Lymphangioma Circumscriptum of the Vulva

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Lymphangioma circumscriptum is a benign disease of the lymph ducts and an unusual pathologic process that rarely affects the vulva. The etiology of this lesion is not clear, but obstruction of the lymph vessels has been suggested as a possible cause in some cases. We report the case of a 44-year-old woman with lesions similar to lymphangioma circumscriptum of the vulva and chronic idiopathic lymphedema of the lower right limb. Because there was no obvious cause, we propose that the lymphangioma was caused by the lymphedema.

Lymphangioma circumscriptum was first described in 1878 by Fox and Fox,[1] who called it *lymphangiectodes;* Morris[2] first used the term *lymphangioma circumscriptum* in 1889. Pathological features were described by Noyes and Török[3] in 1890 and by Francis[4] in 1893. Peachy et al[5] suggested the connection of the superficial vesicles with deep lymphatic cisterns. Lymphangioma circumscriptum is clinically characterized by thin-walled, translucent vesicles concentrated in groups most commonly located on the axila, chest, mouth, and tongue. Presentation in the vulva is unusual. The etiology is not completely clear, but the obstruction of lymph vessels or lymphedema alone has been suggested as a possible cause in some cases.

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Case Report

A 44-year-old woman who had vesicular lesions on the right labium majus for 6 years presented with lesions that produced a slight itch and, occasionally, clear exudate. The lesions had slowly grown in number and size over the years, and there was a lymphedema in the lower right limb that appeared 3 years before the vulval lesions. The patient had no history of sexually transmitted disease, trauma in the area, or cellulitis in the affected limb. Vascular surgeons could not find the cause of the lesions.

Clinical exploration revealed a variety of small, colorless vesicular lesions on the right labium majus (Figures 1 and 2), which released a clear liquid when punctured. The rest of the physical examination was normal.

A histopathologic examination of a biopsy of one of the lesions showed slight epidermal acanthosis and enlarged lymph vessels in the upper dermis (Figure 3). Posterior analyses, including blood and urine tests, a culture of the liquid exuded from the vagina, and chest radiography, were normal. Serologic tests for human immunodeficiency virus, cytomegalovirus, herpes simplex, and syphilis (using the Venereal Disease Research Laboratory test and the *Treponema pallidum* hemagglutination assay) were negative. Cytology of the cervix showed inflammation without evidence of dysplasia, and abdominal and pelvic echography did not offer data of interest.

A lymphography of the affected limb was performed to determine whether a relationship existed between the right leg lymphedema and the vulval lesions. It showed the access of the tracer to the venous circulation and its accumulation in 2 ganglionic structures of the popliteal space of the right

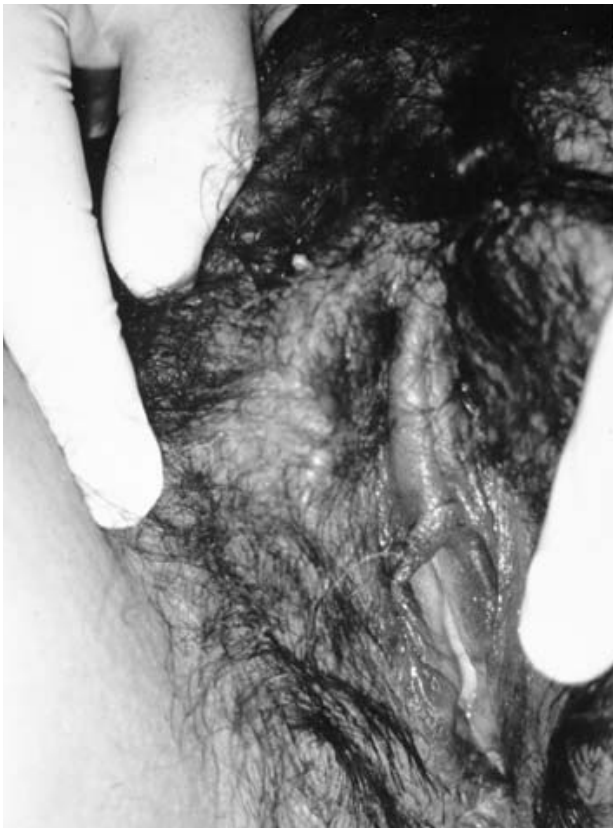

Figure 1. Small translucid vesicles on the right labium majus.

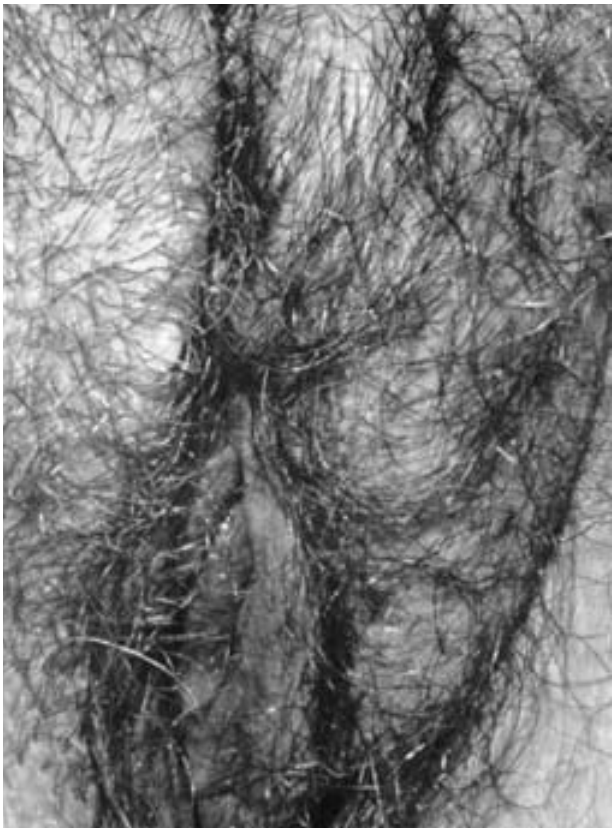

Figure 2. Clinical image showing another group of the vesicles.

knee. We conclude from these data that the lymphangioma was either acquired or lymphangiectasis and was probably secondary to the leg lymphedema. However, it is impossible to prove any cause for the lymphatic obstruction.

Comment

Lymphangioma lesions are characterized by a local eruption of isolated or grouped translucid, thin-walled vesicles filled with a clear liquid. Hyperkeratosis is found incidentally. Histopathologic examination shows enlarged lymph vessels in the dermis connected to deep lymphatic cisterns surrounded by a layer of muscular fibers. Nuclear magnetic resonance has improved the knowledge of the anatomy of the lesion and the extent of the area affected and provides valuable information about damage to the sebaceous tissues.[6]

The complications of lymphangioma include a clear exudate, secondary infections, and minor hemorrhages, and a case of a squamous cell carcinoma on lymphangioma lesions has been described.[7] The appearence of lymphangiosarcoma is discussed in literature,[7,8] but it appears to be related to the presence of lymphedema that has evolved over a long period.

The classification of lymphangioma, considered to be a developmental defect that is restricted to the lymph vessels of the dermis, has been controversial over the years. In a review of 65 cases, Peachy et al[5] describe the 2 main forms as classic and localized, while Lever and Schaumburg-Lever[9] recognize 4 types: localized circumscriptum, classic circumscriptum, cavernous, and progressive. The lesions of classic lymphangioma present at birth or early in life and show predilection for the proximal part of the limbs and areas adjacent to the limb girdles. In contrast, localized lymphangioma appears at any age and has no predilection for particular sites. Lever and Schaumburg-Lever[9] describe lesions of lymphangiectasis, which are indistinguishable from classic circumscriptum and present when local lymph vessels become obstructed. This lesion was also studied in 1970 by Fisher and Orkin,[10] who termed it *acquired lymphangioma* or *lymphangiectasis*.

The vulva is a rare location for lymphangioma. Of the 13 cases of vulval lymphangiectasia we found in the literature, 8 were related to surgery and/or radiotherapy of cervical tumors,[10-16] 2 to lymph node tuberculosis,[17,18] 2 to Crohn's disease,[12] and 1 to recurrent cellulitis.[19] We found only 6 cases of lymphangioma circumscriptum without evidence of secondary

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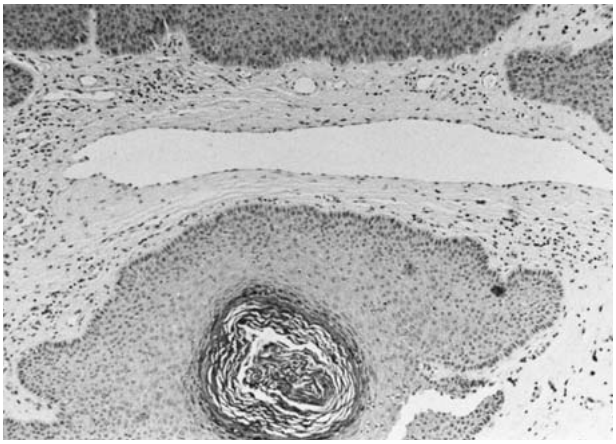

Figure 3. Slight epidermal acanthosis and an enlarged lymph vessel lined by a single layer of cells in the upper dermis (H&E, original magnification ×40).

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lymphatic damage.[8,20-23] In our case, the lymphedema of the lower limb may have played a pathogenic role in the appearence of the vulval lesions, but we could not find any cause of the lymphatic obstruction, and none of the aforementioned antecedents existed.

Although diagnosis is not difficult if clinical data are clear, the presence of hyperkeratosis may make it difficult to distinguish between lymphangioma and genital warts.[16] Other diseases, including molluscum contagiosum, tuberculosis verrucosa cutis, venereal lymphogranuloma, and filariasis, should also be considered in the differential diagnosis.

Lymphangioma circumscriptum is treated for cosmetic reasons, when there is recurrent infection, or when a persistent exudate of lymphatic liquid or blood exists. Many different types of therapy, including electrocoagulation, radiotherapy, cryosurgery, surgical extirpation, argon laser surgery, and carbon dioxide laser, have been used but are associated with a high degree of recurrence unless the deep lymphatic cisterns are correctly treated. Carbon dioxide laser may produce the best results; although the deep cisterns are not treated in this technique, the superficial lymph vessel is pulverized so that the deep cisterns are sealed.[11,24]

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