Myeloid Sarcoma in a Child with Acute Myeloblastic Leukaemia

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INTRODUCTION

Myeloid (granulocytic) sarcoma or chloroma is an extra medullary tumour composed of immature malignant white blood cells or myeloblasts with or without maturation.¹ It affects older adults more frequently. Simple infiltration by myeloid blasts in any part of the body to form tumour mass is not labelled as myeloid sarcoma unless the tissue architecture of the part is lost. This tumour may precede or occur concurrently with acute or chronic myeloid leukaemia. It can also occur with other myeloproliferative disorders (MPD) or myelodysplastic syndrome (MDS). Sometimes, it is the initial manifestation of relapse in a previously treated case of acute myeloblastic leukaemia or herald the onset of blast transformation in a case of CML.¹ Myeloid sarcoma is a rare condition, especially its occurrence in a child in association with AML.² ³ This is a case report of this rare condition.

CASE REPORT

A 7 years old male child presented in Fauji Foundation Hospital, Rawalpindi, in January 2008, with proptosis and swelling in left parotid region for 40 days. He had 3 weeks history of generalized weakness and 2 weeks history of high grade fever.

Physical examination revealed a weak looking child with large left parotid region swelling about 4 x 5 cm in size (Figure 1). It was hard in consistency and not fixed to underlying structures. Overlying skin was normal looking and not fixed to this growth. There was also bilateral proptosis especially marked in the left eye. Pallor was positive. Lymph nodes, liver and spleen were not palpable. Fundoscopy showed established papilloedema. Ear examination showed left conductive deafness due to collapse of left external auditory canal (pressure effect of tumour). CT scan of brain and neck showed locally infiltrative mass in nasopharyngeal region with intraorbital and intracranial extension (Figure 2). A differential diagnoses of rhabdomyosarcoma, lymphoma and meningioma were suggested by the radiologist. Fine needle aspiration cytology of parotid region swelling showed benign epithelial lesion. CSF examination showed no abnormality.

Blood CP revealed TLC 14.8 x 10⁹/1, HB 7.8 g/dl, platelets 32 x 10⁹/l. Peripheral blood smear showed 65% blasts, some showing Auer rods. Liver and renal function were in normal limits.

Bone marrow aspiration revealed hypercellular marrow with 85% blast cells some showing Auer rods. The blasts were Sudan black B positive. A diagnosis of acute myeloblastic leukaemia FAB type M₂ was made.

Induction chemotherapy (ADE regimen: injectable cytosar, daunorubicin and etoposide) was started in first week of February 2008. He went into complete clinical and haematological remission. The bone marrow
aspiration report in March 2008 showed < 5% blast cells. Parotid swelling and proptosis disappeared. He was then lost to follow-up.

**DISCUSSION**

Myeloid (granulocytic) sarcoma has been called chloroma, derived from greek word chloros meaning green due to cut surface appearance caused by the myeloperoxidase enzyme.4 These tumours can virtually affect any organ, but subperiosteal bone structures like skull, paranasal sinuses, ribs and vertebrae are more common sites. Skin and lymph nodes may also be involved. This patient had a mass in paranasal sinuses which extended intracranially and retro-orbitally and manifested with proptosis due to pressure effect. A similar case of myeloid sarcoma associated with CML, presenting with proptosis and blindness has been reported.2 Paraparesis due to extradural myeloid sarcoma in association with AML was reported in another patient.3

Myeloid sarcoma can present de-novo as a primary tumour or concurrent with AML (as in this patient) or with a myeloproliferative disorders.1 Incidence of myeloid sarcoma in AML is 3-5%.5 The latest classification of AML by WHO considers de-novo myeloid sarcoma as a separate entity and the detection of de-novo myeloid sarcoma is taken as equivalent of diagnosis of AML.1 De-novo myeloid sarcoma eventually develops AML (median time of 7-12 months) and these transformed cases show very poor prognosis.5 Patients of AML in remission, who develop myeloid sarcoma, almost always have bone marrow relapse (within a median time of 7 months) and have a poor prognosis.4 Myeloid sarcoma is characterized according to the predominant cell type, most common being granulocytic sarcoma which is composed of myeloblasts, neutrophils and neutrophil precursors. Other forms include monoblastic sarcoma and tumour with trilineage hemato-poiesis.1 In this patient, tumour was difficult to access so tissue diagnosis was not attempted.

The most common differential diagnosis of myeloid sarcoma is non-Hodgkin’s lymphoma which can pose a diagnostic dilemma.6 In one study 47% of myeloid sarcoma cases were initially misdiagnosed as malignant lymphoma.7 A patient from Pakistan initially diagnosed as T-cell non-Hodgkin’s lymphoma was later reviewed and found to have de-novo myeloid sarcoma.8

Definite diagnosis of myeloid sarcoma is helped by morphology, cytochemistry, and immunophenotype of tumour cells. On immunohistochemistry, CD 68KP1 is the most commonly expressed marker followed by MPO, CD117, CD 99, CD 34, AND TdT.1

In this patient we had a concurrent diagnosis of AML-M2 with myeloid sarcoma, so induction chemotherapy was started immediately which resulted in prompt relief of symptoms. There is no standard protocol of chemotherapy for de-novo myeloid sarcoma and standard chemotherapy against AML is only moderately effective in suitable cases.4 The clinical behaviour and response to treatment is not influenced by factors such as age, gender, anatomical site, pattern of presentation, histological, cytogenetic and immunophenotypic features.1,9 However, some studies suggest that systemic chemotherapy against AML can help reduce rate of leukaemic transformation of de-novo myeloid sarcoma.5 Only bone marrow transplant results in prolonged survival.9

To conclude, myeloid sarcomas are rare tumours which usually present in association with myeloid leukaemias. When presenting as primary tumours, they may be misdiagnosed and only a high degree of suspicion and thorough workup lead to correct diagnosis and prompt treatment thus preventing serious complications.

**REFERENCES**


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