Eosinophilic leukaemia with trisomy 8 and double gammopathy

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Abstract

Prolonged eosinophilia of unknown cause has generally been described as the hypereosinophilic syndrome, and is characterised by peripheral blood and bone marrow infiltration and frequent multisystem disease. The nature of this disorder has been questioned, and the clinical features are quite variable, suggesting its heterogeneity and probable neoplastic aetiology.

A patient with severe eosinophilia, karyotype abnormalities, serum gammapathy and massive organ disease is reported. The clinical course was aggressive despite cytoreduction of eosinophils and terminated in multisystem failure.

These findings are consistent with a diagnosis of eosinophilic leukaemia, and it is suggested that chromosome and cell culture studies might be useful in the early diagnosis of this controversial entity.

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Eosinophilic leukaemia is rare and its differentiation from the hypereosinophilic syndrome, myeloproliferative disorders, and acute leukaemia with eosinophilia is in many cases controversial. Cell culture studies of peripheral blood may help to distinguish malignant disease from the idiopathic hypereosinophilic syndrome.1-3 Cytogenetic analysis with banding techniques has disclosed many abnormalities in eosinophilic leukaemia.4-4

We report a case of hypereosinophilia, with features consistent with a leukaemic process, abnormal karyotype, and double gammopathy; as far as we are aware, the latter has not been described in association with eosinophilic leukaemia.

Case report

A 75 year old man presented with a three week history of fever, vomiting, diarrhoea and hiccups. He smoked 20 cigarettes daily and had recurrent urinary tract infections in the previous months. There was no history of allergies, he was not taking regular medica-

Figure 1 Pronounced eosinophilia with aberrant forms; myelocytes and metamyelocytes are prominent (Peripheral blood; May-Grünwald stain).

Figure 2 Quantification of the monoclonal bands by densitometric scanning of the immunofixation pattern.
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platelets $37 \times 10^9/l$; there were numerous hyper-segmental eosinophils, with giant forms and degranulation (fig 1). A neutrophil alkaline phosphatase score was 85. Liver and renal function tests yielded normal results; a coagulation screen was normal.

Total protein concentration was 77 g/l; neutrophil albumin 27 g/l; of the globulins, $a_1$ 1–5 g/l; $a_2$ 2–9 g/l; $\beta$ 8 g/l; $\gamma$ 27–9 g/l. IgG was 20–5 g/l (normal value 8–17 g/l); IgA was 5–09 g/l (normal value 1–4–9 g/l); IgM was 2–34 g/l (normal value 0–5–3–2 g/l). The serum protein electrophoresis disclosed an abnormal pattern and immunofixation showed an IgG $\kappa$ band of 0–5 g/l and a distinct IgG $\lambda$ band of 1–5 g/l (fig 2). Other immunological studies were normal. Bone marrow aspirate showed infiltration with eosinophils, depressed erythropoiesis, and occasional megakaryoblasts.

An ECG showed a right bundle branch block and a left anterior fascicular block. A Doppler echocardiogram showed left atrial enlargement with mitral regurgitation. A chest x ray picture was normal. Blood and urine cultures were negative. Serological investigations for helminthic diseases proved negative.

Cytogenetic studies in the peripheral blood showed trisomy 8. The patient was treated with leukapheresis and hydroxyurea and had blood and platelet support. There was a substantial reduction in the peripheral eosinophil counts after therapy, with persistent anaemia and thrombocytopenia. The clinical condition deteriorated progressively, with mental confusion, renal and liver failure, and disseminated intravascular coagulation. The patient died 21 days after treatment had been started. Permission for necropsy was refused. The aggressive clinical course terminating in multigang failure associated with eosinophilia and karyotype anomaly was consistent with a diagnosis of eosinophilic leukaemia; the finding of a double gammopathy has been described in others conditions, but to our knowledge has not yet been reported in this situation and its clinical importance is not known.

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