

Coexistence of Lymphangioma Circumscriptum and Angiokeratoma in the Same Area: A Case Report

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Abstract

Lymphangioma is a rare benign malformation of lymphatic vessels. Lymphangioma Circumscriptum (LC) is the most common type. Angiokeratoma is a vascular lesion which is considered to be caused primarily by vascular ectasia in the papillary dermis and epidermal changes developing secondarily. Here we presented a 51-year-old patient developing angiokeratoma and LC in the same area with literature's data.

Previously, one case pointing out to the coexistence of LC and angiokeratoma in cutaneous tissue has been reported in the literature. In our case, the lesions' general clinical appearance was consistent with angiokeratoma and dermoscopic examination revealed the findings of angiokeratoma in some areas, histopathologically one of the patients' biopsies specimens revealed angiokeratoma, the other was consistent with lymphangioma.

Coexistence of these two entities can lead to difficulty for diagnosis, histopathological examination is essential for differential diagnosis. Since the late onset of LC can be associated with localized obstructive lymphatic disorders, there is a necessity for the clinician to perform further investigations, establishing the diagnosis of LC has critical importance. Therefore our case points out that distinguishing between LC and angiokeratoma may be challenging for clinicians with only clinicodermoscopic findings, histopathological examination with advanced techniques is mandatory in making a differential diagnosis between these.

Keyword: Angiokeratoma; Lymphangioma circumscriptum (LC); Malformation; Lymphatic disorders

Introduction

Lymphangioma Circumscriptum (LC), is a rare benign proliferation of the lymphatic tissue [1]. Solitary angiokeratoma is an angiokeratoma type developing as a reaction of papillary vein wall to chronic irritation and trauma [2]. Previously, only one case pointing out to the coexistence of LC and angiokeratoma in cutaneous tissue has been reported in the literature [2]. Herein we report a 51-year-old female patient developing angiokeratoma and LC in the same area.

Case Report

A 51-year old female patient presented to the clinic with a red bulging on her right breast which was present since childhood and gradually enlarging during the last two years. Comprehensive anamnesis revealed that the lesion had been scabbing, shedding and bleeding from time to time. The patient had no history of nipple discharge, pain, pruritus, local trauma, surgery or radiation. However, she mentioned recurrent cellulitis attacks during lactation periods. She had no background story of disease and medication. On dermatological examination; localized herpetiform erythematous vascular and maculopapular lesions, ranging from 1 mm and 15 mm

in diameter, were detected on the upper quadrant of the right breast (Figure 1). On dermoscopic examination, multiple clustered red lacunar structures, some with white squams on them, were observed (Figure 2). The systemical examination was normal.



Figure 1: Herpetiform erythematous vascular and maculopapular lesions on the right breast upper quadrant.



Figure 2: Multiple clustered red lacunar structures, some with white squams on them.

A punch biopsy was performed and histopathology was consistent with lymphangioma. Considering angiokeratoma clinically and dermoscopically, two more punch biopsies were executed on two more different areas in the lesion and stained. Histopathologically, thin-walled vascular structures accompanying dilated lymphatic channels were observed and it has been observed that they were stained positively by the lymphatic endothelial marker D2-40 (Figure 3). In the patient's second biopsy, ectatic thin-walled vascular ducts which have been filled with erythrocytes, elongating rete tips partially covering the vascular ducts and slight hyperkeratosis were seen (Figures 4 and 5).

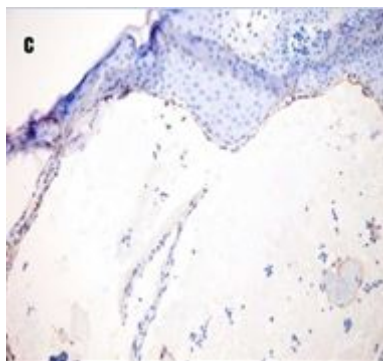


Figure 3: Lymphatic channels were stained positively by the lymphatic endothelial marker D2-40.

The patient was diagnosed as lymphangioma circumscriptum accompanied by angiokeratoma in the light of clinical, dermoscopic and histopathological findings. Concerning the lymphatic disorders in the chest, chest X-ray, thorax tomography, mammography, breast, and axillary lymph node ultrasound were performed and the case was consulted with the department of general surgery. Also, no pathology finding has been detected by the thoracic department during their examination for thoracic mass and tuberculosis.

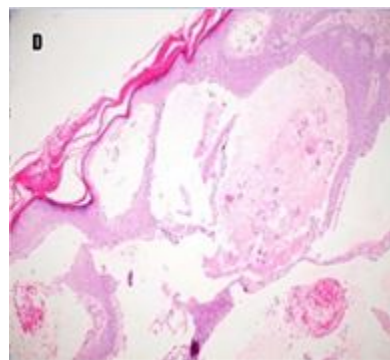


Figure 4: Dilated lymphatic channels in the upper dermis (HE X100).

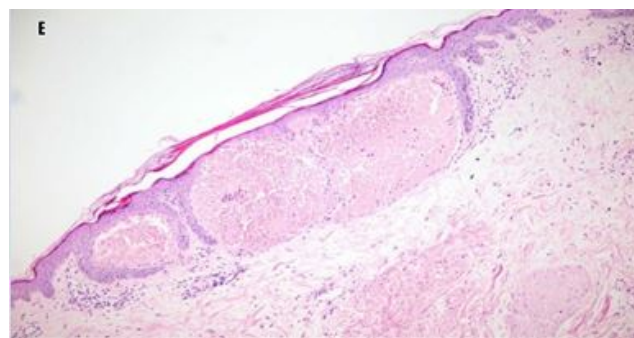


Figure 5: Thin-walled vascular structures accompanying dilated lymphatic channels in the papillary layer of the dermis (HE X100).

Discussion

Lymphangioma is a rare benign malformation of lymphatic vessels. Widened lymphatic ducts with different sizes. Three types of lymphangioma including capillary, cavernous and cystic types exist. LC is the most common type [1,3]. Primary LC is a lymphatic malformation which emerges in the early years of life; secondary LC occurs as a result of the chronic obstruction of the lymph [4,5]. In our case, considering the early disease onset and the absence of an accompanying pathology the patient was diagnosed as primary LC.

Clinically, LC is observed as small sized, thin-walled, grouped pseudo vesicles containing clear liquid. While the liquid contained sometimes might turn to pink because of bleeding in the vesicle, sometimes it might be observed as skin color or black as a result of the coagulation of this blood. Mostly affecting the proximal areas of the extremities, abdomen, femoral, and the thigh; vulva, perineum, scrotum, tongue and cheek mucosa are effected seldomly [3]. While etiology has not been clarified yet, it has been suggested that lymphangiogenic growth factors, vascular endothelial growth factors-C and D, and their receptors might play a role in the development of LC [6].

In the histopathology, wide lymphatic structures surrounded by endothelium has been observed. The differential diagnosis includes angiokeratoma, verrucous hemangioma, verru and melanoma [3,7].

Histopathological examination, staining with lymphatic endothelial markers provide the conclusive diagnosis. Recurrent infections and malignant transformation can occur. While no standard cure exists, electrocoagulation, sclerotherapy, laser, and surgical resection are some of the treatment options [8,9].

Angiokeratomas, are vascular lesions where secondary epidermal changes like hyperkeratosis and acanthosis due to the widening of papillary dermal vessels are observed histopathologically [10]. Angiokeratomas are classified into five categories as Angiokeratoma corporis diffusum, Mibelli's angiokeratoma, angiokeratoma circumscriptum, Fordyce angiokeratoma, and solitary-multiple angiokeratoma. It's considered that vascular ectasia develops first and the epidermal changes occur secondarily as a reaction.

The etiopathogenesis of angiokeratoma is still unknown, it has been suggested that it might stem from the underlying arteriovenous fistula or might develop following a local trauma. In the histopathology hyperkeratosis, acanthosis, vascular dilatation in the papillary dermis and congestion are seen. Among the treatment options are Cryotherapy, laser, and excision [11,12].

We have established the diagnosis of angiokeratoma with coexistent congenital-onset LC in our case. Coexistence of these two entities has been firstly reported by Kim et al. [13] in literature and until now it has been the only case, except for ours, where both LC and angiokeratoma have been observed in cutaneous tissue. In the case reported, solitary angioma and LC has been diagnosed by a biopsy from a dark brown nodule in the right flank area which existed since the early childhood of a 15-year-old male patient and from the surrounding multiple transparent vesicular lesion [2]. In the case, it was suggested that angiokeratoma might evolve in the underlying LC by repeated injuries [14]. Angiokeratoma has been reported to develop overlying an arteriovenous fistula and in areas of LC after local injuries [15]. As for our case, it is possible that these two diseases might have developed independently from one another, as reported before, there might be an etiopathogenic common factor or angiokeratoma might have developed depending upon the chronic trauma based on LC. Recurrent cellulitis attacks of our patient in the area concerned might be another trigger for angiokeratoma. Another remarkable point in our case has been that the lesion was clinically and dermoscopically more compatible with angiokeratoma in monomorphic character. Diagnosis of lymphangioma becomes crucial with regard to the underlying obstructive pathologies.

Conclusion

Our case has been reported because of its rarity and it is the second case report in published data where LC and angiokeratoma have been

observed together. Also, our case points out that distinguishing between LC and angiokeratoma may be challenging for clinicians with only clinical and dermoscopic findings, histopathological examination with advanced techniques is mandatory in making a differential diagnosis between lymphangioma and angiokeratoma. Late onset of LC can be associated with localized obstructive lymphatic disorders and there is a necessity for the clinician to perform further investigations, establishing the diagnosis of LC has critical importance.

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