VERRUCOUS LYMPHANGIOMA CIRCUMSCRIPTUM IN A CHILD

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ABSTRACT

BACKGROUND

Lymphangioma circumscriptum is characterised by dilation of lymphatic vessels in the skin and subcutaneous tissue. It presents with asymptomatic vesicles filled with lymphatic fluid and characteristic frog spawn appearance. The common sites are axillary folds, shoulder, upper arm, scrotum, penis, rectum and vulva. It needs to be differentiated from certain common conditions like herpes zoster, molluscum contagiosum, haemangioma, verruca vulgaris and angiokeratoma. Surgical modality is the first choice of treatment. A case of 5-year-old girl having verrucous plaque on right knee joint diagnosed as lymphangioma circumscriptum on histopathology is presented here.

KEYWORDS

Lymphangioma Circumscriptum, Verrucous, Lymphatics.

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BACKGROUND

Lymphangioma circumscriptum (LC) is a benign lymphatic malformation characterised by dilatation of lymphatic vessels in the skin and subcutaneous tissue.[1] It was first described by Fox and Fox in 1879 as lymphangiectodes and later on renamed as lymphangioma circumscriptum by Morris in 1889.[2]

It can be congenital or acquired. Approximately, 90% cases are diagnosed before age of 2 years, but can occur at any age. Congenital variety occurs due to malformation of lymphatics, while acquired variety has various causes. Clinically, it presents with asymptomatic vesicles filled with lymphatic fluid and has a characteristic frog spawn appearance. LC may resemble a number of disorders including lymphangiosarcoma, syringocystadenoma papilliferum, lupus vulgaris, actinomycosis, herpes zoster, viral warts and molluscum contagiosum.[3] Diagnosis is mainly by clinical and histopathological examination. Definitive treatment is surgical removal, but various other modalities are also available like sclerotherapy, cryotherapy, laser and electrocautery. We report a case of 5-year-old girl having plaque with verrucous morphology on right knee joint diagnosed as lymphangioma circumscriptum on histopathology.

Case History

A 5-year-old girl presented with elevated lesions over right knee joint since 6 months. History of gradual increase in lesions, which extended to leg as well as adjacent thigh were present. H/O on and off bleeding and oozing was also present after any type of trauma while playing or during fall. No pain or itching from the lesions. No h/o fever or any respiratory complaint was present. On examination, multiple grouped papules, few verrucous forming a plaque of almost 8 x 6 cm size present over right knee joint extending to adjacent lower thigh and leg. [Figure 1] Few discrete papules were seen at the margin with two hypopigmented ill-defined plaques over right thigh. A biopsy was taken from one of the papule keeping tuberculous verrucosa cutis, common warts and lymphangioma circumscriptum as differentials. Histopathology showed hyperkeratosis, moderate acanthosis with presence of numerous dilated lymphatics in superficial and papillary dermis containing many red blood cells and lymphocytes. Marked periappendageal and perivascular lymphoplasmacytic infiltration seen. [Figure 2a & b]which was suggestive of lymphangioma circumscriptum. Patient was advised cryotherapy for the same, but the patient wanted quick response so was referred to Laser Clinic, as we did not have the facility for the same.

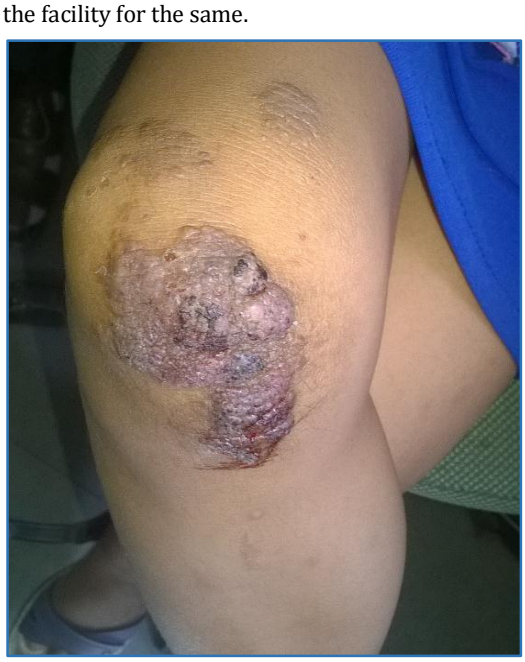

Figure 1. Multiple Grouped Papules, Few Verrucous forming a Plaque over Right Knee Joint

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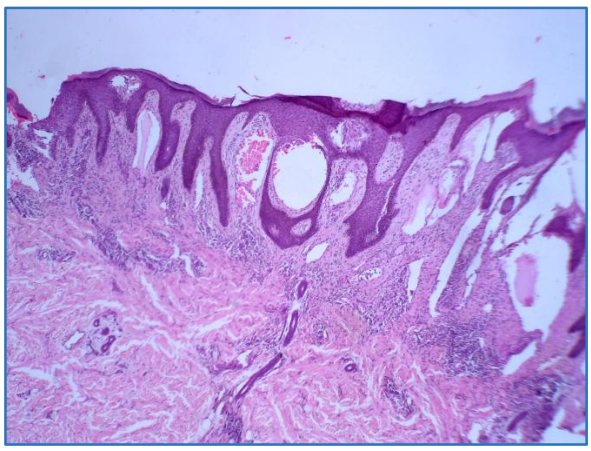

Figure 2a. Hyperkeratosis, Moderate Acanthosis with Presence of Numerous Dilated Lymphatics in Superficial and Papillary Dermis containing many Red Blood Cells (H & E Stain 4x)

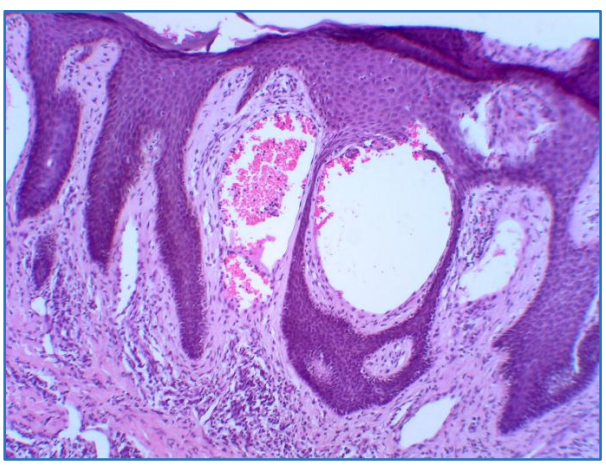

Figure 2b. 10x

DISCUSSION

Lymphatic malformations or lymphangiomas are rare benign hamartomas composed of dilated lymphatic channels filled with a proteinaceous fluid and generally do not have connections to the normal lymphatic system and result from maldevelopment of primitive lymphatic sacs.[4] It is commonly seen over chest, mouth, axilla and tongue.[1] Lymphangioma can occur at any age, but majority are seen in children. Approximately, 50% presents at birth and 90% are diagnosed before the age of 2 years.[1] Lymphangiomas can be divided into lymphangioma circumscriptum, which are superficial cutaneous lesions and cavernous lymphangioma, which are more deep-seated.

LC also known as "capillary lymphangioma," "lymphangiectasia" and "dermal lymphangioma" is a rare benign skin disorder involving hamartomatous lymphatic malformation of deep dermal and subcutaneous lymphatic channels.

Peachey et al[5] classified LC into two main forms: Classic and localised. The classic form usually appears at or soon after birth and involve proximal limbs and is thought to derive from muscular lymphatic cisterns, which failed to segment during embryonic development. They usually appear as vesicular and do not progress into warty plaque. On the contrary, localised form is seen in any age and has no site predilection.

LC can also be divided into congenital and acquired form. Congenital LC results from local malformation of lymphatics and manifest at birth or before 5 years of age, while acquired form occurs secondarily due to obstruction of lymphatics commonly in the vulvar region and can manifest at any age secondary to pelvic surgery, radiation therapy, infection such as tuberculosis, Crohn's disease, etc.[6] The common sites of LS are axillary folds, shoulder, upper arm, scrotum, penis, rectum and vulva.

The exact pathogenesis of LC is as a result of collection of subcutaneous lymph cisterns 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, whereas acquired variety develops due to injury to deep collecting lymphatics caused by radiotherapy damage or infections such as filariasis, lymphogranuloma venereum or tuberculosis in advanced age. The exact aetiology of LC is unknown, but various growth factors such as Vascular Endothelial Growth Factor-C (VEGF-C) and VEGF-D and their receptors on the lymphatic endothelial cells may have a role.[7]

LC is usually asymptomatic. Vesicles that contain lymphatic fluid are compared with frog spawn. Less commonly, it can present as diffuse swelling or with verrucous alterations giving them a warty appearance as was seen in our case and if there is significant hyperkeratosis, the swelling may clinically resemble condyloma acuminate. It can be complicated by excessive drainage and recurrent cellulitis.

Differential diagnosis of LC includes genital warts, herpes zoster, molluscum contagiosum leiomyoma, haemangioma, verruca vulgaris, angiokeratoma and lymphangio-endothelioma.[8] As the clinical presentation of LC may vary from pseudovesicles to nodules or wart-like lesions, correct clinical diagnosis is usually not possible but requires histopathology.

Histology shows dilated lymphatics, which contains red and white blood cells in the epidermis and the papillary dermis lined by flat endothelial cells. Sometimes, the epidermis shows acanthosis and hyperkeratosis with widening of papillary dermis. The deeper dermis shows wide ectatic channels with a lining of endothelium containing lymph.[9]

LC is treated to prevent complications such as cellulitis and for cosmetic reason. The definitive treatment for lymphangiomas is surgical excision. Other treatment options include sclerotherapy, cryotherapy, superficial radiotherapy, pulsed dye laser, intense pulse light, electrocautery, CO2 laser, Nd: YAG laser, electrodessication.[10,11,12] The most common postoperative complication is recurrence of LC with an incidence of 25% to 50% within three months, which is usually due to an improper surgical approach or inadequate excision of the tumour.

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