Massive myeloid sarcoma affecting the central nervous system, mediastinum, retroperitoneum, liver, and rectum associated with acute myeloblastic leukaemia: a case report


Myeloid sarcoma, formerly known as granulocytic sarcoma, was described by Rappaport in 1966. It is an extramedullary solid tumour composed of myeloblasts and other granulocytic precursors, with or without an associated haematological malignancy. This tumour was originally referred to as chloroma, because of its green colour, secondary to the presence of myeloperoxidase. Single myeloid sarcomas are present in about 2% of patients with acute myelogenous leukaemia (AML). The diagnosis of myeloid sarcoma can be difficult. Traweek advised the use of immunohistochemical staining of paraffin wax embedded tissue sections, using antibodies to CD20, CD33, CD68, and myeloperoxidase, and reported an accuracy of 96%. In most patients, myeloid sarcoma associated with AML affects no more than two sites. We report a highly unusual patient with multifocal tumours affecting five different anatomical sites, namely: the rectum, the right lobe of the liver, the mediastinum, the kidney, and the brain.

‘‘The diagnosis of myeloid sarcoma can be difficult’’

CASE REPORT

A 37 year old man was admitted in September 1999 with a two month history of weight loss, symptoms of anaemia, and rectal bleeding. On examination, he appeared pale, underweight, and tachycardic, with petechiae and ecchymosis on the lower limbs. Fundoscopic examination showed bilateral papilloedema and several retinal flame haemorrhages. A seventh left cranial nerve lesion was found. The liver edge was 5 cm below the right costal border. Laboratory studies showed a haemoglobin concentration of 45 g/litre, white blood cell count of 4.1 × 10⁹/litre, platelet count of 50 × 10⁹/litre, and 35% blast cells; some blast cells contained Auer rods. Lactate dehydrogenase was 12 931 IU/litre (normal range, 91–180). Chest radiography showed mediastinal expansion. Computed tomography of the brain showed a solitary hyperdense subdural nodule, 2.9 cm in diameter in the occipital region (fig 1). Computed tomography of the abdomen and pelvis showed a solid mass, 10 cm in diameter, with regular edges and central hypodensity, compatible with necrosis of the right lobe of the liver. The liver–kidney space was occupied by tumour. The left kidney showed a 4 cm tumour mass. The rectum and sigmoid were infiltrated by an irregular 8 cm diameter tumour (fig 2A, B).

Bone marrow aspiration was compatible with AML M2 (fig 3A, B). Flow cytometry showed that the peripheral blood was positive for CD4, CD11, CD13, CD14, CD33, CD45, and HLA-DR. A bone marrow karyotypic study revealed an 8;21 translocation. The presence of multiple solid tumours in AML is a rare event. Enhanced expression of cell adhesion molecules may be the reason why some patients develop myeloid sarcomas.
remission, which was maintained for 18 months. Despite an allogeneic bone marrow transplant, he subsequently relapsed and at relapse had myeloid sarcomas in the inferior pole of the right kidney, at the right ureterovesical junction, and the rectum.

**DISCUSSION**

Myeloid sarcoma is an extramedullary tumour composed of granulocyte precursors.\(^2\) It is frequently found in bones, peritoneum, lymph nodes, skin, and epidural structures.\(^6\) Myeloid sarcoma in patients with t(8;21) was first reported in 1984 by Swirsky \textit{et al}, who found that seven of 30 patients with t(8;21) had solid leukaemic deposits, principally in the mastoid, orbital cavities, or thoracic spine (extradural).\(^7\)

Because myeloid sarcoma can be seen in the absence of AML, evaluation of unusual tumours by immunohistochemical staining for CD33, CD68, and myeloperoxidase is advised.\(^3\)

"The presence of neural cell adhesion molecule on the surface of blast cells has been shown to enhance their propensity for tissue penetration into the central nervous system."\(^8\)

Solid tumour masses are uncommon in AML and their presence might be attributable to overexpression of cell adhesion molecules. The presence of neural cell adhesion molecule on the surface of blast cells has been shown to enhance their propensity for tissue penetration into the central nervous system, in particular into the cerebellum.\(^9\)
Allogeneic bone marrow transplantation has been advocated in patients with a solitary myeloid sarcoma, but was not of longterm benefit in our patient with AML and multiple myeloid sarcomas. The patient gave his permission for this case report to be published.

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Take home messages

- We describe an unusual case of myeloid sarcoma associated with acute myelogenous leukaemia (AML) with tumour formation at five anatomical sites.
- The anatomical sites affected were: the rectum, the right lobe of the liver, the mediastinum, the retroperitoneum, and the central nervous system.
- The presence of multiple solid tumours in AML is rare.
- The development of myeloid sarcomas may be associated with enhanced expression of cell adhesion molecules.