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Full Length Case Report

UNCOMMON PRESENTATION OF LYMPHANGIOMA CIRCUMSCRIPTUM: CASE SERIES

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ABSTRACT

Introduction: Lymphangioma circumscriptum (superficial lymphatic malformation) is a congenital malformation of the superficial lymphatics presenting as a group of deep seated vesicles like papules.

Method: a 12 yr old male presented with multiple non scaly slowly progressive asymptomatic plaques of variable size and shape over the right forearm and a single plaque on left arm and right leg since 8 years in skin OPD, SMS hospital and clinically was thought to be 1) Cutaneous Leishmaniasis 2) Borderline tuberculoid leprosy. A biopsy from the lesion was taken and submitted for histological examination.

Result: on performing histopathology, these lesions came out to be of lymphangioma circumscriptum. Discussion: this case presents with different presentation as verrucous lesions, wherein usually it presents with clear vesicles. On microscopy it is composed of numerous dilated lymphatics in the superficial and papillary dermis with the presence of clear fluid. Overlying dermis shows acanthosis and hyperkeratosis. The surrounding stroma shows scattered lymphocytes. This entity is the most common subtype of lymphangioma.

Conclusion: this interesting case of lymphangioma presenting with uncommon clinical features exemplifies a case where utmost vigilance is required for correct diagnosis. This case is taken for better understanding of lymphangioma.

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INTRODUCTION

Lymphangiomas are uncommon, hamartomatous malformations of the lymphatic system that involve the skin and subcutaneous tissues and were first described by Fox and Fox in 1879. Lymphangioma circumscriptum is the most common form of cutaneous lymphangioma. It accounts for 25% of all benign vascular tumors in children (Schwartz *et al.*, 2011). Male to female ratio is 3:1. It arises mainly in infancy but can occur at any age. Typical clinical findings are multiple clear vesicles that may appear pink, red and black. Unusual presentations are multiple verrucous papules and plaques. The common sites involved are proximal extremities, genitals, trunk, axilla and oral cavity (Schwartz *et al.*, 2011).

Case report

Two males aged between 10-13 years presented with long standing multiple asymptomatic plaques with verrucous

surface over the extremities which was progressive in nature to the skin opd, SMS medical college, Jaipur and clinically diagnosed as Tuberculosis verrucosa cutis. Skin punch biopsy was taken and sent for histopathological examination. A 25-year-old female was admitted in surgical unit with huge progressive swelling of left foot since childhood. On gross examination revealed huge mushroom like mass measuring 30cm x 20cm, overlying skin was folded thickened, verrucous, hyper pigmented and lichenified. There were multiple scattered clusters of vesicles over the involved skin, varying in color from whitish to blackish.

Microscopy: Irregular acanthosis, hyperkeratosis with follicular plugging. Superficial dermis is showing many dilated lymphatic channels with presence of clear fluid. Occasional vessels show RBC. Stroma shows scattered lymphocytes. No epithelioid cells or necrosis is seen.

DISCUSSION

Lymphangiomas are malformations of the lymphatic system. There is benign proliferation of lymph vessels separated into

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Figure 1. Gross photograph showing large verrucous mass in lower limb

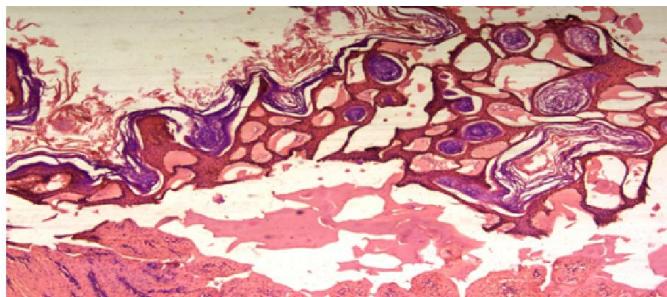


Figure 2. microphotograph showing dilated lymphatic channels in superficial dermis (100X)

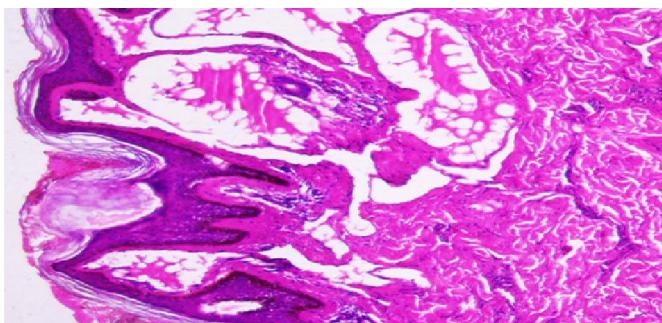


Figure 3. microphotograph showing acanthosis, hyperkeratosis, follicular plugging and dilated lymphatic channels (400X)

two categories i.e. the superficial includes lymphangioma circumscriptum; deeper group includes cavernous lymphangioma and cystic hygroma (Patel et al., 2009). Lymphangiomas account for 4 % of all the vascular tumors and 25 % of all the benign vascular tumors in children (Schwartz et al., 2011). The etiology is uncertain. There is collection of lymphatic cisterns in the deep subcutaneous plane and contraction of smooth muscle cells that line these cisterns causes dilatation and protrusion of lymphatic vesicles from the skin. Cisterns are separated from the normal network (Whimster, 1976). Lymphangiomas may represent vascular malformations during embryonic development rather than as true neoplasms. Vascular endothelial growth factor (VEGF)-C and VEGF receptor-3 are active in the formation of lymphangiomas (Schwartz et al., 2011). The differential diagnosis includes lymphangiectasis, metastatic carcinoma to the skin, verrucae, molluscum, hemangioma, angiokeratoma, or lymphangioendothelioma (Patel et al., 2009).

Conclusion

We would like to emphasize the importance of including lymphangiomas in the differential diagnosis of verrucous lesions of the extremities.

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