SKIN CANCER (OTHER THAN MELANOMA)

BILATERAL BLINDNESS COMPLICATING A WIDESPREAD NECROBIOTIC XANTHOGRANULOMA

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Background: Necrobiotic xanthogranuloma (NXG) is a very rare non-Langerhansian histiocytosis. It is a systemic disease, clinically characterized by indurate yellowish to brown papules or nodules, most commonly in the periorbital region. Approximately, 130 cases of NXG have been described in the literature till now. We report a case of disseminated NXG associated with uveitis evolving towards blindness.

Observation: A 45-year-old woman presented to our skin department with a 2-year history of extensive annular lesions that had resisted to high-dose systemic corticosteroids. On examination, there were a symmetrical, pruriginous and painful annular lesions involving large areas of her face with markedly a periorbital distribution, trunk and extremities. Ophthalmological examination revealed an anterior uveitis.
Her biochemical parameters were within normal limits mainly there was neither diabetes nor hypercholesterolemia. Several biopsy specimens of cutaneous lesions revealed dermo-hypodermic infiltrates with eosinophilic necrobiosis material including some foamy histiocytes as well as cholesterol clefts. Features were suggestive of NXG. Both Serum electrophoresis and immunoelectrophoresis showed no paraproteinaemia. Thoracoabdominal-pelvic computed tomography (CT) scan did not find any other visceral locations. Although Chlorambucil and systemic corticosteroids have been introduced, her ophthalmic involvement progressively deteriorated towards bilateral vision loss.

Key message: Our patient presented lesions covering large areas of her body, which was exceptionally described. Ocular manifestation affects more than 50–80% of subjects. However, progression to blindness has been reported rarely (only one case in the larger series of 17 NXG seen by Wood et al). Monoclonal gammopathy was found in 23% of patients by Ugurlu et al. in their study. However, no paraproteinaemia was observed in our patient. Treatment of NXG is difficult. To our knowledge, a universally effective treatment has never been reported. In our patient, Chlorambucil and systemic corticosteroids were administered without success.