

Cutaneous Lymphangioma Circumscriptum

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Abstract

Cutaneous Lymphangioma Circumscriptum (CLC) is a rare, benign, vascular malformation of dilated superficial lymphatic channels. It is characterized clinically by localized clusters of thin-walled translucent or red-purple colored vesicles that contain clear or hemorrhagic lymphatic fluid. The role of imaging is important to determine the extent of CLC, particularly the deep cisterns. The treatment options include surgery, sclerotherapy, radiotherapy, and laser therapy. We present the characteristics of a case of this rare entity.

Introduction

Cutaneous Lymphangioma Circumscriptum (CLC) is a relatively uncommon, benign, disorder of the lymphatic channels with unclear etiology [1]. They account for 4% of all vascular tumors, but comprise 25% of benign vascular growths in children [2]. The disorder is characterized by groups of thin-walled translucent or red-purple colored vesicles [1]. Management of lymphatic malformations is challenging due to the lack of satisfactory treatments [3]. We describe the clinical and dermoscopic features of a case of this malformation occurred in a young girl.

Case Report

A 14-year-old girl admitted to our department with slowly growing lesions on the left trunk which first appeared during infancy. On physical examination, we observed multiples translucent vesicles, grouped into a bouquet. As well as, clusters of pink to purple papules scattered over the right trunk with some hemorrhagic crusts (Figure 1). Dermoscopy showed light brown lacunas with pale septa and some linear vessels (Figure 2). The rest of the clinical examination was normal. The abdominal ultrasound was without abnormalities. Considering the limited extent of the disease and age of patient, we decided to abstention therapeutic.

Comments

Cutaneous lymphangioma Circumscriptum (CLC) was described by Fox and Foxin 1878. Lymphangiomas are hamartomatous, congenital malformations of the lymphatic system that may involve the skin and subcutaneous tissues [4]. Major symptoms of CLC include oozing, bleeding,



Figure 1: Multiples translucent vesicleslike and clusters like of pink to purple papules scattered over the right trunk.



Figure 2: Dermoscopy showing light brown lacunas with pale septa and some linear vessels.

itching, pain, swelling in affected area, and secondary infection [3]. Cutaneous lymphangioma circumscriptum is typically seen as a small number of translucent cutaneous vesicles at or soon after birth, although possible at any age. They may range in size from minute vesicles to larger bullae. Specifically, 2-4 mm clusters of vesicles with pink, red, or black discoloration as a byproduct of hemorrhage are common. These vesicles often resemble frog spawn and tend to increase in number and size with occasional rupture leading to bleeding or drainage [4]. Lesions are usually seen on buttocks, inguinal region, axillary folds, proximal extremities, and oral cavity [3]. Dermatoscopic examination of CLC shows a lacunar/saccular pattern. Presence of clear fluid appears as light brown lacunas with pale septa. If blood has infiltrated in a patchy manner, scattered areas of redness appear within the lacunas. However, if there is homogeneous distribution of blood in the CLC, a pink hue overwhelms the lacunar

structures [5]. Histopathology is characterized by acanthosis and papillomatosis of epidermis with numerous ectatic lymphatic channels in the upper dermis which may often extend through deep layers [6]. Radiological exploration is necessary to look for deep localization. Treatment is typically undertaken for cosmetic reasons or complications such as fluid drainage, pain, and infection risk that can negatively impact quality of life [1]. The treatment options include surgery, sclerotherapy, radiotherapy, and laser therapy. Given the risk of recurrence, abstention therapeutic may be justified [7].

Conclusion

We aim to present the clinical and dermoscopic features of a particular and unusual lymphatic malformation, in order to be easily recognized by young clinicians.

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