IJD CASE REPORT

# A case of long-lasting misdiagnosed lymphangioma circumscriptum masquerading as dermatitis

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Lymphangioma circumscriptum is a rare benign lymphatic malformation, clinically characterized by clusters of translucent vesicles. Uncommon manifestations such as verruciform or zosteriform have also been reported in the literature, yet eczematous-like presentation is extremely rare. The present study describes the case of a young woman with a 16-year history of an exudative eczematous-like plaque, mistreated as dermatitis which converted to a semi vesicular lesion during the treatment of super infection and was later diagnosed as lymphangioma circumscriptum by histopathology study. It is suggested that since lymphangioma circumscriptum may resemble other diseases such as dermatitis, histology, as the gold standard for diagnosis, should not be forgotten in the clinical context of this entity.

**Keywords:** lymphangioma circumscriptum, dermatitis, pathology

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## INTRODUCTION

Lymphangiomas are malformations of the lymphatic system located on the skin and subcutaneous tissues. Depending on the size and depth of the lymphatic vessels, lymphangiomas are separated into two categories: the superficial group, containing only lymphangioma circumscriptum (LC), and the deeper group which is comprised of cavernous lymphangioma and cystic hygroma [1]. Of the three types of lymphangioma, lymphangioma circumscriptum (LC) is the most prevalent [2]. Clinically, the lesion presents as a cluster of small, cutaneous, translucent vesicles, which resemble frog spawn [3]. However, myriad cases with unusual presentations have been reported in the literature, mostly mimicking viral skin infections (e.g. herpes zoster, warts and molluscum contagiosum) and tumors. Herein, we describe a case of LC falsely treated as eczematous dermatitis for years due to its atypical clinical presentation.

## CASE PRESENTATION

A 33-year-old female presented with an extensive, exudative, erosive plaque over her left arm, which appeared 16 years earlier and gradually acquired its current size. She complained of pain, bleeding, discharge, and pruritus. The patient claimed that since the onset of her skin lesions, she had visited several dermatologists and had been diagnosed with dermatitis. During this period, she had repeatedly received topical corticosteroid therapy with no improvement occurring in her condition. She had no significant past medical history and denied any use of tobacco or drugs.

 

Physical examination revealed a 20 ×15 cm sized erythematous, erosive plaque covered with crusts and exudates on the lateral side of her arm, extending toward her shoulder as well as axilla (Figures 1 & 2). The involved area was well-defined and accompanied by secondary hyperpigmentation. The remainder of the skin examination was essentially normal, and the overall clinical aspect was suggestive of infected eczematous dermatitis.

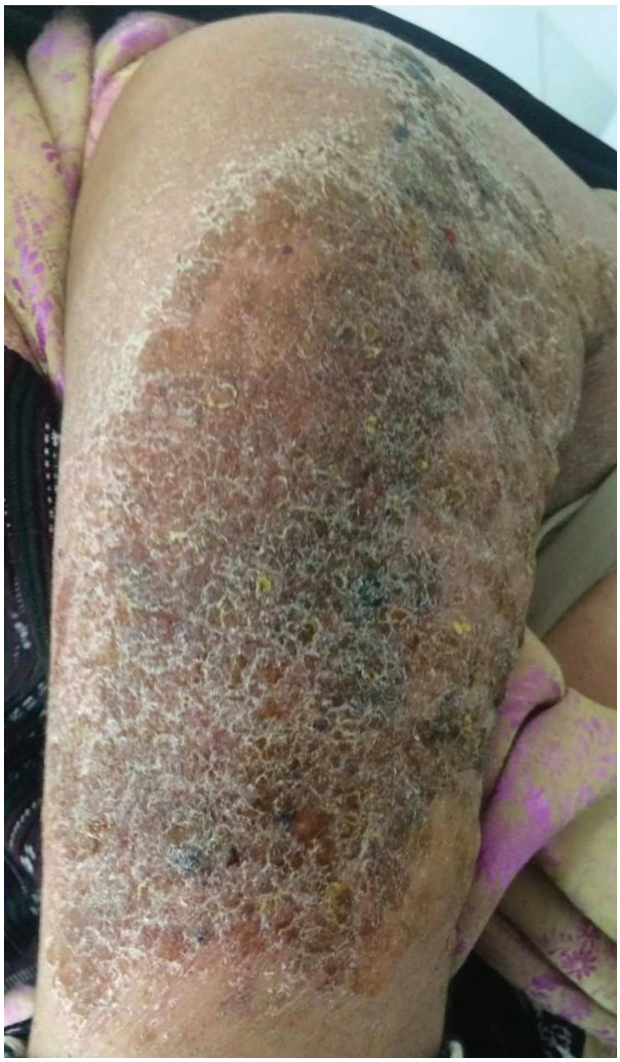

**Figure 1**. Exudative, crusted plaque on lateral side of the arm

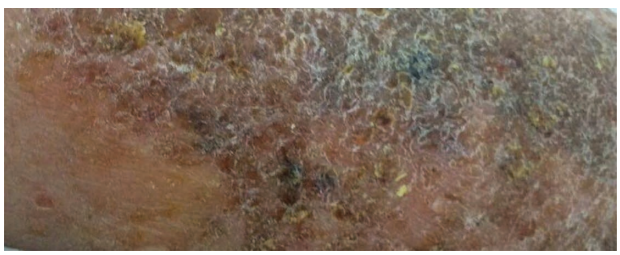

**Figure 2**. Crusted plaque on lateral side of the arm

Exudate culture evaluation was positive for klebsiella. Routine laboratory blood tests were normal except for the elevated values of erythrocyte sedimentation rate. ASO and rheumatoid factor were negative.

Based on the antibiogram, antibiotic therapy was initiated. Interestingly, during the treatment of the infection, the clinical feature of the lesion was transformed to semi vesicular (figures 3 & 4) and areas of hemorrhage appeared on the lesion. This new appearance raised suspicion of lymphangioma circumscriptum, which was later confirmed to be the definitive diagnosis by histopathology.

Histology report revealed grouped ectatic lymphatics located in the papillary dermis. The channels abutted closely on the overlying epidermis and were thin walled, consisting predominantly of an endothelial lining, and vessels containing eosinophilic proteinaceous lymph. These findings were consistent with LC (Figure 5).

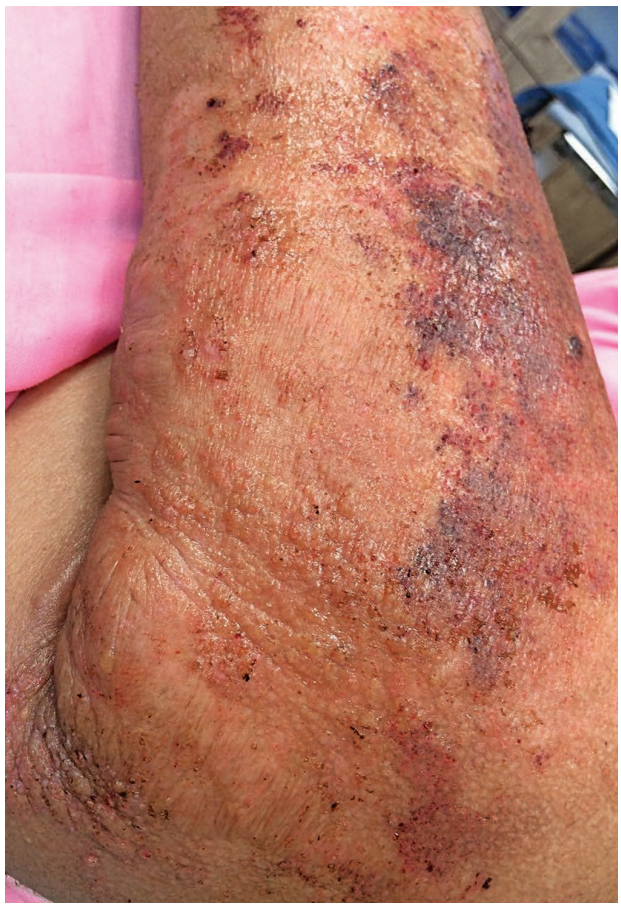

**Figure 3.** The clinical manifestation of the plaque following partial antibiotic therapy

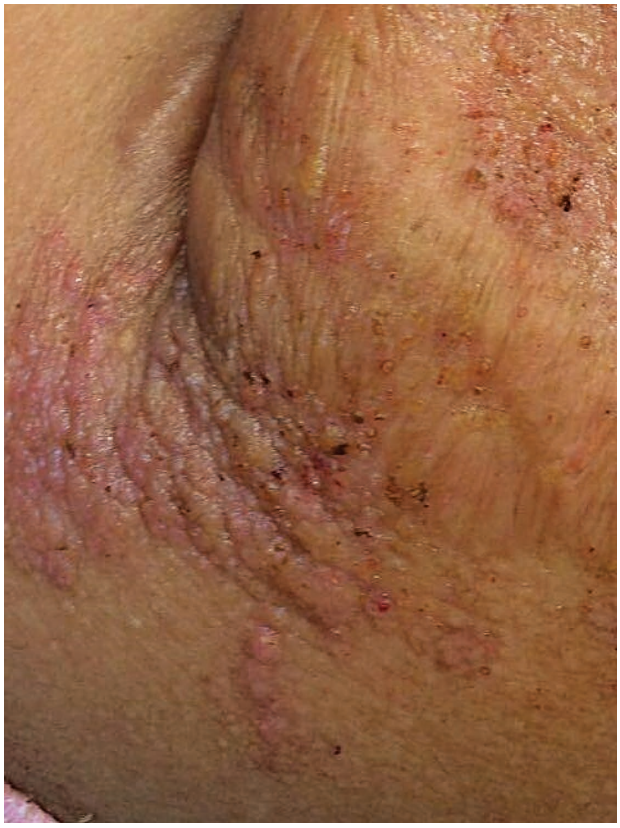

**Figure 4.** The vesicular lesions

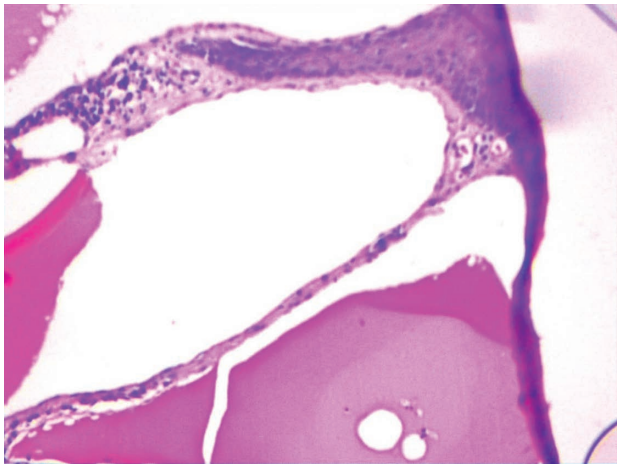

**Figure 5**. Histology of LC showed dilated thin-walled lymphatic channels (H & E stain ×40)

## DISCUSSION

Lymphangioma circumscriptum (LC) was first described by Fox in 1879 as vesicle-like elements in groups as lymphangiectodes [4]. It is now described as hamartomatous malformation of lymphatic channels of the skin [5]. Histologically, it is characterised by multiple dilated lymphatic channels, more often than not, located in the superficial dermis. The condition often extends into the epidermis, which can be acanthotic, and accompanied by a stromal lymphocytic infiltrate [6]. LC is asymptomatic in its localized form, but the most common symptom is oozing of clear fluid mixed with blood, which occurs spontaneously or following minor trauma [7]. The most common complications are bleeding, pain and infection by staphylococcus aureus causing cellulitis. The common sites of involvement are axillary folds, shoulder, upper arm, scrotum, penis, rectum and vulva [7]. The diagnosis of LC is most often made by clinical inspection, yet skin biopsy conduces to differentiation from other vesicular disorders such as herpes infections.

LC may mimic a number of disorders like genital warts, herpes zoster, molluscum contagiosum and even leiomyoma [8]. Reviewing the existing literature, it is evident that most LC case reports describe atypical cases of vulva and scrotum. For instance, Sinha et al presented a case of LC of vulva occurring in a 60-year-old female. The patient presented with multiple genital wart-like papular lesions on the vulva. Biopsy of lesion revealed LC, and she was treated with vulvectomy and no sign of recurrence has been observed since [8].

In 2016, Hassan et al reported a case of LC of buttock mimicking lupus vulgaris. The disease was presented with a red brown plaque, composed of multiple nodules which subsided after the completion of anti-tubercular therapy; however, the disease started spreading 3-4 months following the completion of the treatment [9].

Furthermore, only one case of LC mimicking

**Table 1.** A comparison between Mendiratta et al's case and ours

| Author | Age | Sex | Location | Size | Appearance | Duration | Symptoms |
|---|---|---|---|---|---|---|---|
| Mendiratta et al | 10 | Female | Deltoid region of the left arm | 8×6 cm | A crusted plaque with small deep vesicles | 4 years | Pruritus + oozing |
| Our case | 33 | Female | Lateral side of the left arm | 20×15cm | Erythematous, erosive plaque covered with crust and exudate | 16 years | Pruritus + pain + bleeding + discharge |

dermatitis has been reported in the literature [10]. In 1999, Mendiratta et al described a ten-year-old patient with a crusted plaque over her left upper arm, studded with small deep vesicles initially treated as irritant dermatitis, later identified as lymphangioma circumscriptum via histology. The table below compares our case with Mendiratta's (Table 1).

The late appearance of the lesions and their atypical crusted appearance and associated itching led to the misdiagnosis of dermatitis in both cases.

## CONCLUSION

LC is a benign lymphatic malformation with few complications. However, it poses a diagnostic challenge, the risks of which are misdiagnosis and mistreatment. We presented this case for its atypical presentation to increase diagnostic vigilance. It is to be noted that the diagnosis of LC should be suspected in presumed long-lasting eczema cases with failure to respond to the appropriate corticosteroid treatment.

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**Conflict of Interest:** None declared.

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