# Benign Lymphangioendothelioma Manifested Clinically as Actinic Keratosis

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*Benign lymphangioendothelioma is an acquired lymphangiectatic lesion that must be recognized and differentiated from angiosarcoma, early Kaposi's sarcoma, and lymphangioma circumscriptum. We report the case of a 68-year-old woman with the clinical presentation of a possible actinic keratosis and the typical histologic findings of benign lymphangioendothelioma and an overlying actinic keratosis.*

Benign lymphangioendothelioma is a recently described acquired lymphangiectatic lesion. Clinically, it usually appears as a dull pink to reddish brown macule or plaque.[1] We report a case in which the clinical presentation was possible actinic keratosis, with typical histologic findings of benign lymphangioendothelioma and an overlying pigmented actinic keratosis. Recognition of this entity is vital because the histologic differential diagnosis includes angiosarcoma, early Kaposi's sarcoma, and lymphangioma circumscriptum.

## Case Report

A 68-year-old white woman in generally good health presented with a small, light brown patch on the extensor surface of her right forearm that grew radially over a 2-year period. The lesion was asymptomatic, but the patient was concerned about cosmesis and requested that it be removed.

The patient had no history of malignant or premalignant lesions of the skin. For one year, she had had asymptomatic white patches on the mucosal surface of her lower lip; a biopsy demonstrated that the patches were leukoedema.

Cutaneous examination revealed tan skin, and her right extensor forearm had a 2.4×1.0-cm light brown patch with a slightly rough texture (Figure 1). Clinically, a pigmented actinic keratosis or lentigo was suspected. A punch biopsy specimen showed delicate, thin-walled, endothelium-lined spaces and clefts in the upper dermis, with an overlying pigmented actinic keratosis (Figure 2). These vascular channels ran parallel to the epidermis and contained no or few erythrocytes in their lumina. The endothelial cells outlined collagen bundles. Furthermore, there was no erythrocyte extravasation, hemosiderin deposition, or significant inflammation, and no abnormal muscular lymphatic vessels were detected. Ulex europaeus agglutinin I (UEA-I) stained both the lesion and the normal dermal blood vessels. On the other hand, von Willebrand factor antigen (vWF:Ag) stained normal dermal blood vessels but not the lesion vessels. The lesion was fully excised, and the patient was stable 1 month after the procedure.

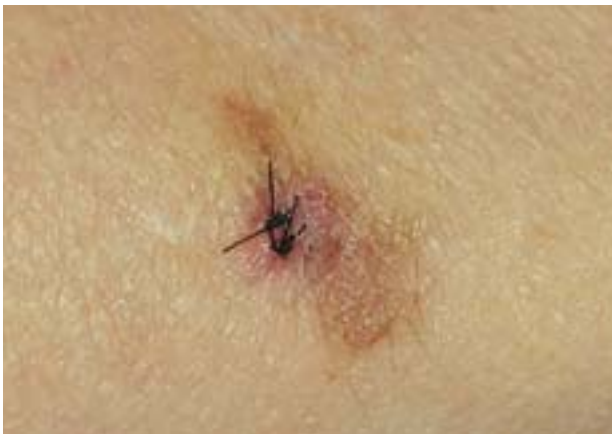

**FIGURE 1.** Benign lymphangioendothelioma at extensor forearm after punch biopsy.

## Comment

Benign lymphangioendothelioma was described in 1990 by Jones et al[1] as an acquired lymphoendothelial lesion. The histologic findings of the lesion in our case are similar to those described by Jones et al, except that the overlying epidermis in our case con-

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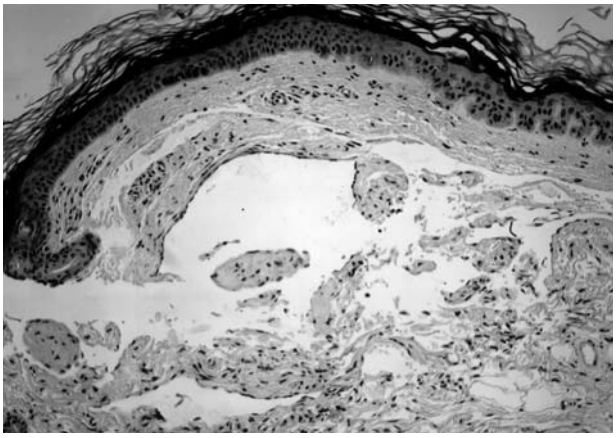

**FIGURE 2.** Delicate, thin-walled, endothelium-lined spaces and clefts in upper dermis. Note also overlying actinic keratosis (H&E, original magnification ×160).

tained a pigmented actinic keratosis. This epidermal change was consistent with the clinical diagnosis of a pigmented actinic keratosis or lentigo. Thus, the clinical appearance of our patient's lesion was different from the pink or red-brown color noted by Jones et al. The lesion was, however, in a typical location—on an extremity.

Lymphangioendothelioma is an important entity to identify because of its histologic and clinical resemblance to angiosarcoma, early Kaposi's sarcoma, and lymphangioma circumscriptum.[2,3] Clinically, angiosarcoma usually appears on the face or scalp of elderly patients[4]; histologically, it has a much greater degree of nuclear atypia and increased mitotic activity. Results of staining with UEA-I and vWF:Ag in lymphangioendothelioma and angiosarcoma are usually similar and therefore offer little help in differentiating the 2 entities.[1]

The early patch stage of Kaposi's sarcoma may clinically resemble lymphangioendothelioma, but the latter is usually solitary and more static. Histologically, both show lymphangiomalike cell dissection of collagen, and early Kaposi's sarcoma may have little associated cell atypia or spindle cell proliferation. Extravasated erythrocytes and hemosiderin are generally present in Kaposi's sarcoma but are absent in lymphangioendothelioma. Similar to lymphangioendothelioma, early Kaposi's sarcoma has vascular channels that are positive for UEA-I and negative for vWF:Ag.[5] Therefore, histologic differentiation between lymphangioendothelioma and early Kaposi's sarcoma may be impossible without clinical correlation.

Typically, lymphangioma circumscriptum is characterized by vesicular, clear, or blood-filled lesions in a single region, with gradual development of peripheral lesions. Histologically, lymphangioma circumscriptum usually has large and dilated endothelium-lined spaces, with a deeper component showing muscular lymphatics.[6] Lymph vessel proliferation with collagen dissection is minimal or absent in lymphangioma circumscriptum.

Benign lymphangioendothelioma can be cured by surgical excision, in contrast to the poor prognosis for diseases such as angiosarcoma and Kaposi's sarcoma.[1,7,8]

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