A Case of Lymphangiectasia Arising at the Site of an Operative Scar

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Lymphangiectasia(acquired lymphangioma) is characterized clinically by the presence of a circumscribed eruption of thin-walled, translucent vesicles which closely resemble frog spawn in appearance. This rare disorder may arise as a result of acquired lymphatic obstruction secondary to surgery, irradiation, chronic recurrent infection, chronic scarring, or trauma. Herein, we report a case of atypical lymphangiectasia at the site of an abdominal scar in a 70-year-old female. She developed a dark red-colored, pedunculated papule, 2 years after a total abdominal hysterectomy and post-operative irradiation for carcinoma of the uterine cervix.

Histopatholgical findings showed multiple irregularly shaped cystic dilatated cells lined by a single layer of endothelium in the dermis. Immunohistochemical staining with factor VIII-related antigen showed negative results. A lymphangiogram showed signs of acquired lymphatic obstruction. (Ann Dermatol 10:(1) 56~60, 1998).

Key Words : Lymphangiectasia, Acquired lymphangioma

The cutaneous lymphangiomas consist of dilated lymphatic channels that may be found in the superficial dermis and may extend into the subcutaneous fat to the level of deep fascia. These are rare skin lesions, the majority of which are of congenital origin. Occasionally, they arise from obstruction of lymphatic flow from an extrinsic cause[1], and thus they are more appropriately termed lymphangiectasias. Clinically, the most common lesions were circumscribed groups of tense, thin-walled vesicles, often filled with clear fluid.

In the present case, a single dark red-colored, pedunculated papule was developed on the operative scar after total abdominal hysterectomy and post-operative irradiation for carcinoma of the uterine cervix. To our knowledge, this is a rare case showing an unusual manifestation of lymphangiectasia.

Received October 28, 1997.
Accepted for publication December 8, 1997.
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This case was presented at the 49th Annual Meeting of the Korean Dermatologic Association on April 15, 1997.

CASE REPORT

A 70-year-old woman was referred to our OPD for the evaluation of a pedunculated papule on an abdominal scar, in July 1994. Eleven years previously, she had undergone a total abdominal hysterectomy and received post-operative irradiation for carcinoma of the uterine cervix. Three months after the operation, she had developed painless swelling of the lower left extremity and labia majora. Thirteen months later, a cutaneous lesion developed on the abdominal scar.

On physical examination of the abdomen, a dark red-colored, painless 0.5 × 0.5cm sized pedunculated papule was observed on the middle portion of the surgical incision scar (Fig. 1). When the papule ruptured, it exuded clear fluid. Results of the following laboratory tests were within normal limits or negative: a complete blood count,

liver function test, urinalysis, chest X-ray, electrocardiogram, and VDRL/TPHA.

A total excisional biopsy was performed. H&E staining showed a large number of irregularly shaped cystic dilatated cells lined by a single layer of endothelium in the dermis (Fig. 2, A). On immunohistochemical staining, factor VIII-related antigen showed a negative reaction in the dilated endothelial cells (Fig. 2, B). Two years after excision, the skin lesion had not recurred in the same region, but the lymphedema of the lower left extremity had persisted. About three months after excision of the single abdominal skin lesion, numerous translucent papular lesions varying between 2 mm to 4 mm in diameter developed on the labia majora (Fig. 3). There was slight edema of the labia majora. She complained of discharge from her labia majora, and on examination had several tense vesicles, some of which were oozing clear fluid. A translucent papule was biopsied and H&E staining showed elongation of the rete ridge and large irregularly shaped cystic dilatated cells lined by a single layer of endothelium in the upper dermis (Fig. 4, A). On immunohistochemical staining, factor VIII-related antigen showed a negative reaction in the dilated endothelial cells (Fig. 4,

Fig. 1. A 0.5×0.5cm sized dark reddish pedunculated papule in the middle of the incision scar on the lower abdomen.

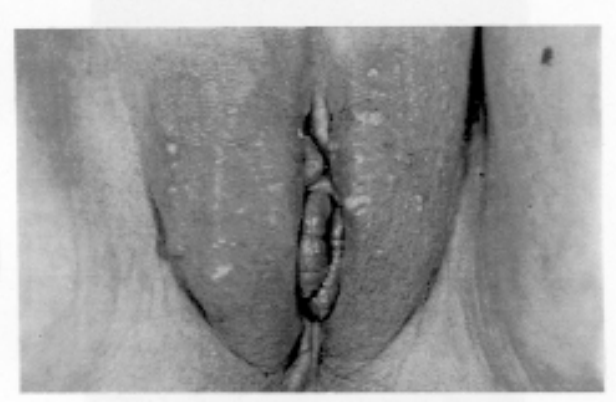

Fig. 3. Three months after excision of the abdominal skin lesion, numerous translucent papules developed in her labia majora.

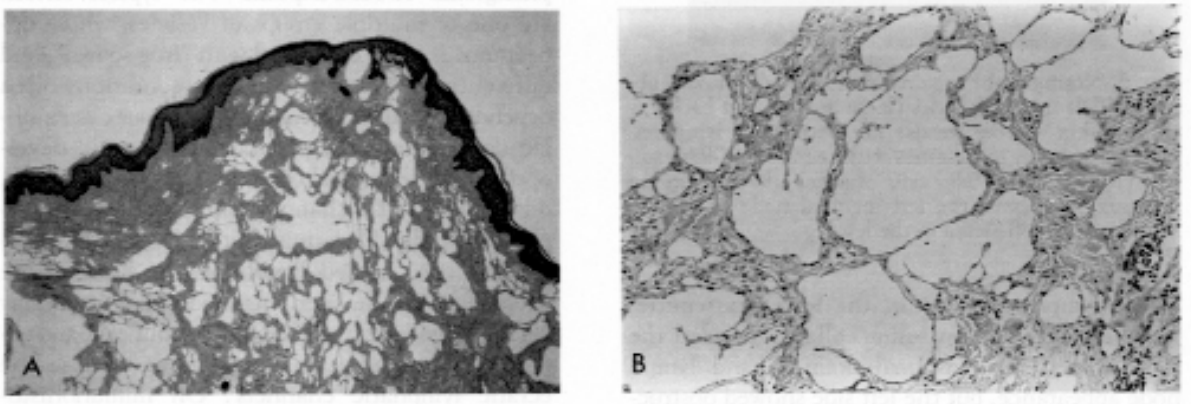

Fig. 2. A biopsy specimen from the abdomen. (A) Multiple cystic lymph vessels are lined by a single layer of endothelium in the dermis (H&E, ×20). (B) Factor Ⅷ-related antigen stain was negative on dilated endothelial cells (Factor Ⅷ-related antigen stain, ×100).

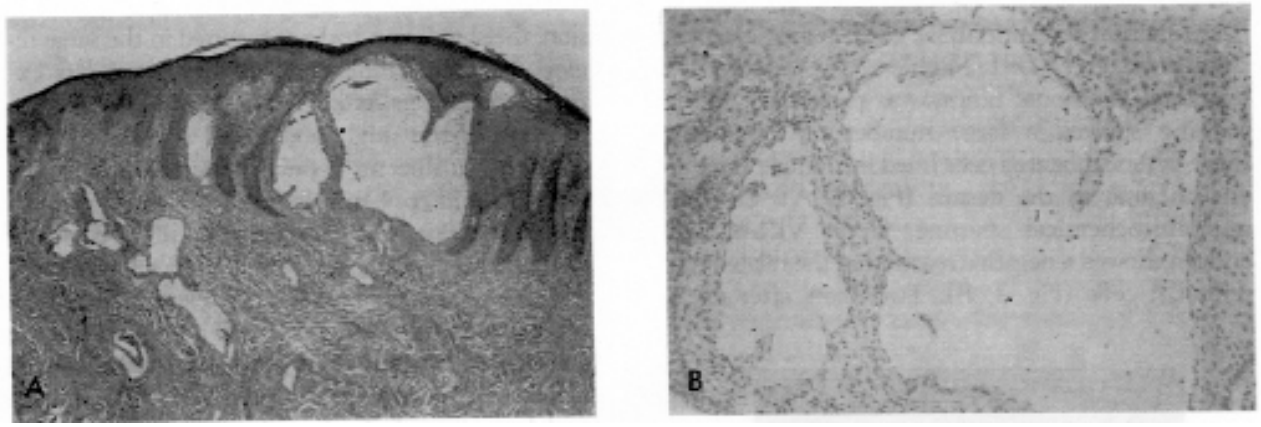

Fig. 4. A biopsy specimen from the labia majora. (A) Elongation of the rete ridge and large irregularly shaped cystic dilatation lined by a single layer of endothelium in the upper dermis (H&E, ×40). (B) Factor VIII-related antigen stain was negative in dilated endothelial cells (Factor VIII-related antigen stain, ×100).

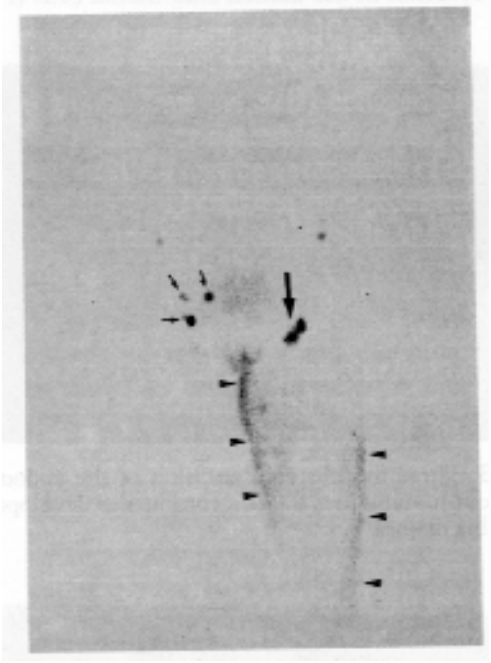

Fig. 5. Normal right inguinal nodes (small arrows), abnormal left inguinal nodes (large arrow), and backflow to the skin (arrow heads) are shown by lymphangiogram of lower extremities with technetium-99m sulfur colloid. The left side showed obstruction in drainage just above the left inguinal node level with significant lymphedema in the left leg.

B). A lymphangiogram of the lower extremities with technetium-99m sulfur colloid, revealed the right side to have normal drainage and lymph node appearance, but the left side showed obstruction in drainage just above the left inguinal node level with significant lymphedema in the left leg (Fig. 5). Thus, the condition was diagnosed as lymphangiectasia of the vulva. We decided to treat the patient using the carbon dioxide laser, but the patient declined any further treatment.

DISCUSSION

Abnormal dilatation of the lymphatics of the skin can be either a primary structural abnormality known as lymphangioma, or a secondary event following lymphatic obstruction, known as lymphangiectasia (acquired lymphangioma). Lymphangioma has been classified by Peachey[2] into classical lymphangioma circumscriptum and localized lesions.

The clinical and histological findings of lymphangiectasias are identical with those of lymphangioma circumscriptum. The typical lesions are one or multiple groups of vesicles, whose appearance is often compared with "frog spawn", and differentiation between the two conditions often depends upon the clinical history[3]. In our case, a single dark red-colored pedunculated papule developed on the abdominal operative scar. Histologically, dilated lymphatic channels are present in the superficial and mid dermis, but only one, or occasionally two, dilated lymphatics are seen in the deep dermis[4]. The overlying epidermis can display varying degrees of hyperkeratosis, acanthosis, and papillomatosis, and it may appear to enclose the ectatic lymphatic channels[4]. On immunohistochemical staining, the endothelial cells in lymphangiomas are usually negative for factor VIII-re-

lated antigen[4]. In our case, factor VIII-related antigen showed a negative reaction in dilated endothelial cells. The lymphangiographic findings in our patient indicated near complete obstruction of lymphatic drainage just above the left inguinal node. There was no history of pre-existing lymphatic abnormality. The accompanying lymphedema probably may have occurred as a result of this lymphatic obstruction.

Lymphangiectasias have been reported after radical mastectomy and radiotherapy[5], radical hysterectomy and radiotherapy[6], thoracotomy[7], keloid[8], scrofuloderma[9], scleroderma[10], penicillamine dermopathy[11], and arthrotomy[12]. Several cases of lymphangiectasia have been reported in the Korean dermatologic literature[13-18]. It is suggested that the vesicles develop as saccular dilatations of superficial lymphatics, secondary to raised intralymphatic pressure due to compromised lymphatic drainage of the involved part[6]. This mechanism would explain the accompanying lymphedema[6]. The present case was associated with considerable lymphedema. Lymphangiectasias have not been reported to undergo malignant change.

The ablative modalities used in the treatment of lymphangiectasia, such as surgery, electrocautery, cryosurgery, and carbon dioxide laser treatment are associated with frequent recurrences unless the deep lymphatic cisterns are adequately treated[19-20]. The carbon dioxide laser has the advantage of being independent from the content of erythrocytes and sealing the lymph vessels at least partially[21,22]. In our case, single lymphangiectasia on the abdomen was successfully excised, and there was no evidence of recurrence at follow-up 2 years later. The vulvar lesions in our patient were complicated by oozing fluid but the patient refused any more treatment. She was to keep her legs elevated in order to reduce the amount of edema.

We consider that the factors contributing to the lymphatic obstruction in our patient were the total abdominal hysterectomy and post-operative irradiation. Herein, we report a case of lymphangiectasia secondary to scarring from a total abdominal hysterectomy and post-operative irradiation for carcinoma of the uterine cervix.

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