Gelatinous degeneration presenting as a preleukaemic syndrome

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Abstract
Gelatinous degeneration of marrow is a rare histological disorder associated with chronic debilitating diseases, such as anorexia nervosa, AIDS and postchemotherapy aplasia. Solid tumours have been associated with this condition but it has been reported in only two patients with leukaemia. In these cases leukaemia and gelatinous degeneration were diagnosed simultaneously. In the case reported here, a 48 year old man, gelatinous degeneration was the only histological finding observed more than two years before the diagnosis of acute myelogenous leukaemia with monosomy 7. The significance of hyaluronic acid deposition remains uncertain. Two hypotheses have been put forward: (1) that gelatinous degeneration occurs during tissue repair; and (2) that gelatinous degeneration inhibits haemopoiesis by altering the microenvironment of the bone marrow. In the case reported here, the presence of monosomy 7 suggests that myelodysplasia was the underlying disorder which finally evolved into acute leukaemia.

Keywords: gelatinous degeneration, acute myelogenous leukaemia, monosomy 7.

Gelatinous degeneration is generally diagnosed in bone marrow biopsy specimens by the presence of a focal or generalised extracellular deposition of a gelatinous material, identified as hyaluronic acid, in association with atrophy and marrow hypoplasia. This disorder has been referred to as serous atrophy, mucoid degeneration and starvation marrow. Gelatinous degeneration has been classically observed in association with chronic debilitating disorders, such as anorexia nervosa, starvation, malignancy, chronic infections, systemic lupus erythematosus, and myxoedema. Recently, it has been widely reported in patients with AIDS and a variant form of the classical degeneration has been reported after the administration of chemotherapy.

In the case reported herein, gelatinous degeneration was diagnosed in a previously healthy man, with no evidence of an underlying disorder. To our knowledge, this is the first reported case in which gelatinous degeneration preceded the diagnosis of acute myelogenous leukaemia (AML).

Case report
A 48 year old healthy, well nourished, white man was admitted to hospital in August 1992 because of fever and pancytopenia. He worked as a driver for a chemical company, smoked...
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Munich, Germany) was administered and a mild increase in the patient’s neutrophil count was observed. In June 1993, a bone marrow biopsy specimen showed an increase in the amount of mucoid material present and dysplastic changes were observed in the three haemopoietic lines. Repeated tests for occult disease during this period were negative.

In December 1994, blast cells were detected on peripheral blood smears and a bone marrow aspirate revealed immature cells with a myeloid appearance, vacuolated cytoplasm and visible nucleoli, comprising 28% of the total cell population. Peroxidase, non-specific esterase and periodic acid Schiff stains were negative. Immunological markers in peripheral blood showed expansion of immature myeloid precursors with expression of CD34, CD33, CD56, and CD13. Cytogenetic studies showed monosomy 7 in all metaphases studied. Examination of a bone marrow biopsy specimen revealed proliferation of immature myeloid cells with reticulin fibrosis; mucoid material was not detected (fig 2). Two cycles of daunorubicin and cytarabine were administered without response. The patient died in May 1995.

Discussion
To our knowledge, gelatinous transformation of the bone marrow has been described in only two patients with acute leukaemia. However, in
We speculate that deposition of hyaluronic acid resulted from a metabolic imbalance generated by altered bone marrow function. The presence of monosomy 7 might indicate that myelodysplasia was the underlying haematological disease in our patient, which finally developed into AML. In fact, although the bone marrow aspirates were too scanty for an accurate haematological diagnosis, myelodysplastic changes could be detected at the later stages of the patient's clinical course.

Anaemia or moderate pancytopenia are the most common findings described in patients with gelatinous transformation. In that regard, Seaman et al suggested that hyaluronic acid is a putative inhibitor of haemopoiesis which acts by altering the microenvironment of the bone marrow. Indeed, deposition of hyaluronic acid in the bone marrow of our patient may have lead to the development of AML, although we believe that this is unlikely.

In patients with anorexia nervosa the gelatinous material may disappear when their nutritional status improves. It also disappears in patients with postchemotherapy aplasia, when haemopoietic recovery takes place. These observations support a role for hyaluronic acid in tissue repair. Interestingly, in our patient the gelatinous ground material disappeared when he developed leukaemia.

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