymphangioma Circumscriptum, Lymphatic Channels.

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BACKGROUND

Lymphangioma circumscriptum, the common form of cutaneous lymphangioma, is characterised by persistent multiple clusters of translucent vesicles that usually contain clear lymph fluid (often compared with frogspawn).(3) These vesicles represent superficial saccular dilations from underlying lymphatic vessels that occupy the papilla and push upward against the overlying epidermis. Each skin lesion may range from a minute vesicle to a small bulla-sized lesion. These vesicles can be clear or vary from pink to dark red due to presence of serosanguineous fluid or haemorrhage. These vesicles often are associated with verrucous changes, giving them a warty appearance.

Case Report

An 18-year-old unmarried female patient presented with a painful, mildly pruritic swelling on the left labia minora of 1-year duration with watery discharge from the swelling. She also gave history of left lower limb oedema since childhood. She had similar swelling 10 years back, for which she was operated. The reports of the previous surgery were not available. Menstrual history was unremarkable. On clinical examination, a solitary flat pedunculated swelling was seen arising from left labia minora measuring 5 x 4 cms with 3 cms width of the base. Skin over the swelling was reddish-brown with no other secondary skin changes. The swelling was soft in consistency and watery/serosanguineous discharge was oozing from it. Ultrasound examination of abdomen and pelvis revealed normal study.

The patient was posted for surgery, subsequently we received the excised specimen. The specimen was fixed in 10% neutral buffered formalin, routinely processed and multiple sections were submitted for histopathological examination.

Gross specimen was received as multiple skin covered grey-brown soft tissue masses, largest measuring 6 x 3 x 1 cms and smallest measuring 1 x 1 x 0.5 cms. Cut sections of all the masses showed tan white-to-yellow areas (Figure 1).

Figure 1

Sections were submitted from the masses for histopathological examination. Light microscopy showed stratified squamous epithelium with acanthosis, elongated rete ridges with subepithelium showing multiple, ectatic, cystically dilated, thin-walled lymphatic channels with few of the lumina filled with pale eosinophilic secretions. Stroma showed myxomatous degeneration and sparse chronic inflammatory infiltrates (Figure 2). Differential diagnosis of haemangioma was considered. Confirmation was done by running Immunohistochemistry marker of CD-34 (Clone-QBEnd/10) which showed negativity in the lymphatic channels, whereas strong positivity in the blood vessels (Figure 3).

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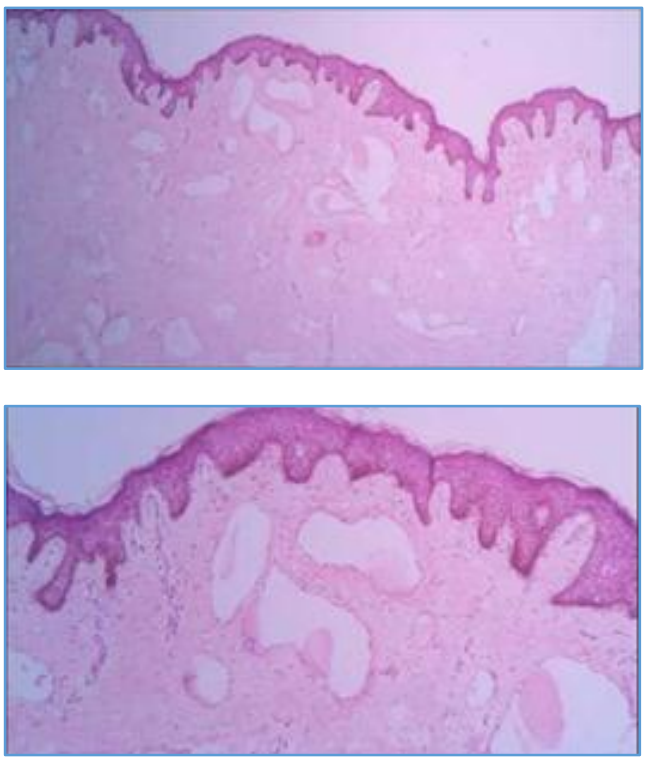

Figure 2

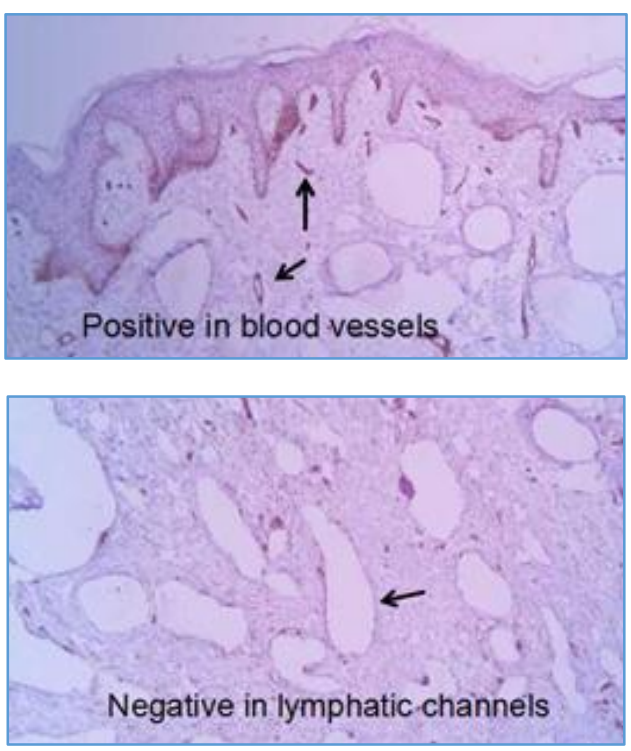

Figure 3

DISCUSSION

Lymphangioma accounts for approximately 4% of all vascular tumours.[4] It is classified into lymphangioma circumscriptum, cavernous lymphangioma, cystic hygroma and benign lymphangioendothelioma. The term lymphangioma circumscriptum was coined by Morris.[5]

Lymphangioma circumscriptum is a rare, uncommon, hamartomatous benign lesion of no specific aetiology involving the lymphatic channels in the deep dermal and subcutaneous layers. In lymphangioma circumscriptum, the underlying lesions constitute abnormal dilated lymph vessels involving the upper part of the dermis. The typical picture consists of a small number of vesicles on the skin, which are usually present at birth or appear in early childhood.[6] In subsequent years, they tend to increase in size and number.[7] The sites of predilection are the proximal extremities, trunk, axilla and oral cavity, especially the tongue. Involvement in other areas such as the scrotum is not uncommon. Lymphangioma circumscriptum has a high recurrence rate after excision, because of its deep component.

In 1976, Whimster studied the pathogenesis of lymphangioma circumscriptum and proposed the basic pathologic process to be collection of lymphatic cisterns in the deep subcutaneous plane. These cisterns are separated from the normal network of lymph vessels, but they communicate with the superficial lymph vesicles through vertical, dilated lymph channels. These cisterns arise from a primitive lymph sac that fails to connect with the rest of the lymphatic system during its embryonic development. The vesicles formed in lymphangioma circumscriptum are outpouchings of these dilated projecting vessels.

The lesions are often present at birth or appear early in life (< 30 years). In localised form which occurs less frequently and appears as small, discrete lesions (< 1 cms) have no predilection for any site. The lesions may appear at any age. Vulvar lymphangioma circumscriptum can be either congenital or acquired.[8] Most common cause of acquired form is surgery and radiotherapy, especially for cervical carcinoma.

Different modes of treatment which includes surgical modalities are done.[9] These constitute local excision, simple or extended vulvectomy and abrasive modalities like carbon dioxide laser, liquid nitrogen, 5-fluorouracil, electrocoagulation, or sclerosing therapy with OK-432s.[10] Currently, surgery is an effective and well-tolerated treatment for most women. Recurrence is the major problem due to incomplete excision.[11] Our patient was treated surgically with complete excision.

CONCLUSION

Lymphangioma circumscriptum is a hamartomatous benign lesion of nonspecific aetiology. Involvement of labia minora is very rare. A differential diagnosis of lymphangioma circumscriptum should be considered for recurrent swellings in this region with unilateral limb oedema.

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