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Extensive Lymphangioma Circumscriptum of Lower Leg: A Rare Case Report

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Abstract: Lymphangioma circumscriptum is a rare benign lymphatic malformation arising from sequestration of lymphatic tissue which is not communicated with the rest of the lymphatic channels. It was classified into localized (smaller), classical (extensive), and spongy subtypes. Most common locations are proximal part of limbs, axilla, shoulders, groins and buttocks but distal part of limbs is a rare site. We report a case of 25 year old female with an extensive subtype of lymphangioma circumscriptum of distal part of lower leg.
Keywords: Lymphatic malformation, Lymphangioma circumscriptum, Extensive type

INTRODUCTION

Lymphangioma circumscriptum(LC) is a rare benign hamartomatous malformation of the lymphatic channels affecting the skin and subcutaneous tissue.It is arising from sequestration of lymphatic tissue which is not communicate with the rest of the lymphatic channels [1]. It is usually present as persistent, multiple clusters of translucent vesicles that are mainly due to superficial saccular dilations from underlying lymphatic vessels that occupy the papilla and push upward against the overlying epidermis [2]. It was classified into localized (smaller), classical (extensive), and spongy subtypes [3].

We report a case of extensive subtype of lymphangioma circumscriptum of distal part of left lower leg in a young female, managed by above knee amputation**.**

CASE REPORT

A 25- year- old female was admitted in surgical unit with huge swelling of distal part of left lower leg. The swelling was since childhood with gradual increase in size. She had discomfort and difficulty in walking.

On physical examination revealed a huge soft spongy swelling of the left lower leg, involving the whole foot and ankle. The overlying skin was corrugated, roughened and pigmented. X-rays of lower leg showed a soft tissue swelling with no evidence of bone erosion.Due to morbidity reason above knee amputation was done on patient's request.The surgical specimen was sent to histopathology department.The macroscopic examination revealed a huge mushroom like mass involving the whole foot and distal part of left leg measuring 30x20 cms in size, overlying skin was folded thickened, hyperpigmented and lichenified at some places (Fig.1). The rest of leg was atrophied. On cutting through the skin, deeper tissue and bone appears unremarkable.

On microscopic examination, epidermis shows hyper keratosis, acanthosis with complex papillary folding. Beneath the epidermis there are dilated irregular lymphatic channels occupying the papillary dermis and protrude into epidermis lined by attenuated and flattened endothelium. There are areas of lymphocytic infiltration throughout dermis and subcutaneous tissue with occasional dilated lymphatic channel in the deep dermis (Fig. 2). These findings were consistent with lymphangioma circumscriptum.

DISCUSSION

The pathogenesis of lymphangioma circumscriptum (LC) was first described by Whimster in 1976. According to him, lymphatic cisterns in the deep subcutaneous tissue arise from early lymph tissue sacs during embryonic development. These sacs were distinct entities that did not connect to the remainder of the lymphatic system. Finally, these sacs are lined by muscle fibers that contract and induce pressure that causes outpunching's in the walls of the sacs. These outpunching's eventually protrude from the skin surface to become what are clinically described as vesicles [4].

LC is most commonly affects proximal part of limbs, axilla, shoulders, groins, buttocks that contains extensive lymphatic network, rarely affects distal limb, ankle and foot region. Clinically it present as soft tissue

swelling and overlying skin show persistent, multiple cutaneous, clusters of translucent vesicles over large area. It is usually present since childhood and can be congenital with no sex predilection [5]. In our case, the swelling was noticed from childhood involving the distal part of lower leg and foot.

It is divided into smaller lesions, localized form (less than 7 cm) and extensive lesions, classic form (more than 7 cm). Extensive type of LC shows thickened, hypertrophied, hyperpigmented, lichenified skin, clusters of vesicles may be scattered over the involved skin [6]. In the present case, the swelling was of size 30x20 cm and showed features compatible with extensive type of LC.

The histopathologic features of lymphangioma circumscriptum include dilated lymph vessels in the upper dermis that may extend into the subcutis. These dilated vessels cause expansion of the papillary dermis.There may be acanthosis and hyperkeratosis of the overlying epidermis [7]. These finding were also noted in our case.

The definitive treatment for lymphangiomas is surgical excision of both the superficial and deep components. The main indications for treatment include both its cosmetic appearance and prevention of complications such as cellulitis [8]. Our patient was a young girl presented with huge swelling of distal part of lower leg since childhood with marked morbidity for that above knee amputation was done. The other palliative treatments are superficial x-ray therapy, radiotherapy, argon laser, and sclerotherapy [9, 10].

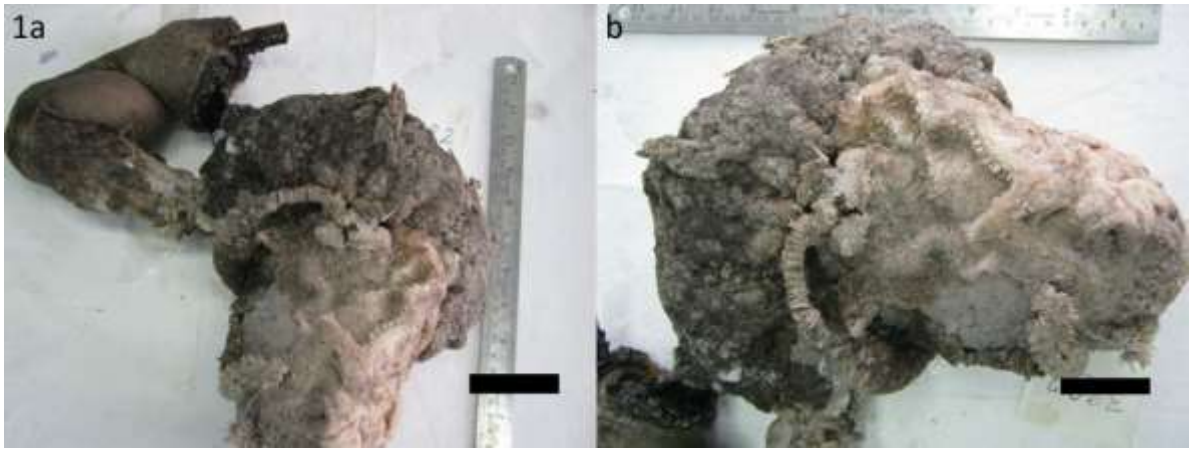

Fig. 1a & b: Gross of above knee amputation showing huge mushroom like mass involving the whole foot and distal part of left leg, overlying skin was folded thickened, corrugated

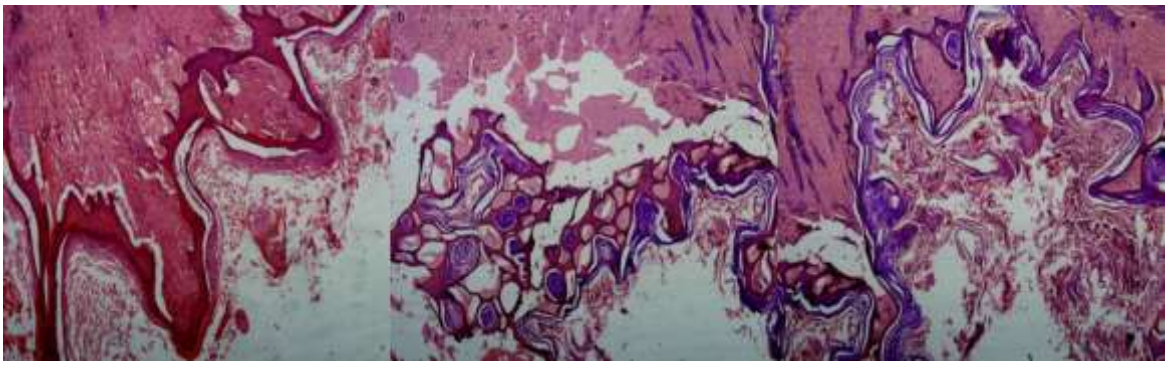

Fig. 2a, b & c: Low magnification view shows dilated irregular lymphatic channels in dermis and protrude into epidermis. Epidermis shows hyper keratosis, acanthosis with complex papillary folding. Dermis shows lymphocytic infiltration [H&E]

CONCLUSION

In conclusion, Extensive type of lymphangioma circumscriptum is uncommon and this entity in distal part of lower limb is still rare. An early diagnosis and treatment will help in improving functional, psychological disturbances and cosmotic disfigurement.

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