Case Report

Idiopathic adult onset Lymphangioma circumscriptum of scrotum: An unusual case Scrotal lymphangioma circumscriptum

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Abstract

Lymphangioma circumscriptum (LC) is a lymphatic hamartoma that is usually congenital but may be acquired. It is seen in axilla, oral cavity, tongue, vulva, scrotum etc. Here we report a middle aged male who presented with multiple grouped vesicles in the scrotum with no prior history of radiotherapy or surgery. There are very few cases of idiopathic LC of scrotum in the published literature. The case is being reported because of rarity of presentation in adulthood, unusual site and absence of any underlying etiology.

Keywords: Idiopathic, dermal lymphatics, adult.

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Introduction

Skin normally doesn't have lymphatic channels in the papillary dermis, however in Lymphangioma Circumscriptum these become dilated causing protrusion of the lesion above the surface of skin [1]. Lymphangioma circumscriptum (LC) is a rare, benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatics [2].It can occur anywhere in the skin and mucous membranes, the common sites being axillary folds, shoulders, neck, proximal part of the limbs, tongue, vulva, buccal mucus membrane and uncommonly the scrotum [3]. Penoscrotal lymphangioma is an unusual entity of which only around ten cases have been reported to date [4].

Case

A36 year old North Indian male, labourer by occupation presented to the dermatology OPD with complains of multiple, grouped lesions in the scrotum since 4 years. There was no history of fever or weight loss. He was married with single sexual partner. On examination multiple grouped vesicles were seen involving almost the complete scrotal skin (Figure 1). An occasional vesicle had ruptured however there was no evidence of local inflammation or cellulitis.A provisional diagnosis of filariasis, condyloma accuminatum or molluscum contagiosum was made. Hematological profile did not show eosinophilia and he was non-reactive for HIV, HBs Ag or VDRL. Fluid from vesicle did not stain for any microorganism and culture was sterile. Ultrasound was done to rule out a deeper systemic involvement. A 4mm deep biopsy was taken from one of the intact vesicles and sent for histopathology. On microscopic examination the epidermis was thinned out and atrophic. Superficial dermis showed many dilated angiolymphatic channels lined by flattened endothelium (Photomicrograph 1,2). Few of these channels were filled with clear fluid. There was no endothelial swelling (as seen in lymphangitis), granulomatous reaction or adult worm in the section examined and a diagnosis of lymphangioma circumscriptum was made. The patient was sent to Plastic Surgery department for surgical excision of lesion.

Manuscript received: 14th March 2018
Reviewed: 18th March 2018
Author Corrected: 24th March 2018
Accepted for Publication: 31st March 2018

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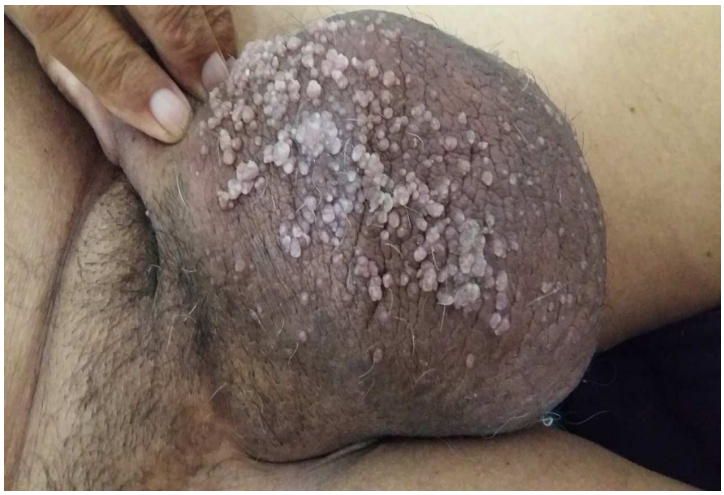
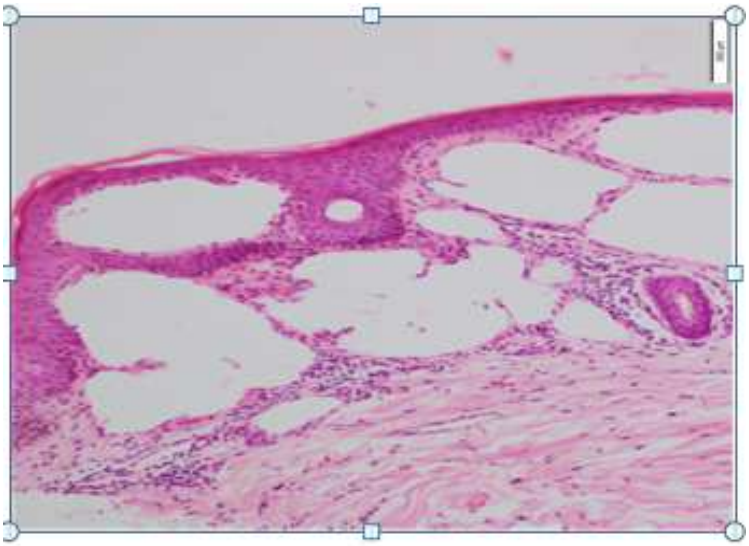
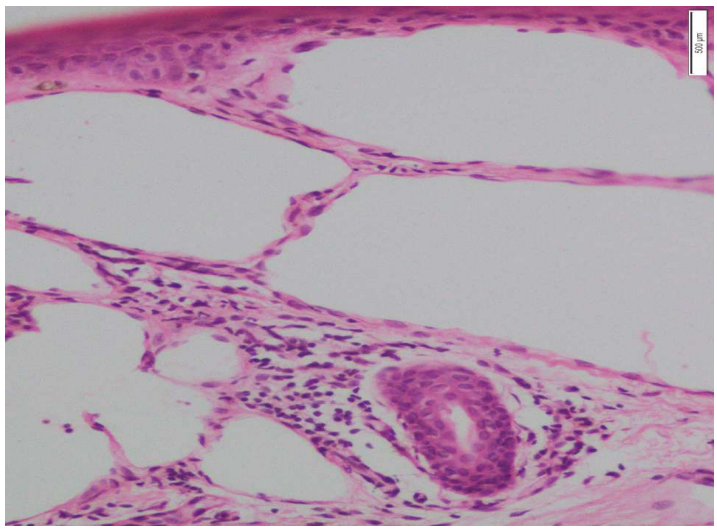


Figure 1: Multiple pearly white grouped vesicles in scrotum filled with clear fluid



Photomicrograph 1: H&E: 100X Thinned out epidermis with numerous dilated angiolymphatic channels in superficial dermis



Photomicrograph 2: Dilated dermal lymphatics lined by flattened endothelium

Discussion

Lymphangioma circumscriptum is a congenital malformation of superficial lymphatics present at birth or soon after. It presents as a group of vesicles containing clear fluid (resembling frog's spawn), the size of which can be up to 5 mm in diameter [3]. Sometimes the vesicles can be hemorrhagic. Hemorrhage can occur spontaneously or secondary to trivial trauma. The proximal part of the extremities is the commonest site involved**.** It may be classic (extensive), localised (smaller), and spongy [3].

Whimster first proposed the pathogenesis of LC in 1976. He postulated that LC is a collection of subcutaneous lymph cisterns, which arise during embryonic development, that are not connected to the lymphatic system and therefore unable to drain the lymph received from surrounding tissue. The cisterns are lined with muscle that contracts and, by applying pressure, produces protrusions on the skin which manifest as grouped vesicles [2].

Acquired LC develops in advanced age, possibly due to injury to deep collecting lymphatics, caused by trauma, surgery, radiotherapy or infections such as filariasis, lymphogranuloma venereum, donovanosis or tuberculosis [5]. LC is clinically identified by translucent or hazy vesicles of different sizes which are grouped like frog spawn or, less commonly, as diffuse swelling to a particular area. These swellings frequently have accompanying verrucous alterations giving them a warty appearance and, if there is significant hyperkeratosis, the swelling may clinically resemble lymphogranuloma venereum [5]. A strong clinical suspicion is needed for diagnosis of this disease with histopathological confirmation. Ultra sonography and CT scan of the abdomen or pelvis are helpful in patients who have suspicious extensions of cystic lesion to the retroperitoneum or pelvis.

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. The most common postoperative complication is its recurrence with an incidence of 25–50% within 3 months, which is usually due to improper surgical approach or inadequate excision of the tumor [8].Other complications are edema, prolonged lymphatic drainage, and local infection. [9] Malignant transformation to lymphangiosarcoma has also been reported [11].

Conclusion

Since LC in scrotum is rare and tends to mimic certain infectious diseases like filariasis and molluscum, the clinicians should be aware of LC in adult males without a prior disease to avoid missing the diagnosis and to prevent inappropriate treatment. Adequate surgery including removal of subcutaneous tissues and fascia is the treatment of choice to prevent recurrences[12].

Ethics: Informed consent was obtained from the patient in order to publish this case report

Authors Contributions

NS: Concept and design of the study, reviewed the literature, manuscript preparation and critical revision of the manuscript.

RJ: Conceptualized study, literature search, review of study

Funding: Nil, **Conflict of interest:** None initiated, **Permission from IRB:** Yes

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How to cite this article?

Shirazi N., Jindal R. Idiopathic adult onset Lymphangioma circumscriptum of scrotum: An unusual case Scrotal lymphangioma circumscriptum. Trop J Path Micro 2018;4(1):63-66. doi: 10.17511/jopm.2018.i1.11.

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