Benign Lymphangiomatosus Papules and Plaques After Radiotherapy

T Martín-González,a A Sanz-Trelles,b J del Boza, and E. Samaniegoa

To the Editor:

Lymphangiomas are tumors that normally appear at birth. They are formed from dilated lymph vessels that may extend to the subcutaneous cellular tissue. A number of causes of acquired lymphangiomas such as radiotherapy and surgery have been reported. The area irradiated during radiotherapy may develop benign vascular proliferations such as acquired progressive lymphangioma or malignant processes such as high-grade angiosarcoma, even when low doses of radiation are used.1,2

Within what are considered acquired blood cell count of 8200 cells/mL, 70% neutrophils, and left shift (7% band forms). Globular sedimentation rate was 25 mm/h and the chest x-ray revealed no significant abnormalities. Histology of a biopsy sample taken from a lesion on the forearm showed the presence of a subepidermal blister with intense neutrophilic inflammatory infiltrate, but no signs of leukocytoclastic vasculitis. Serology was negative for syphilis, herpes simplex, and hepatitis B and C, but positive for HIV-1 in an enzyme-linked immunosorbent assay. This finding was confirmed in a second test. The skin lesions improved after initiating treatment with a tapering course of oral prednisone (beginning at 50 mg/d) for 6 weeks, with no subsequent relapse. The patient was referred to the infectious diseases department of the hospital, with initial analysis showing a low CD4+ T-cell count (285 cells/mL) and a viral load of 100 000 copies/mL.

The association of Sweet syndrome with HIV infection has rarely been described in the literature,2-7 and only twice has Sweet syndrome been reported as the first manifestation of HIV infection.2,5 The CD4+ T-cell count in the cases described varied between 368 and only with HIV infection has rarely been described in the literature.2-7 and only twice has Sweet syndrome been reported as the first manifestation of HIV infection.2,5

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References

lymphangiomatous lesions, benign lymphangiomatous papules after radiotherapy have specific characteristics.

Our patient was a 54-year-old woman with a history of stage T2 N1b M0 infiltrating ductal carcinoma in the right breast, diagnosed in 1998, and treated by tumorectomy and right axillary lymphadenectomy, chemotherapy, hormone therapy, and external radiotherapy of the entire chest wall and right breast at a dose of 50 Gy. She attended our clinic for the evaluation of progressive asymptomatic lesions on the irradiated skin of the right breast. The lesions had appeared approximately 1 year earlier (6 years after receiving radiotherapy).

Physical examination of the right breast showed yellowish skin coloring. This skin was covered with multiple erythematous papules that coalesced in places to form small vesicular plaques and lesions filled with clear or occasionally bloody fluid (Figure 1). There was no associated lymphedema.

Biopsy of one of the vesicles revealed marked vascular dilation in the papillary dermis that extended into the epidermis (Figure 2). As the vessels penetrated deeper into the dermis, they got narrower and more irregular and tortuous, and took on a lymphatic appearance (Figure 3).

The vascular spaces were covered by a single discontinuous strand of endothelial cells with oval hyperchromatic nuclei that protruded towards the lumen and that coalesced to form small plaques. Typically, the vascular lumen was empty, as is the case for lymphatic vessels, although at times a proteinaceous material and some red blood cells could be found, as well as endothelium-lined papillary projections. The endothelial cells were strongly positive for CD31 and CD34, as well as for D2–40, a marker specific to lymphatic vessels. In accordance with these clinical and histologic findings, we diagnosed lymphangiomatous papules and plaques after radiotherapy.

In 1994, Finenberg and Rosen described benign vascular proliferations in the skin of the breast and chest wall after postoperative radiotherapy for breast cancer. Over the last 20 years, a range of terms have been used to describe these lesions, such as atypical vascular lesions, acquired lymphangiectasis, progressive acquired lymphangioma, circumscribed lymphangioma, and benign lymphangiomatous papules. Díaz-Cascajo et al proposed the term benign lymphangiomatous papules following radiotherapy for benign skin lesions that show a predominance of lymphatic vessels and that present clinically as erythematous papules related to radiotherapy treatment.

In recent years, new cases of this condition have been described. All patients have been women aged between 33 and 72 years, and the most common primary tumor has been breast cancer. In all cases of breast cancer, the primary tumor was removed by partial or radical mastectomy, and all patients received external postoperative radiotherapy at doses between 46 and 50 Gy.

The lesions present as erythematous papules measuring less than 1 cm in diameter and that coalesce to form small plaques. Sometimes, as was the case in our patient, vesicles may be present. The latency period between radiotherapy and the onset of the first lesions is long, between 3 and 20 years. Characteristically, the patients do not report associated symptoms or present lymphedema.

Histology reveals marked vascular dilation in the papillary dermis that may extend into the epidermis, thereby giving the lesion a tuberous appearance. The vascular lesion is relatively well circumscribed, although not encapsulated, and may reach the deep dermis and occasionally extend to the subcutaneous cellular tissue, although this happens more often with malignant tumors such as angiosarcomas.

The vascular spaces are covered by a single discontinuous thread of endothelial cells that may have flat or large, oval nuclei that are hyperchromatic and that protrude towards the lumen. Occasionally, small nucleoli may be present. Mitotic and atypical cells are not present.

In summary, we believe that, in agreement with Díaz-Cascajo et al, lymphangiomatous papules and plaques after radiotherapy are a variant of acquired lymphangioma. Likewise, the term benign lymphangiomatous papules...
or plaques after radiotherapy is the most appropriate because it makes reference to the clinical presentation of the lesions, their nature, and their relationship with radiotherapy.

References