PhotoDermDiagnosis

Multiple ulcers in a young boy

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A 12-year-old boy presented to us with multiple painful ulcers over the extremities of 6 months duration. Child was completely asymptomatic 6 months back when he started developing multiple erythematous tender nodules on the upper and lower limbs associated with fever and malaise. The nodules turned into hemorrhagic bullae and ruptured to form ulcers covered with slough. There was no history of any hypopigmented anesthetic patches, neuritis, sensory, motor weakness, epitaxis or nasal stuffiness. Cutaneous examination revealed multiple tender subcutaneous nodules on the back, upper (Figure 1) and lower limbs with post inflammatory hyperpigmented macules. Multiple ulcers of varying sizes having a well-demarcated border and purulent discharge were seen on the forearms and thighs. Ichthyotic changes were seen on the legs with pedal edema. Peripheral cutaneous nerves were not thickened and there was no sensory or motor deficit. Laboratory investigations revealed the child to be anemic with normocytic normochromic anemia. The ESR was markedly raised. Urinalysis, liver and kidney function tests, complete iron profile were normal. Biopsy from the tender nodule showed dense neutrophilic infiltrate in the upper dermis, perivascular and periappendageal location and subcutis with collection of foamy macrophage granulomas in the dermis and subcutis (Figure 2).

What is your diagnosis?

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Diagnosis

Necrotizing erythema nodosum leprosum (NENL).

Discussion

Reactions in leprosy are divided into two types. Type 1 reaction is associated with alterations in cell-mediated immunity and is seen in tuberculoid and borderline groups; type 2 lepra reaction occurs in lepromatous leprosy. Erythema nodosum leprosum (ENL) in addition to the other features of systemic involvement such as arthritis, myositis, and nephritis, is the most characteristic lesion of type 2 reaction, although it is not seen in all cases. Classic ENL is characterized by the appearance of crops of tender painful subcutaneous nodules that are evanescent and last for 7-10 days, associated with fever and other constitutional symptoms. As the lesions subside they become purplish. Ulceration may occur and lesions heal with scarring. 50% of lepromatous leprosy patients and 25% of borderline leprosy patients experience an ENL reaction. In 15-50% cases, ENL is encountered during therapy of patients with lepromatous leprosy or borderline lepromatous leprosy usually within the first year, or it may even occur as the presenting manifestation of the disease. The usual triggers for ENL include surgical operations, pregnancy, parturition, lactation, menstruation, trauma, intercurrent illness, vaccination, physical or mental stress and sometimes even therapy. Precipitating drugs include iodides, bromides, diaminodiphenyl sulfones and chaulmoogra. Vesiculobullous, pustular, ulcerated, hemorrhagic and erythema multiforme-like lesions have been reported in ENL. A bullous-type reaction mimicking pemphigus in lepromatous leprosy has been reported from India however the patient did not show neuritis or ENL lesions, and histopathological examination and immunofluorescence studies were not performed to support the diagnosis. Necrotic ulcerative lesions of long standing untreated Lucio leprosy of Mexicans can be mistaken for necrotic ENL. The lesions of Lucio phenomenon appear on the extremities as painful tender red patches on the skin, which become purpuric; the centers of the lesions develop necrotic ulcers covered with black crust healing with superficial atrophic scars. Excellent results are seen with rifampin and good results with dapsone and steroids. NENL appears in crops along with constitutional symptoms such as fever, joint pain, and malaise, and it occurs in treated patients of lepromatous leprosy. NENL responds very well to thalidomide as well as to systemic steroids but not to rifampin and dapsone. Apart from NENL, ulceration in leprosy can also occur in a lepromatous nodule per se from pressure from within, in severe type 1 reaction and in lazarine leprosy. Lazarine leprosy was considered earlier to be occurring in atypical tuberculoid cases in the borderline spectrum near the tuberculoid pole, but in recent times it has been described as a subpolar form of lepromatous leprosy characterized by extensive ulceration of the lesions involving the trunk and extremities some deep enough to expose muscles and tendons. Cutaneous necrotizing vasculitis can also present as similar necrotic lesions. They can be differentiated by the absence of other features of leprosy, negative slit skin smears, and negative
findings in tissue biopsy. The diagnosis of NENL in our case was confirmed with a slit skin smear of BI 3+, MI 0% and biopsy showing characteristic macrophage granulomas. Our patient was managed with oral steroids which were gradually tapered over a period of 2 months, oral antibiotics along with MB-MDT and daily dressing for the ulcers with which the lesions healed.

References