

What Is Your Diagnosis?



A 19-year-old woman presented with an umbilical mass of 5 months' duration that had grown in size. Physical examination revealed a 1×1-cm brownish, pedunculated, cauliflower-shaped lesion on the umbilicus. There were no other signs or symptoms of disease. The patient's personal and family disease history were unremarkable. An excisional biopsy was performed.

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The authors report no conflict of interest.

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The Diagnosis: Acquired Lymphangiectasia

On histopathology numerous dilated channels lined by a single flat layer of endothelial cells were noted within the dermis. The overlying epidermis was papillomatous and acanthotic (Figure 1). The endothelial cells lining the dilated channels were D2-40 positive (Figure 2). Furthermore, the channels contained a pinkish amorphous material and a few red blood cells. The surrounding stroma showed scattered lymphocyte infiltration. These findings were consistent with lymphangiectasia. The lesion has not recurred 4 years following total excision.

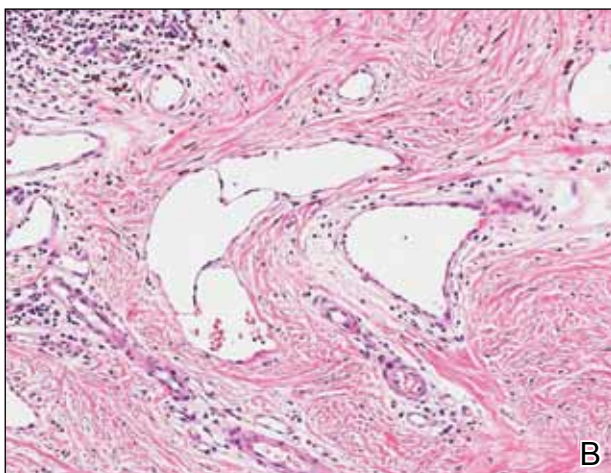
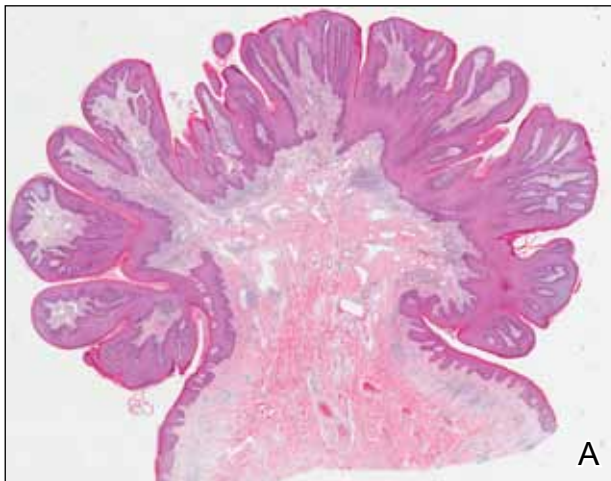


Figure 1. Numerous dilated channels lined by flattened, single-layer endothelial cells in the dermis with overlying epidermal papillomatosis and acanthosis (A and B) (H&E, original magnification $\times 40$ and $\times 200$).

Acquired lymphangiectasia is known by various names, including lymphangioma, acquired lymphangioma, and acquired lymphangioma circumscriptum, which has led to confusion.¹ Acquired lymphangiectasia, which is characterized by dilated superficial lymphatics, develops following damage to previously normal lymphatic channels, leading to a buildup of lymph pressure and backflow.² Acquired lymphangiectasia has been reported as clinically and histologically indistinguishable from lymphangioma circumscriptum²; however, unlike in lymphangiectasia, the suffix *-oma* denotes a tumor. Our case matched more closely with the typical concept of lymphangiectasia rather than lymphangioma.

Clinical findings of acquired lymphangiectasia usually include translucent, flat or slightly raised, 2- to 5-mm, flesh-colored papules and vesicles.^{3,4} Acquired lymphangiectasia has been described with lesions that have verrucous surfaces mimicking warts, condyloma acuminata, or molluscum contagiosum.^{5,6} Our case suggests that acquired lymphangiectasia also can present with a pedunculated cauliflowerlike appearance. In general, it develops

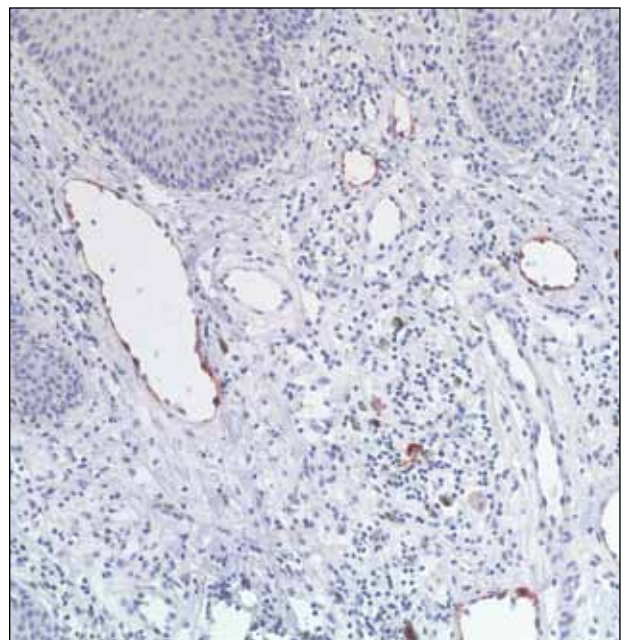


Figure 2. The flattened, single-layer endothelial cells that lined multiple dilated channels were positive on D2-40 immunochemical staining (original magnification $\times 200$).

secondary to certain conditions such as recovery from trauma or surgery, postsurgical fibrosis, and irradiation. Lymphangiectasia often is seen on the arms, axillae, chest wall, and genital area in women and the scrotum, penis, thighs, and pubic region in men, both who have undergone radical surgery and irradiation for treatment of breast and prostate cancer, respectively.³ Our patient did not report any history of trauma to the umbilicus.

On histopathology acquired lymphangiectasia typically shows edematous polypoid nodules with dilated lymphatics. The overlying epidermis usually shows a spectrum of proliferation ranging from mild acanthosis to florid pseudoepitheliomatous hyperplasia with marked hyperkeratosis and parakeratosis. The distinctive finding of lymphangiectasia is the presence of dilated lymphatic spaces within the dermis. The dilated channels are filled with lymphatic fluid and often red and white blood cells. The single layer of flattened endothelial cells generally exhibits immunoreactivity to D2-40 and CD31.¹

Treatment of lymphangiectasia is focused on reducing the pressure within the lymph vessels and managing consequent lymphedema with compression dressings. Simple surgical excision of lesions on sites such as the vulva or legs often is effective.³ If surgical intervention is not an option, cryotherapy, sclerotherapy, cauterization, and treatment with CO₂ lasers also have been utilized with good outcomes.⁷ In the current case, total surgical excision was performed, which provided good results.

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