SKIN MANIFESTATIONS OF INTERNAL DISEASE

NECROBIOTIC XANTHOGRANULOMA WITH PARAPROTEINEMIA: A CASE REPORT

Tj Lee [1] - Ij Chang [1]

Taipei Medical University, Department Of Dermatology, Taipei Medical University Hospital, Taipei, Taiwan [1]

Background: Necrobiotic xanthogranuloma (NXG) is a rare slowly progressive granulomatous disorder of the skin. The clinical course is chronic and often progressive. NXG is closely associated with paraproteinemia with a monoclonal gammopathy. Other hematologic or lymphoproliferative diseases may also appear in association with NXG. The management of NXG is challenging and the recurrence is frequent.

Observation: We present a case of NXG with associated monoclonal gammopathy. There were hypertrophic and atrophic lesions only on the extremities at first and then the skin lesions progressed to periorbital area 10 years later. Laboratory data revealed pancytopenia. Moreover, serum and urine immunoelectrophoresis revealed monoclonal gammopathy of the IgG kappa type and bone marrow examination showed no significant findings.

Key message: NXG is a rare disorder and the typical presentation is yellow-orange, red-brown, or violaceous papules, nodules or infiltrated plaques. The cutaneous findings of yellowish to brown papules and plaques in periorbital area strongly suggest NXG, especially there are central atrophy and telangiectasias. Most patients with NXG have a monoclonal gammopathy with or without multiple myeloma. Evaluation of NXG includes a review of systems and laboratory exams to look for the underlying disease or systemic involvement. NXG may mimic necrobiosis lipoidica and plane xanthomas clinically and histologically. The management of NXG is challenging and the recurrence is not rare. Because of the rarity of the disease, data on the treatment of NXG are limited. Future studies for the pathogenesis and therapeutic outcome are required.