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CASE FOR DIAGNOSIS

Papules and Pustules on the Knee of a Young Woman

Pápulas y pústulas en la rodilla de una mujer joven

Medical History

A 22-year old woman with no relevant past history attended the emergency dermatology clinic for tender papular lesions that had been present on the left knee for 2 months. She had experienced mild fever and joint pain in the 2 previous days. Her primary care physician had provided treatment with corticosteroids and topical fusidic acid, oral cloxacillin, and a tapering course of deflazacort (initial dose, 30 mg), with no improvement.

Physical Examination

Physical examination revealed between 5 and 7 polymorphic lesions on the left knee, mostly papules of 0.5 to 1.5 cm in diameter, on an erythematous base (Figure 1).

Histopathology

The skin biopsy showed a dense polymorphic inflammatory infiltrate including a number of large cells with large nuclei and prominent nucleoli. Direct immunofluorescence was negative.

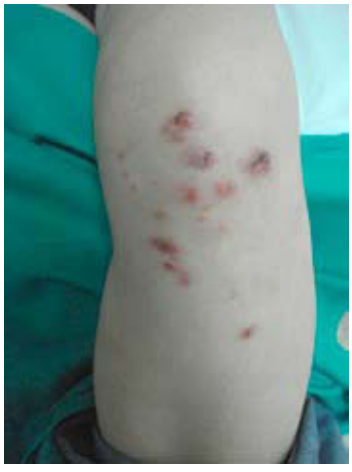

Figure 1

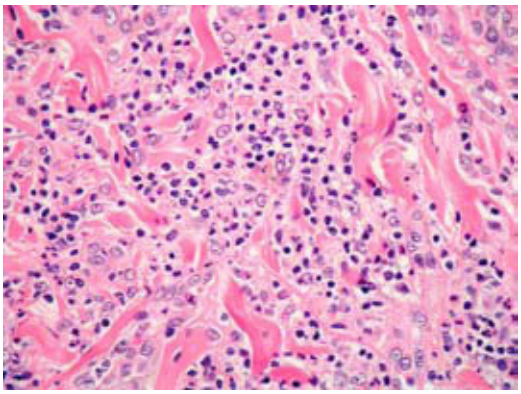

Figure 2 Hematoxylin-eosin, original magnification, 200.

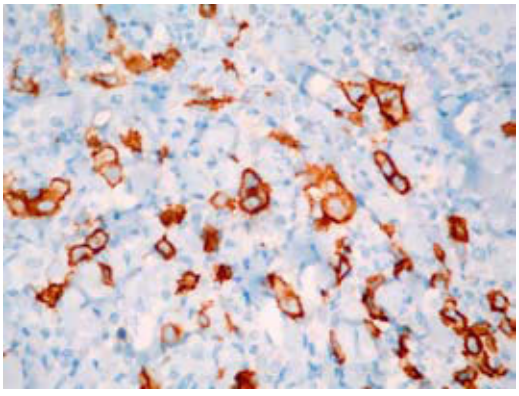

Figure 3 CD30, original magnification, 400.

Additional Tests

Standard tests were requested including liver and kidney function, erythrocyte sedimentation rate, C-reactive protein, antinuclear antibodies, antineutrophil cytoplasmic antibodies, immunoglobulins, complement, protein electrophoresis, urine sediment, and a culture of exudate from a vesicle. All the results were normal or negative.

What Is Your Diagnosis?

Diagnosis

Localized lymphomatoid papulosis (LyP) type A.

Clinical Course and Treatment

The biopsy result was followed by a more exhaustive clinical examination that ruled out the presence of other skin tumors, enlarged lymph nodes, or enlargement of the spleen. Genome analysis revealed no clonal T-cell receptor (TCR) gene rearrangement. In accordance with the protocol used in our unit,[1] further diagnostic tests were requested to study lymphoma spread, including: chest x-ray; Epstein-Barr virus, cytomegalovirus, Borrelia, and human immunodeficiency virus serology; biochemistry with lactate dehydrogenase and β2-microglobulin; peripheral blood smear; and flow cytometry. These all gave normal or negative results. The lesions had resolved spontaneously 1 month later, leaving a residual purpuric pigmentation. There was no recurrence.

Comment

Primary cutaneous CD30+ lymphoproliferative syndromes form a heterogeneous clinical-pathological spectrum with LyP and CD30+ anaplastic large cell lymphoma at the 2 extremes and a range of intermediate or borderline cases in between.[2] CD30 antigen is the characteristic marker of CD30+ lymphoproliferative syndromes, but certain inflammatory responses (reactions to insect bites, toxicoderma) and viral infections, as well as other hematological and nonhematological neoplasms, can present CD30+ cells and form "CD30+ pseudolymphomas" that must be differentiated from LyP.[3]

LyP is a chronic disease that occurs in outbreaks, with the appearance of asymptomatic, frequently ulcerated papules that spread over the trunk and limbs. Lesions in different stages commonly coexist. A negative TCR gene rearrangement test result does not rule out a diagnosis of lymphoma.[4]

Localized LyP is a clinical variant with only 16 reported cases.[5] This form of LyP tends to present around the age of 28 years, most commonly in the type A or histiocytic form, as was the case in our patient. Classic LyP has been reported to progress to an aggressive form of lymphoma in 5% to 24% of cases, depending on the review.

In localized forms of LyP, treatment does not appear to influence disease progression as the lesions show a tendency to resolve spontaneously in 1 to 3 months, sometimes with residual scarring. It is therefore reasonable to provide no treatment in most cases. Topical corticosteroids or local radiotherapy have been used for more serious cases, although these measures do not prevent further outbreaks.[6]

We present a case of localized LyP and draw attention to the need to consider this condition in the group of disorders that present with regional papular eruptions. This is an entity with a favorable prognosis, although further outbreaks are possible and a more aggressive form may develop in rare cases.

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