

Penile Lymphangioma Circumscriptum-A Rare Case Report

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Abstract

Lymphangioma circumscriptum is a rare disorder of lymphatic channels characterised by multiple grouped thin translucent vesicles. The lesions are often seen around the neck, upper trunk, proximal extremities and tongue. Genital involvement in females has been reported but penis is a rare site. On the other side congenital variety is extremely rare. Here we are presenting a rare case of congenital penile lymphangioma circumscriptum.

Keywords: Congenital; Penis; Lymphangioma circumscriptum

Introduction

Lymphangioma circumscriptum (LC) is a rare disorder of lymphatic channels characterised by multiple grouped thin translucent vesicles. Reported for the first time by Fox and Fox in 1870 under the name Lymphangiectodes [1], the entity was coined as "Lymphangioma circumscriptum" by Malcolm Morris in 1889. The lesions are often seen about the neck, upper trunk, proximal extremities and tongue. Genital involvement in females [2] has been reported but penis is a rare site of involvement. Though a few case reports of acquired genital lymphangioma circumscriptum [3-4] are available in the literature only occasional case reports of congenital or idiopathic variety of congenital penile lymphangioma circumscriptum are available. We report one such case.

Case report

A 10 year old male child presented in the skin department with complaints of painless vesicular lesions over penis starting at the age of 1 year. Over the course of subsequent 9 years the lesions gradually spread over the entire penis and scrotum and partly over the groins bilaterally.



Figure 1: Showing multiple vesicular lesions involving entire scrotum, penis, and adjoining groin and thigh.



Figure 2: Showing Multiple vesicular lesions involving entire scrotum, penis, and adjoining groin and thigh.

There was no history of itching, haemorrhage, swelling in the lower limbs, abdominal upset or difficulty in urination. Patient was a strict vegetarian with no history of alcohol or tobacco use. On general physical examination patient was of normal build. There were no signs of malnutrition, edema, icterus, cyanosis or lymphadenopathy. On local examination the lesions were warty and vesicular, evenly and densely distributed all over the penis and scrotum and scantily over the groin regions bilaterally. There was gross enlargement of scrotum and penis almost twice normal size for the age. He did not belong to endemic area of filariasis (Figures 1-9).

When vesicles are punctured, clear serous fluid comes out. On culture, this fluid was sterile in nature and on smear examination no microorganisms were found.

Patient was referred to the radiology department for Ultrasonography (USG) and Doppler evaluation to assess the degree of depth of soft tissue involvement and vascularity of the lesion.

On USG, scrotum and proximal half of the penis showed preservation of normal architecture with only thickening of the skin and subcutaneous tissues. The corpora cavernosa and spongiosum were normal in size and architecture. Dorsal penile artery showed normal peak systolic and end diastolic velocity and waveform on colour and pulse Doppler. Distal half of penis showed almost complete architectural distortion and was constituted by heterogeneously echogenic hypertrophic subcutaneous tissue. No evidence of increased vascularity was seen in the lesion on lying or standing position. Bilateral testes were normal in size architecture and vascularity. There was no inguinal lymphadenopathy. On USG abdomen and pelvis there was no lymphadenopathy or any organomegaly seen. Excisional biopsy of a single papule from inguinal lesions revealed hyperkeratosis and papillomatosis in the epidermis, dilated lymphatic vessels lined by endothelium throughout the papillary and reticular dermis and epidermis.

Discussion

Lymphangioma circumscriptum (LC) clinically presents as a circumscribed area of tense deep seated vesicles mostly filled with colourless fluid but occasionally tinged with varying quantities of blood giving variable colour from pink, red to black to the vesicles. It may be classic (extensive), localised (smaller), and spongy [5].

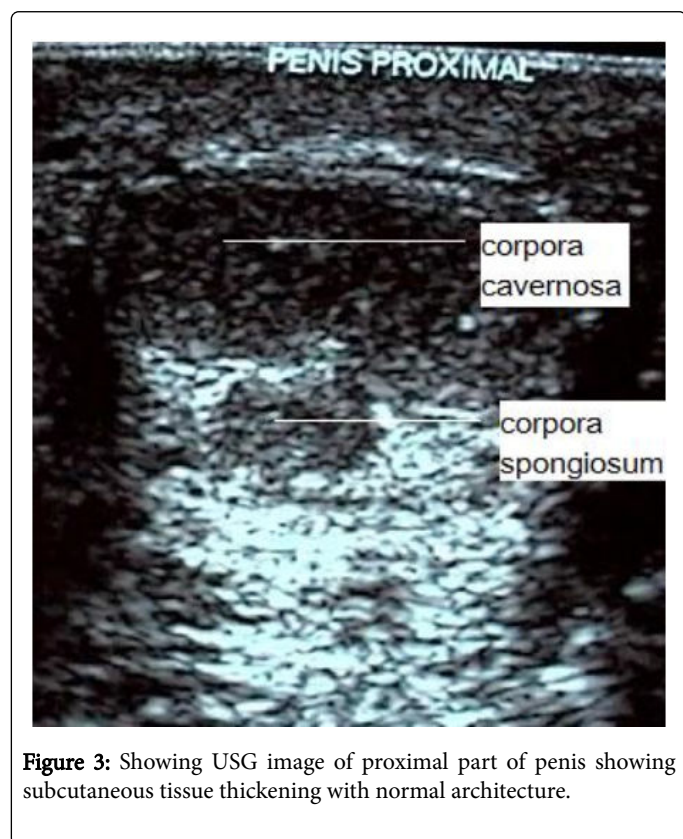


Figure 3: Showing USG image of proximal part of penis showing subcutaneous tissue thickening with normal architecture.

Classic LC is present at birth or appears soon afterward increasing progressively throughout childhood and persisting indefinitely. Pathologically the lesion is a sequestered cluster of large muscular

coated lymphatic cisterns, lying deep in the subcutaneous plane communicating via dilated superficial dermal lymphatics with the superficial vesicle. It is believed that contractions of the muscle layer of cisterns generate pulsations which transmit the increased pressure to abnormal superficial dermal lymphatics which subsequently over the course of time dilate to appear as vesicles on the skin. Clinically the lesions are usually symptomatic with oozing, minor bleeding and secondary infection, are usually larger than 1 cm² and are associated with thickening of subcutaneous tissues beneath the lesions. They often involve the proximal parts of limbs and adjacent limb girdle. USG, Doppler study and MRI may be required in extensive lesions to rule out vascular neoplasm and to determine the extent of the disease.

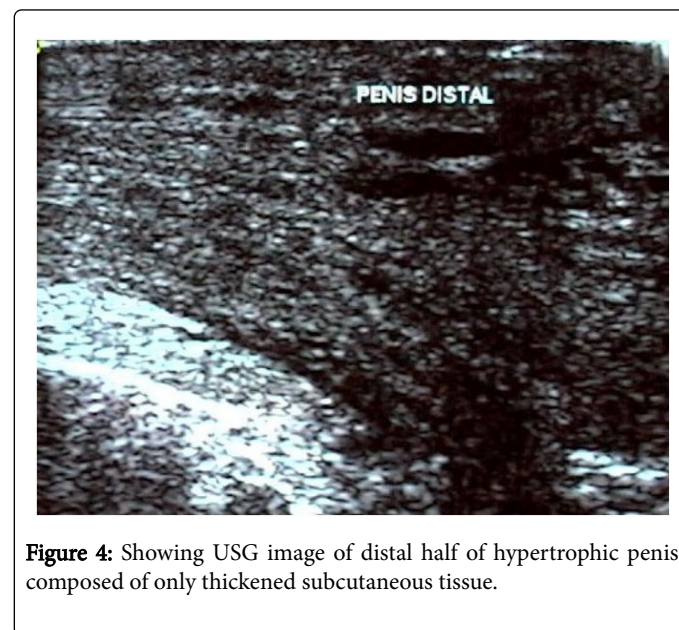


Figure 4: Showing USG image of distal half of hypertrophic penis composed of only thickened subcutaneous tissue.

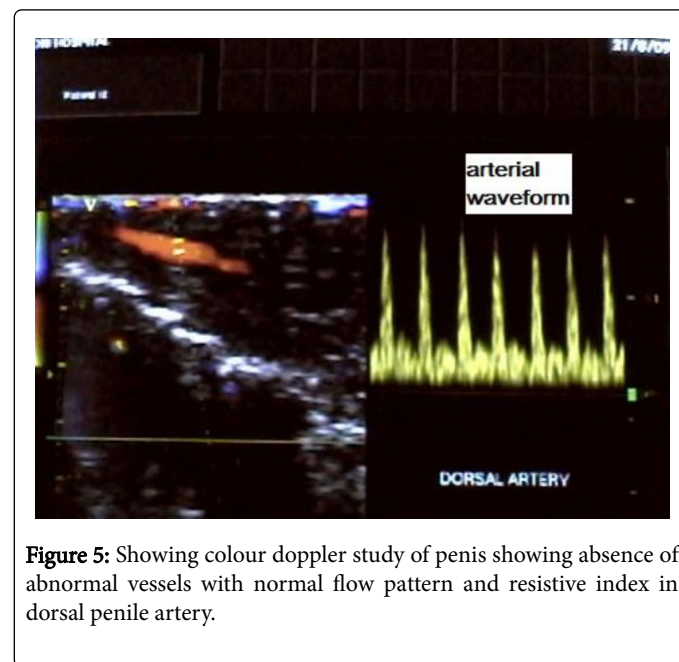


Figure 5: Showing colour doppler study of penis showing absence of abnormal vessels with normal flow pattern and resistive index in dorsal penile artery.

Local type of LC is acquired with a definite cause, is usually less than 1 cm² in size, has no definite site of predilection and is usually asymptomatic. Lymphatic obstruction secondary to post mastectomy

lymphedema, congenital unilateral lymphedema as a possible cause has been proposed for acquired cases of LC.

Lesions occurring at genitalia may create a fear of STD in the patient or be mistaken for either molluscum contagiosum or condyloma accuminatum or esthiomene by physicians.

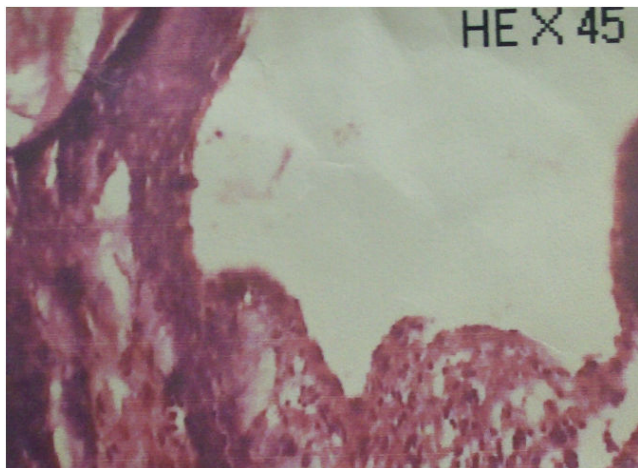


Figure 6: Showing skin biopsy showing hyperkeratosis and papillomatosis of epidermis with lymphatic vessels lined by endothelium in upper dermis s/o Lymphangioma circumscriptum.

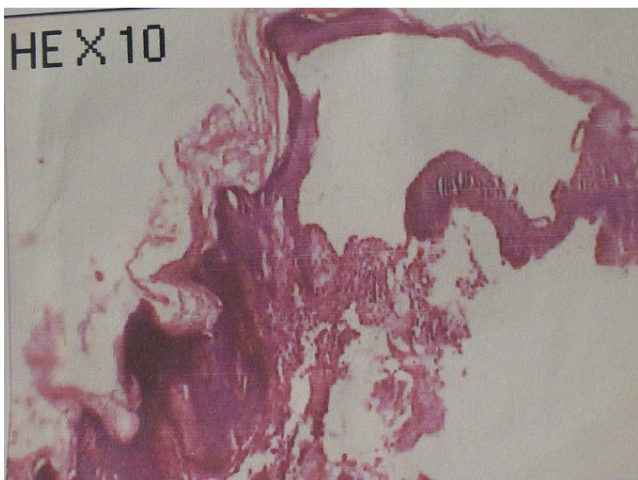


Figure 7: Showing skin biopsy showing hyperkeratosis and papillomatosis of epidermis with lymphatic vessels lined by endothelium in upper dermis s/o Lymphangioma circumscriptum.

Symptomatic treatment with topical silver sulfadiazine on ruptured lesions to prevent superimposed infections, palliative treatment in form of electrofulguration, cryosurgery, carbon dioxide laser for destruction of visible vesicles have been used. However, recurrence of lesions is very common. Radical resection or intralesional administration of sclerosants in penile lesions is often difficult, requires highly skilled soft tissue reconstruction and effect on sexual function is not known. One of the treatment options proposed after understanding the pathology of the lesions is to excise the subcutaneous cisterns right down to the level of deep fascia and leave the overlying skin intact for more successful and better cosmetic results [6]. Short follow up of nine such surgeries provided by Whimster showed excellent results with no new vesicles appearing and in fact partial regression of vesicles previously present.

Involvement of penis is extremely rare, only 11 cases of penile involvement reported till 2007 [7] and only one case report of penile LC with thickening of subcutaneous tissues and enlargement of penis and scrotum is reported. Possible cause of subcutaneous thickening in our case could have been compression of normal lymphatics by extensively distributed subcutaneous lymphatic cisterns of LC. We are reporting this case due to rarity of such kind of presentation and we should think this possibility even in adults in some circumstances.

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