

Clinical and Dermoscopic Hallmarks of an Acquired Lymphangiectasia Following Surgery and Radiotherapy of Breast Cancer

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Abstract

Acquired lymphangiectasia (AL) is a permanent dilation of lymphatics, which develops after a wide range of scarring processes. The etiopathogenesis of this entity is still unknown but mostly due to lymphatic damage. It was described essentially after surgery and/or radiotherapy for cervical or breast cancers. It is frequently associated with lymphedema. We report an AL in a 45-year-old woman who had undergone a radical mastectomy, radiotherapy, and chemotherapy. After 2 years of combined therapy, she developed multiple multi-colored vesicles and small papules without lymphoedema. Dermoscopy and skin biopsy confirmed the diagnosis of lymphangiectasia. Despite being a harmless condition, it arises worrisome thoughts for the patient and the doctor. Hence a deep comprehension of this entity is important to prompt the right diagnosis and comfort the patient.

Keywords: Breast cancer; Dermoscopy; Acquired lymphangiectasia.

Introduction

Benign and malignant cutaneous vascular proliferation has been reported in the literature due to surgery and radiotherapy for certain malignancies. A wide spectrum of diseases has been described including high-grade angiosarcomas and benign entities such as AL and atypical vascular lesions. Medical history, clinical examination, dermoscopy findings, and immune-histochemistry (e.g., c-MYC, anti HHV8,

Ki69) are essentials for the diagnosis. For instance, some benign lesions like atypical vascular proliferation and benign lymphangioendothelioma are diagnostic dilemmas for pathologists. They may simulate malignancy as patch stage Kaposi sarcoma or well-differentiated angiosarcoma [1]. AL is also called acquired lymphangioma circumscriptum (LC), despite the absence of clinical or histological criteria to distinguish the two of them, some authors believe that the subcutaneous muscle-coated cisternae

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characteristic of LC are absent in AL [2]. In our case, we described the entity as an acquired lymphangiectasia, as the term lymphangioma circumscriptum refers to localized malformation of lymphatics of the superficial dermis arising mostly in infancy while AL is due to the obstruction or destruction of the lymphatic drainage system and has been described as a result of interference of previously normal lymphatics secondary to radiotherapy or surgery [1,3].

Case Report

A 45-year-old woman with a history of infiltrating duct cell carcinoma of the right breast in 2011. She underwent a radical

mastectomy followed by chemotherapy and radiotherapy. She has presented 2 years after the surgery-radiotherapy treatment, multiple grapelike whitish and purple vesicles on the anterior chest. On dermatological examination, on the site of the Y-shaped mastectomy scar, we observed multiple pseudo-vesicles, vesicles, and small papules, some were polypoid, pedunculated, and hypertrophic. The lesions were purple, whitish, flesh-colored, and pink cherry. They were spread within the field of radiation with a metameric disposition and on the left breast. No oozing of fluid from papulovesicular lesions. We have noticed the absence of lymphedema underneath (Figure 1).



Figure 1: Multiple grouped vesicles and small papules, multi-coloured, some are hypertrophic polypoid on the mammary region.

On dermoscopy, some lesions presented whitish and reddish lacunae separated by white lines. We have observed the hypopyon sign. We have noted a light background

color, yellowish, whitish on some lesions while others were red or multicolored. Scales and vascular structures have been observed too (Figure 2).

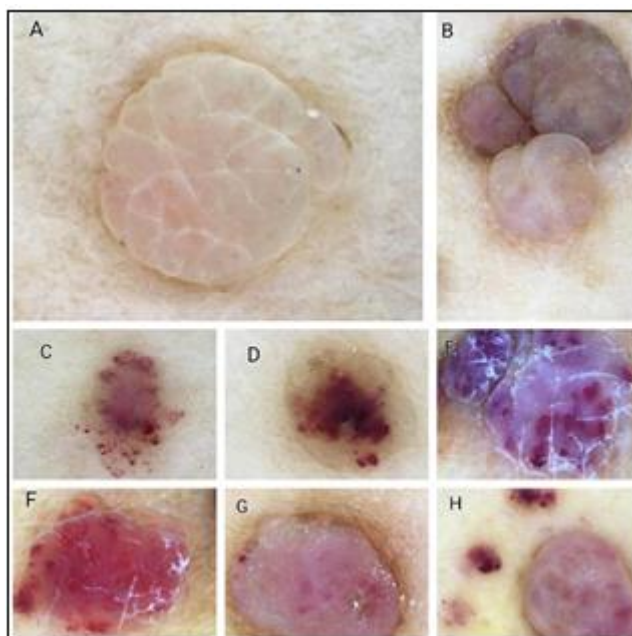


Figure 2: Dermoscopy findings; (a) Whitish-yellow lacunae; (b) White lines multi-coloured lacunae; vascular structures; (c) Vascular structures; (d) Red, dark lacunae, the hypopyon sign; (e) White lines and vascular structures; scales mimicking solitary angiokeratoma; (f) Red lacunae mimicking haemangioma, hypopyon sign; (g) Pinkish lacunae, white lines, (h) Dark lacunae, and vascular structures (some of which are in contact with pinkish lacunae).

The clinical and dermoscopic findings evoked the diagnosis of acquired lymphangiectasia (AL). However, haemangiomas, angiokeratomas, angio-sarcomas, cutaneous metastases were considered in the differential diagnosis. Histopathological examination of a skin biopsy revealed, ectatic lymphatic vessels in the superficial dermis lined by endothelial cells, with mild hyperkeratosis compatible with AL. Immunohistochemistry was positive to anti CD31 and anti-ERG and the patient was treated with a CO₂ laser.

Discussion

Plotnick and Richfield were first to describe AL in 1956, as a complication of radical mastectomy [3]. In a retrospective study, Chiyomaru and Nishigor reported that

external genitalia were the most frequent localization following treatment for malignant neoplasm. They also notified that AL was mostly induced by combined therapy surgery and radiotherapy (RT) in 77% of cases, followed by surgery alone 18% and irradiation alone 5%.

Furthermore, by analyzing the time to onset between completion of therapy and the development of AL, they found that it was shorter after combination therapy (5.8 years) than after surgery (12.2 years) or irradiation (11.8 years) alone [4]. The pathogenesis remains unclear, It's most likely secondary to surgery alone, RT alone, or combined therapy. Surgery induces fibrosis and lymphatic obstruction of the reticular dermis thus an increased pressure by the accumulation of lymph fluid and dilatation

of the superficial lymphatic vessels. RT by targeting the junction of the subcutaneous tissue and reticular dermis causes lymphangiectasia [5]. Moreover, AL without lymphoedema was mostly described as secondary to scarring in scrofuloderma, scleroderma, and cirrhosis [6-8]. Clinically, AL appears as grapelike, small papules, vesicles, or bullae organized in groups and distributed in areas of irradiated skin. The lesions are asymptomatic, thin sometimes thick-walled, translucent, often compared with frog spawn. Hemorrhages within the lesions can create a deep red-purple or black appearance. Rarely, they may appear as wart-like lesions as they may become pedunculated with a hyper-keratotic and verrucous surface. Lymphoedema is usually associated, and trauma may induce infection with a risk of cellulitis [1,3]. Histopathologically, it is correlated to dilated lymph vessels in the papillary and the reticular dermis. The lumina of these vessels show no contents or are filled by homogeneous eosinophilic material. They are lined up by a thin wall composed of a single discontinuous layer of flat endothelial cells and arranged in a “back-to-back” fashion [1,3]. Additionally, the immunohistochemistry of endothelial cells of the neoformed vessels (expresses CD31, but not CD34) and the absence of a peripheral ring of actin-positive pericytes support a lymphatic nature for these neoformed vessels [1,3]. Pedro Zaballo et al reported dermoscopic patterns associated with lymphangioma circumscriptum, which are lacunae and vascular structures in 82% of cases [9]. Different colors of lacunae have been described based on their contents, the lymphatic fluid appears as whitish to yellow,

red blood cells in the dilated lymphatic vessels appear reddish, and thrombosed lacunae can be dark violaceous. The hypopyon sign is a consequence of the sedimentation of blood in ectatic lymphatics giving an aspect of two-tone lacunae [9]. On histopathology, lacunae correspond to dilated vessels in the papillary dermis. The extravasation of red blood cells into lymphatics is still unknown, but micro-shunts may explain this phenomenon thus the presence of vascular structures (red or dark lacunae) in the LC. Other dermoscopic findings were, white lines separating the lacunae secondary to fibroplasia and the presence of scales seen in hyperkeratotic lymphangioma circumscriptum correlated to epidermal hyperplasia [9]. The main differential diagnosis in dermoscopy are haemangiomas and solitary angiokeratomas but the absence of the hypopyon sign and the white or yellowish lacunae permit to dismiss these diagnoses. From our experience, dermoscopy may be labeled to an enhanced non-invasive diagnosis of AL by showing particular hallmarks, thus avoiding the need for skin biopsy and reassuring the patient. Different treatment options have been proposed including surgical excision, cryotherapy, sclerotherapy, electrodesiccation, laser therapy (CO₂, argon, YAG, KTP, PDL) [3]. The combination therapy, like sclerotherapy and radiofrequency [11]. Even though therapeutic options are effective, recurrences are frequent and may be complicated by scarring.

Conclusion

Our interesting findings are the absence of lymphoedema, the presence of lesions beyond the field of irradiation, the short

onset time of AL after combined therapy, and the importance of Dermoscopy features

making this non-invasive instrument a mandatory device in the diagnosis of AL.

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