INTRODUCTION

Necrobiotic xanthogranuloma (NXG) is a multisystem disorder with prominent skin changes first described by Kossard and Winkelmann in 1980. The trunk and limbs may develop orange-red plaques and nodules, which may ulcerate. The lesions around the eyes resemble xanthelasmas except that they are firm, deep and indurated and may extend into the orbit. There is a monoclonal paraproteinemia in the majority of cases. The cause is unknown and the course is chronic and progressive.

The report describes an unusual skin presentation of the condition.

CASE REPORT

A 73-year-old Caucasian lady presented with a 6-year history of skin lesion on her left arm. She then developed further lesions on the other arm, chest, back and legs. The lesions were persistent and asymptomatic. No systemic symptoms were present. She had a course of oral steroids in the past without much success and this treatment was stopped.

On examination, she had well circumscribed, hyperpigmented and erythematous lesions with areas of atrophy resembling morphea (Figures 1 and 2), which were not consistent with NXG. The appendages were normal. There was no lymphadenopathy or hepatosplenomegaly.

The skin biopsy findings included broad zones of hyaline necrobiosis in the upper and deep dermis within granulomatous foci composed of histiocytes, foam cells and multinucleated giant cells of both Touton and foreign body type. Foci of cholesterol clefts were also noted with patchy lymphocytic infiltration. These findings were consistent with NXG.

The laboratory investigations revealed monoclonal IgG lambda paraprotein and cryoglobulins. Bone marrow aspirate showed 6% plasma cells consistent with a diagnosis of monoclonal gammopathy of uncertain diagnosis. Urine for Bence Jones protein was negative. Her latest blood tests showed: Hb of 13.9 g/dl, WBC 3.3 unit, platelets 148 unit, IgA and IgM level were normal at 3.06 g/l and 1.11 g/l respectively. IgG lambda paraprotein 6 g/l, normal renal hepatic profiles, albumin 33 g/l and adjusted calcium 2.45 mmol/l. Her regular repeat blood tests and clinical condition has remained...
stable during her recent reviews. Her present treatment plan therefore, comprises clobetasol (dermovate) for troublesome lesions as required and emollients.

**DISCUSSION**

NXG is a rare, progressive, histiocytic disease that features destructive cutaneous lesions, a close association with paraproteinemia, and multisystem extracutaneous manifestations. Although a causative role of the paraproteinemia is supposed, the pathogenesis of this disease remains obscure. The paraproteinemia in patients with NXG may even reflect a secondary phenomenon than the originating cause. It has a strong association with paraproteinemia which may progress to frank haematological malignancy.

Clinically, there are indurated nodules, yellow-red papules and nodules involving the face and less frequently the trunk and extremities. Histologically there are highly characteristic xanthogranulomas with giant cells and the Touton and foreign body type and degenerate collagen with lymphocytic and plasma cell infiltrate. NXG lesions with lichen sclerosus et atrophius have been reported. In the reported case, the skin lesions were highly unusual with no papules or nodules and instead reminiscent of morphea. This sort of presentation is not reported in the literature.

Patients with NXG may develop systemic involvement. Pulmonary, myocardial lesions involving spleen and giant cell myocarditis have been described. Eyes can be involved with orbital masses, conjunctivitis, keratitis, scleritis, uveitis, iritis, ectropion or proposis. Blindness may finally result. Lymphadenopathy, hepatosplenomegaly and mucosal lesions can occur. The bone marrow may become involved with plasmacytosis, anaemia, leukopenia, myeloma, or myelodysplastic syndromes.

There are different treatment options, none of which is curative. Antiblastics, cytotoxic drugs associated with steroids, alkylating agents like chlorambucil, melphalan and cyclophosphamide have been used in NXG with paraproteinemia

The patient’s prognosis depends on the degree of extracutaneous involvement and the presence of visceral malignancies.

**REFERENCES**