Case reports

Necrobiotic xanthogranuloma with orbital involvement

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Necrobiotic xanthogranuloma (NXG) is a rare non-X histiocytic disease characterized by a conspicuous dermatosis with a predilection for the periorbital tissues. It is associated with a dysproteinaemia, and various haematological abnormalities. We present a case of necrobiotic xanthogranuloma with orbital involvement, and report successful treatment with systemic low-dose Chlorambucil.

Case report

A 41-year-old woman was referred to a consultant ophthalmologist (BAN) in 1988 with a 4-year history of intermittent ocular discomfort. At that time she had unusual xanthelasmata around the medial aspect of both eyes. These had been present for approximately one year, and the patient had noticed a similar lesion in the right axilla.

In May 1990, the patient developed further skin lesions and was referred to the Department of Dermatology, where biopsy indicated the diagnosis of necrobiotic xanthogranuloma. Serum electrophoresis confirmed the presence of an IgG monoclonal paraproteinaemia, but there were no other significant haematological abnormalities.

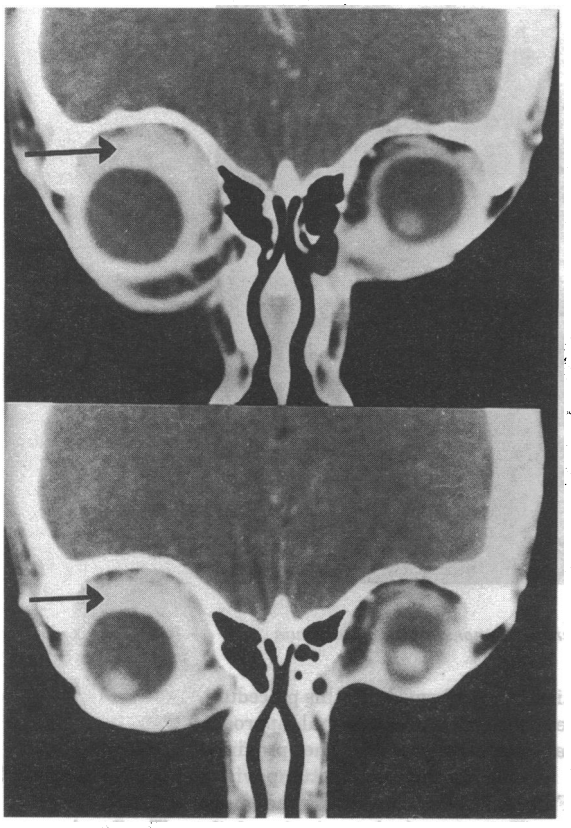

Figure 1. CT scan of head (coronal section) showing extensive soft tissue infiltrate around the right globe (arrow)

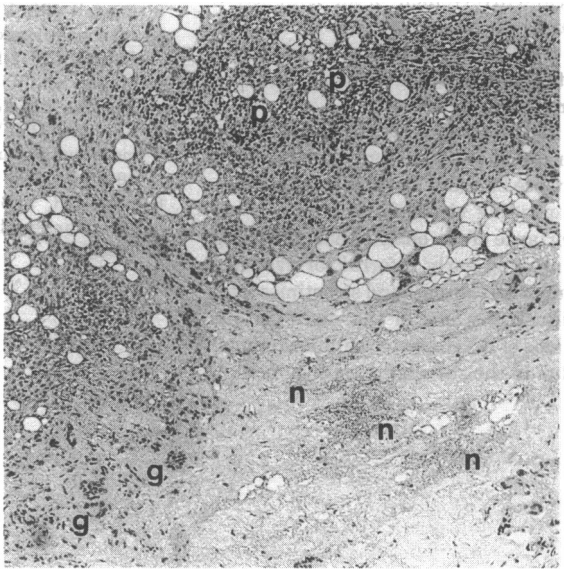

Figure 2. Orbital biopsy showing areas of Panniculitis (p), Giant cell formation (g), and Necrosis (n) with fragmented nuclear debris (H&E,×62)

Three months later, the patient presented to the eye department complaining of diplopia. Examination showed the presence of a firm mass in the superonasal quadrant of the right orbit. The globe was proptosed and depressed and ocular movements restricted, but there was no evidence of an optic neuropathy.

A CT scan (Figure 1) of the orbit showed an extensive soft tissue infiltrate around the globe, causing depression and lateral displacement. The mass extended to the posterior aspect of the globe.

Biopsy of the lesion (Figure 2) showed a diffuse infiltrate of lymphocytes, macrophages, and abundant eosinophils, but no plasma cells. Foamy macrophages were seen. Numerous multinucleate giant cells, some with the morphology of Touton giant cells, were present. Several foci of necrosis with fragmented nuclear debris in the centre were also seen. The diagnosis of necrobiotic xanthogranuloma was confirmed, and the patient was commenced on chlorambucil 4 mg daily.

At review 6 weeks later, the patient reported a subjective improvement confirmed by clinical examination. Her ocular movements were again full, and the proptosis had resolved. A further CT scan of the orbits confirmed this clinical improvement.

Discussion

Necrobiotic xanthogranuloma was first recognized as a discrete clinical entity by Kossard and Winkelmann in 1980[1,2], having been previously described in a number of ways including multicentric reticulohistiocytosis[3] and atypical necrobiosis lipoidica[4].

The cutaneous manifestations begin as painless non-pruritic papules that progress to nodules and plaques that may vary in appearance, but usually have a xanthomatous element. Associated findings include hepatosplenomegaly, a raised erythrocyte sedimentation rate, and a leucopenia. Various treatments have been evaluated, but the most

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successful appears to be the use of a low-dose alkylating agent, such as chlorambucil[5].

Ocular findings in NXG are common, and may dominate the clinical picture. In a series of 16 patients with NXG, 15 had ophthalmic manifestations[6]. The most frequent ocular finding was ocular discomfort. Involvement of the periorbital skin occurred in 13 patients. Other ophthalmic manifestations included anterior uveitis (three patients), keratitis (two patients), and involvement of the conjunctiva (three patients). Orbital involvement was described in two cases, but no treatment results were discussed. The response of orbital involvement to low-dose chemotherapy has not to our knowledge been previously reported.

Orbital radiotherapy has been reported with encouraging results[6], and radiotherapy in conjunction with low-dose chemotherapy may therefore be useful in rapidly progressive orbital disease with impending visual failure.

We reiterate the importance of the ophthalmologist being aware of NXG, thus ensuring early referral for systemic evaluation and treatment of this chronic and sometimes fatal disease. Our patient illustrates one of the more serious ophthalmic manifestations of NXG. Low-dose chlorambucil has seemed to be of value in the successful management of this case.

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Idiopathic megarectum treated by Duhamel's operation

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Chronic constipation with normal transit time may be due to Hirschsprung's disease or idiopathic megarectum. Distinguishing between these conditions requires clinical, radiological and histological studies.

Case report

A 21-year-old man was referred to St Mark's Hospital with a lifelong history of constipation. Over the previous 2 years, he had no spontaneous bowel motion despite regular enemas and laxatives. The only successful technique for emptying his rectum had been by manual evacuation performed under general anaesthetic. He had required 4 manual evacuations in the 18 months. On this occasion he complained of abdominal pain and distension, his last manual evacuation was 3 months previously.

On examination he was a fit young man with gross abdominal distension. Rectal examination revealed a distended rectum full of faeces. A gastrografin enema demonstrated a megarectum which was dilated from the sigmoid colon to the anal canal, with no distal narrowed segment (Figure 1). A study in which radio-opaque shapes were swallowed demonstrated normal colonic transit up to the rectum. Anorectal physiology studies revealed a normal rectoanal inhibitory reflex. He underwent a manual evacuation of faeces and a full thickness rectal biopsy. Histology showed that ganglion cells were present.

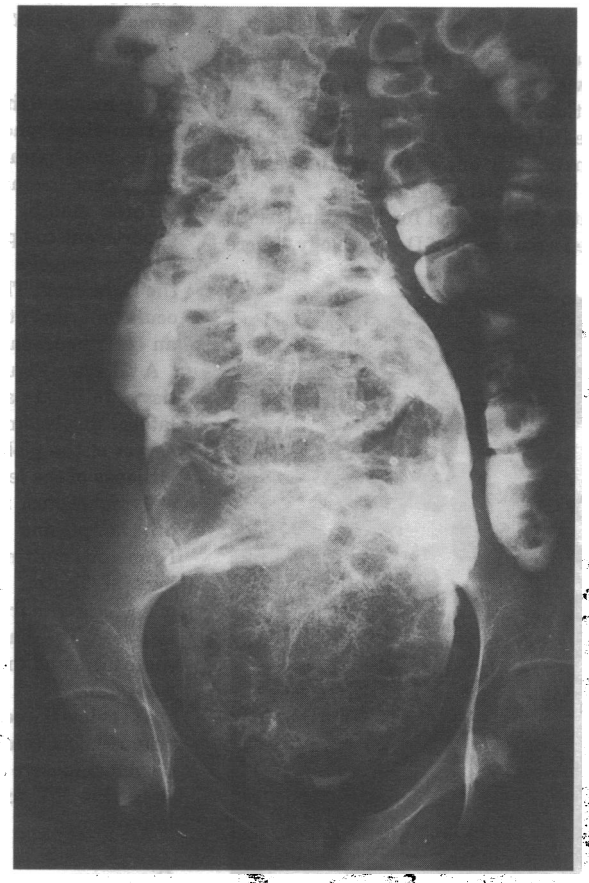

Figure 1. Gastrografin enema demonstrating megarectum

A Duhamel's operation was performed[1]. In this procedure, the megarectum is excised via an abdominal approach and the distal rectal stump oversewn. The proximal is anastomosed end to side to the rectal stump via a perineal approach (Figure 2). Postoperatively he made an uneventful recovery. Three months later, he has daily bowel motions, but still requires laxatives and occasional enemas.

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