Acquired lymphangiectasis
associated with a surgery scar from a congenital hemangioma.

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Summary
Acquired lymphangiectasis (AL) occurs due to the blockage of deep lymphatic vessels. The latter is responsible for dilated dermal lymphatic channels that can induce superficial vesicle formation. We report an 11-year-old girl presented to us with a 10-year history of translucent thick-walled vesicles, which was diagnosed as AL according to the typical clinical lesions.

Key words
Acquired lymphangiectasis; surgery scar; congenital hemangioma.

Acquired lymphangiectasis (AL) occurs due to the blockage of deep lymphatic vessels. The latter is responsible for dilated dermal lymphatic channels that can induce superficial vesicle formation. The development of AL occurs secondarily in the areas of the skin affected by the destruction and/or obstruction of lymphatic drainage. No specific histological criteria can be used to differentiate primary lymphangioma circumscriptum from AL, the only difference being the early onset and the lack of external acquired causes in lymphangioma circumscriptum. We report an 11-year-old girl with a 10-year history of translucent thick-walled vesicles, which were clinically diagnosed as AL.

To our knowledge, there are no previous reports of AL associated with a surgery scar from a congenital hemangioma.

Case report
An 11-year-old girl presented to us with a 10-year history of translucent thick-walled vesicles, 1-5 mm in diameter, on her left upper extremity (Fig. 1). She had undergone surgery for a congenital hemangioma on her left axilla and left upper extremity when she was one year old. After the surgery, she had surgery-related scars in both axilla and upper limb.

About a few months after the surgery, the parents noticed that a few vesicles had appeared on their daughter’s left upper extremity near the surgery scar. It was inferred that these lesions had appeared progressively over the previous year. Two months before the examination, the lesions sometimes drained a clear fluid but were otherwise asymptomatic. No significant lymphadenopathy had developed on the axilla or upper extremity.

On dermatological examination, the patient had multiple, 1 to 5 mm, clear to yellowish, thick-walled, grouped vesicles located on her left upper extremity in an area of approximately 5 cm. Moreover, there were also some scattered vesicles, 2-3 mm in diameter, on the left extremity. The content inside the vesicles was a clear fluid. She also had long congenital hemangioma surgery-
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Fig. 1: Acquired lymphangectasias of the left upper limb in an 11-year-old girl, secondary to surgery performed at the age of 1 year to remove a congenital hemangioma.

related scars on her left axilla and medial aspect of the left upper limb.

Although the patient and her parents refused a skin biopsy, a diagnosis of AL was made according to her medical history and the characteristic clinical lesions.

Discussion

Lymphangiomas may be divided, according to Flanagan and Helwig (9), into two forms: superficial lymphangioma circumscriptum and deep lymphangioma cavernosum. These primary lymphangiomas usually present at birth or soon afterwards.

Acquired dilatation of lymphatic vessels are called AL, which represent an acquired dilatation of lymphatic channels secondary to an external cause. No specific histological criteria can be used to differentiate primary lymphangioma circumscriptum from AL. The pathogenesis of these conditions involves the obstruction and/or damage of deep lymphatic vessels that leads to increased lymphatic pressure and dilatation of the vessels. The diagnosis does not present a significant problem, and it is based mainly on the clinical presentation.

AL have been reported in association with repeated trauma (16), surgery, hysterectomy and radiation therapy for carcinoma (5), resection of regional lymph nodes alone (17), pregnancy, scarring from scrofuloderma (7), scleroderma and keloids, Crohn’s disease treated with marsupialization of perianal fistulae (3), malignancies, tuberculosis (4, 15), topical corticosteroid application (2) and recurrent cutaneous infections (12). Lymphangiectasias can also develop a hyperkeratotic surface that mimics warts (8). Pain and oozing may occur after infection.
Minor trauma and solar elastosis from chronic ultraviolet radiation exposure may be etiologic factors in the development of AL; the latter is associated with decreased immune cell trafficking and cell-mediated immunity (14).

We think that the lesions in our patient are acquired and are due to impaired lymph flow, considering in particular their late onset (11) and the fact that in our patient the lymphangiectasia appeared to be associated with a congenital hemangioma surgery scar that may have altered lymph flow and damaged lymphatic vessels, that resulted in the acquired lymphatic obstruction.

Treatment of AL often includes surgical resection of the involved tissue and closure with skin grafting, sclerotherapy (1), cryotherapy, electrodesiccation and/or carbon dioxide laser vaporization (6).

Although the preferred treatment of AL is complete surgical excision, recurrence may occur because of the persistence of deep lymphatic vessels after treatment (10).

Carbon dioxide lasers can not treat deep vessels, but the surface lymphatic vessels are vaporized and communicating channels to the deeper vessels are sealed (13). Moreover, carbon dioxide laser therapy can be used easily under local anesthesia, so the carbon dioxide laser may be a better choice. However, our patient refused any treatment.
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References