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Lymphadenopathy and hepatosplenomegaly in a patient with acute myelogenous leukaemia

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DESCRIPTION

An 18-year-old man was admitted to our hospital with fever and generalised lymphadenopathy in October 2010. One month prior to admission, a submaxillary nodule appeared, with gradually progression to nodes involving

the entire body. His medical history was unremarkable, and he was not receiving any medication. Physiological parameters were: body temperature, 37.7°C; heart rate, 96 beats/min and regular; and blood pressure, 143/72 mm Hg. Physical examination revealed generalised enlargement of

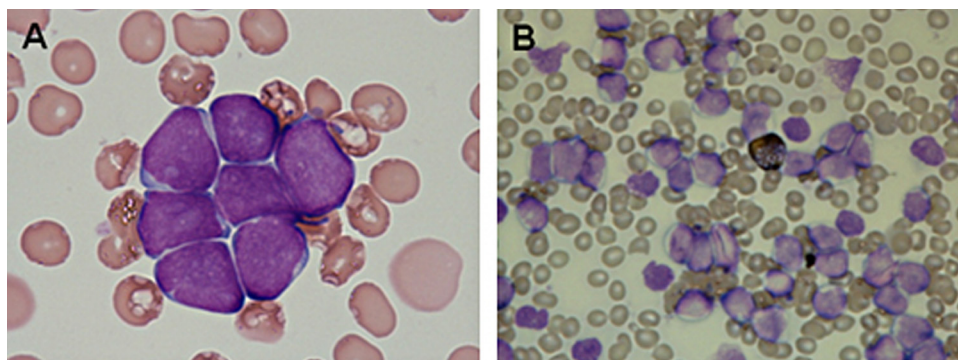


Figure 1 Bone marrow examinations at initial diagnosis (A May-Giemsa stain, 1000×; B Myeloperoxidase stain, 400×)

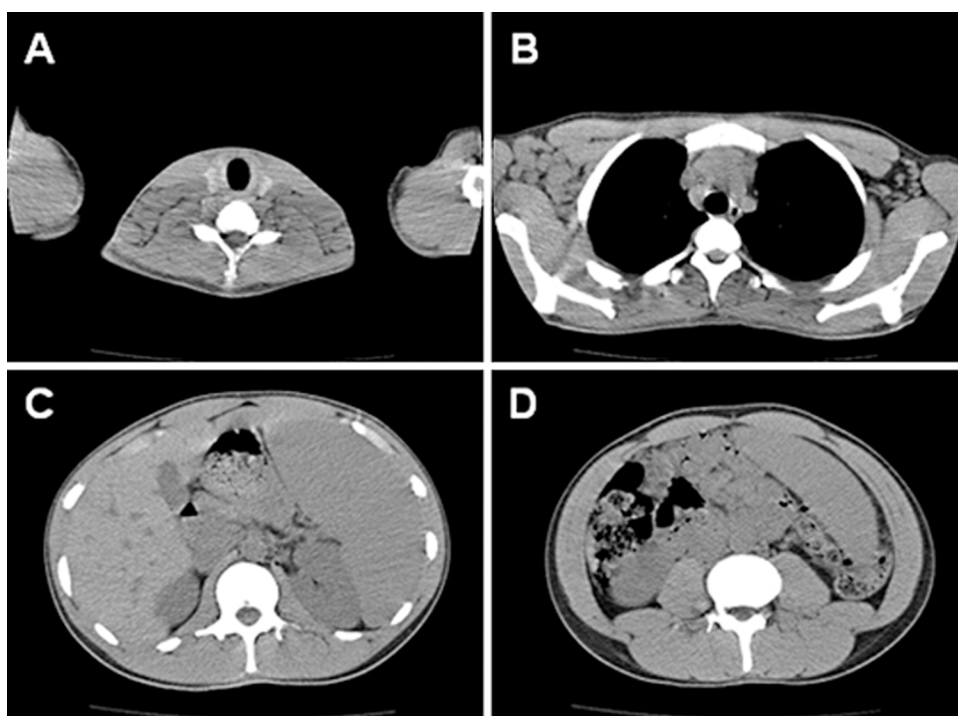


Figure 2 CT at initial diagnosis.

superficial lymph nodes, varying in size from about 1 to 2 cm. Hepatosplenomegaly was identified. Blood analyses showed a white blood cell count of 364 800/ μ l with 99.8% blasts, a haemoglobin level of 13.4 g/dl and a platelet count of 331×10^3 / μ l. Biochemical analysis showed a serum lactate dehydrogenase level of 854 IU/l (normal 101–213 IU/l). Bone marrow was hypercellular, with diffuse infiltration of undifferentiated myeloperoxidase-negative blasts (66%) (figure 1). Blastic cells were positive for CD4, CD7, CD13, CD33, CD34 and HLA-DR on immunophenotyping analysis. These results led to a diagnosis of acute myelogenous leukaemia M0 according to the French-American-British classification. Cytogenetic analysis of blastic cells revealed 46,XY,t(10;11)(p12;q14) (20/20), and molecular analysis also showed clonal immunoglobulin gene rearrangements.¹ CT revealed systemic lymphadenopathy both superior and inferior to the diaphragm, and marked enlargement of the spleen (figure 2). The patient underwent induction chemotherapy with idarubicin (12 mg/m², days 1–3) and cytosine arabinoside (100 mg/m², days 1–7). Rapid regression of lymphadenopathy and hepatosplenomegaly was observed after chemotherapy and complete remission was achieved.

Learning points

- ▶ Skin, gingiva, gastrointestinal tract and central nervous system may often be associated with extramedullary involvement in the setting of AML. Granulocytic sarcoma is also a tumour composed of myeloblasts or monoblasts, and may be found in virtually any location, but is usually localized. Such prominent generalised lymph node involvement combined with hepatosplenomegaly as in this case is extremely rare in AML at onset, in contrast to acute lymphoblastic leukaemia (ALL).
- ▶ Moreover, the translocation t(10;11)(p12;q14), which results in fusion of the clathrin assembly lymphoid myeloid leukaemia (CALM) gene with the AF10 gene, was found in this case. CALM-AF10 fusions have been observed as a rare but recurring phenomenon in patients with ALL rather than AML, and are often associated with poor prognosis.¹ Extramedullary involvement is a typical characteristic among patients with this translocation.

Competing interests None.

Patient consent Obtained.

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1. Caudell D, Aplan PD. The role of CALM-AF10 gene fusion in acute leukemia. *Leukemia* 2008;**22**:678–85.

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