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 patient received combination chemotherapy and craniospinal radiotherapy, and achieved complete haematological response.

Figure 1 Brain computed tomography scan showing a solitary mass at the right occipital region, indicated by a closed arrow.

**Abbreviations:** AML, acute myelogenous leukaemia
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. It is frequently found in bones, peritoneum, lymph nodes, skin, and epidural structures. Myeloid sarcoma in patients with t(8;21) was first reported in 1984 by Swirsky 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). 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. 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. 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.

![Figure 2](A) Abdominal computed tomography scan showing a solid tumour with hypodensities suggestive of necrosis in the hepatorenal space and retroperitoneal cavity, causing a compression effect against the right kidney. (B) Pelvic computed tomography scan showing the presence of a solid tumour at the rectum–sigmoid level, with hypodensities indicated by the numbers 1, 2, and 3.

![Figure 3](A) Low power view of a bone marrow smear (Wright stain) showing a clump of myeloblasts; original magnification, ×40. (B) High power view of a bone marrow smear (Wright stain) showing a clump of myeloblasts; original magnification, ×100. (C) High power view of a touch preparation of the pararenal tumour mass (Wright stain); original magnification, ×100. (D) Touch preparation of the pararenal tumour mass showing Sudan black positive myeloblasts. (E) Cerebrospinal fluid smear showing myeloblast infiltration (Wright stain). (F) Cerebrospinal fluid smear showing myeloblast infiltration (Sudan black stain).
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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REFERENCES

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