IgE myeloma associated with plasma cell leukaemia

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Summary

A patient presented with plasma cell leukaemia and was found to have an IgE secreting multiple myeloma. IgE myeloma is very rare with only 18 cases recorded. However, this case is the fifth recorded of IgE myeloma associated with plasma cell leukaemia. Since plasma cell leukaemia only occurs in 1-6% of all cases of myeloma, it seems certain that this is a significant association.

KEY WORDS: IgE myeloma, plasma cell leukaemia.

Introduction

Nineteen cases of IgE myeloma have been reported including the present one and five have been associated with plasma cell leukaemia (Johansson and Bennich, 1967; Ogawa et al., 1969; Sala et al., 1981; Endo et al., 1981). Plasma cell leukaemia is itself found in only 1-6% of all cases of myeloma (Kyle, Maldonado and Bayrd, 1974).

The number of cases now seems large enough to conclude that a definite association exists between IgE myeloma and plasma cell leukaemia.

Case report

A 67-year-old man with maturity onset diabetes mellitus was admitted complaining of a cough with purulent sputum, recent loss of weight and drowsiness. He had become confused. He denied any bone pain. He was found to be weak, disorientated and dehydrated.

On examination, he was found to have mitral regurgitation but no other abnormal physical signs. Initial investigations revealed a haemoglobin of 11·0 g/litre and a white cell count of 11·8×10⁹/litre of which 20% were plasma cells. He was in renal failure, his blood urea being raised at 24·4 mmol/litre and creatinine at 546 µmol/litre. The plasma calcium level was within normal limits. His plasma viscosity was slightly raised at 2·23 cp. X-rays of his skeleton failed to demonstrate any abnormality. Bone marrow aspirate contained 90% abnormal plasma cells. Total protein was 102·5 g/litre of which 37·7 g/litre was albumin. Serum electrophoresis showed a band in the α-region.

Immunoelectrophoresis showed the presence of a IgE type lambda monoclonal protein in his serum and free lambda light chains (Bence Jones protein) in the urine. The IgE level was 38 g/litre by densitometry, and 28 g/litre by radioimmunoassay. The other serum immunoglobulin levels were depressed.

Examination of the marrow using fluorescein-conjugated antisera specific for each of the immunoglobulin heavy chains and for kappa and lambda light chains, showed that the vast majority of nucleated cells contained IgE-lambda in their cytoplasm. The cells were of varied morphology with most appearing as plasma cells, although numerous large blast-like cells also showed cytoplasmic IgE. In most of the cells, the staining was relatively light and mainly at the periphery. IgG, IgA, IgM and kappa staining cells were decreased in frequency and no IgD plasma cells were seen.

During these investigations, his haemoglobin dropped to 9·2 g/litre and his white cell count rose to 25·8×10⁹/litre of which 61% were plasma cells. His blood urea rose to 33·0 mmol/litre.

He was rehydrated with intravenous fluids and his renal failure gradually improved. Therapy was commenced with melphalan 5 mg a day for 5 days, repeated at monthly intervals. The response was excellent with complete clearance of plasma cells from his peripheral blood in 7 days and resolution of his confusion.

However, after some months, he again became...
confused and was found to have a rising paraprotein level. Plasma cells reappeared in the peripheral blood. In spite of further chemotherapy, he died, 36 weeks after presentation.

Discussion

The features of IgE myeloma are becoming clearer. The age of onset (mean 62 years), sex ratio (53% male) and frequency of Bence Jones proteinuria (63%) are similar to ordinary myeloma (Kyle, 1975). However, anaemia may be more frequent. The haemoglobin at presentation was less than 10 g/litre in 63% of IgE cases compared with 33% overall. In contrast, severe renal failure only occurred in three of the IgE cases although mild renal impairment is common (71% of IgE cases compared with 55% overall). The X-ray findings in IgE myeloma are curious. Normal bone X-rays are more common (33% compared to 21%) and lytic bone lesions are less often seen (44% compared with 70%). Bone sclerosis is very rare in myeloma generally but has been found in two IgE cases (Rogers et al., 1977). Because many case reports have been published before the patient's death, it is difficult to be precise about the prognosis; probably the survival is similar to myeloma generally, a median of about 20 months.

Although the numbers are small, it seems certain that the incidence of plasma cell leukaemia in IgE myeloma (26%) is genuinely much higher than in myeloma overall (1-6%). The factors controlling the release of plasma cells into the blood are little known. It has been suggested that surface IgE increases the propensity for their release into the circulation but the mechanism is unknown.

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References


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