



Case report

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Proptosis in orbital myeloid sarcoma: A precursor to acute leukaemia

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ABSTRACT

Myeloid sarcoma is a rare, aggressive presentation of acute myeloid leukemia which tends to affect bones and soft tissue. It is usually seen in children, and has a poor prognosis. We report a case of orbital myeloid sarcoma presenting with unilateral proptosis in a young adult who subsequently developed acute myeloid leukaemia. An 18-year-old male presented with sudden, progressively increasing right eye swelling over two weeks. Examination revealed right eye proptosis, with a positive relative afferent papillary defect and a right eye visual acuity of 6/60, pinhole 6/18. The right fundus showed optic disc hyperemia, tortuous veins and choroidal folds. The left eye was normal. On initial examination, computed tomography (CT) of the brain and orbit showed an enhancing mass at the lateral aspect of the right orbit measuring 3.8 x 2.1 cm, with optic nerve displacement. Right lateral orbitotomy and excisional biopsy yielded a tissue diagnosis of myeloid sarcoma. The peripheral blood smear was normal at this time. The patient defaulted treatment but presented again three months later with right eye massive proptosis and loss of vision. A repeated CT scan showed a large lobulated mass measuring 104.5mm x 87.21mm x 77.25mm occupying the right orbit, with destruction of the bony orbital walls and intracranial extension. A repeat full blood picture and bone marrow aspiration showed 30% blast cells, consistent with the diagnosis of acute myeloid leukemia. Patient was started on chemotherapy and radiotherapy. The tumour reduced in size after five cycles, but unfortunately, patient passed away due to brain metastases.

1. Introduction

Myeloid sarcoma is an extra-medullary proliferation of cells of myeloid origin. It is usually associated with acute leukemia, but may also be seen in other myeloproliferative disorders and myelodysplastic syndromes[1]. Myeloid sarcoma is more common in children, with a median age of 7 years old[2]. Also known as primary myeloid sarcoma, this condition rarely occurs without a pre-existing or concomitant diagnosis of leukemia; in the majority of cases reported, myeloid sarcoma occurred 1 to 25 months after the diagnosis of leukemia[3]. Our case illustrates a rare presentation of orbital myeloid sarcoma in a healthy adult male.

2. Case report

An 18-year-old Malay boy presented with sudden, progressively increasing right eye proptosis over two weeks. There was associated eye redness, eye pain upon eye movement, headache and giddiness. There was no history of fever, rhinorrhea, coughing, sinusitis or trauma. In addition, there were no symptoms of hyperthyroidism, and he had no underlying medical conditions. He sought treatment at the nearest clinic, and was initially presumed to have idiopathic inflammatory orbital disease. He was started on oral prednisolone 25mg BD for 1 week, which was subsequently tapered. However, the proptosis worsened after the corticosteroids were initiated, and the right eye vision deteriorated, prompting the patient to seek an ophthalmology opinion.

On examination, the visual acuity in the right eye was 6/60, pinhole 6/18. The relative afferent pupillary defect was positive in the right eye, and extraocular movements of the right eye were restricted in all directions. There was right eye non-axial proptosis,

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with conjunctival injection and chemosis (Figure 1). The rest of the anterior segment examination was otherwise normal. The right eye intraocular pressure was 24 mmHg. Fundus examination of the right eye showed optic disc hyperemia, tortuous veins and choroidal folds. The left eye was normal. Complete blood count and leukocyte differential count were normal. Peripheral blood smear did not reveal any blast cells.



Figure 1: Right eye proptosis with conjunctival chemosis, seen from the (A) anterior and (B) anteromedial aspect.

Computed tomography (CT) of the brain and orbit showed an enhancing mass at the lateral aspect of the right orbit measuring 3.8 x 2.1 cm, with displacement of the optic nerve. Right lateral orbitotomy and excisional biopsy were done. Histological examination showed a malignant tumour composed of diffuse infiltrates of blue neoplastic cells under hematoxylin and eosin stain (Figure 2A). Tumour cells were medium-to-large in size, with irregular nuclear contour and scanty cytoplasm. Immunohistochemical studies of the tissue were positive for CD34, CD117, CD56, CD99, CD 15, CD 10 and myeloperoxidase (Figure 2B). These findings were suggestive of myeloid sarcoma. The patient was informed regarding the diagnosis, but he refused chemotherapy and defaulted follow-up to seek traditional medicine.

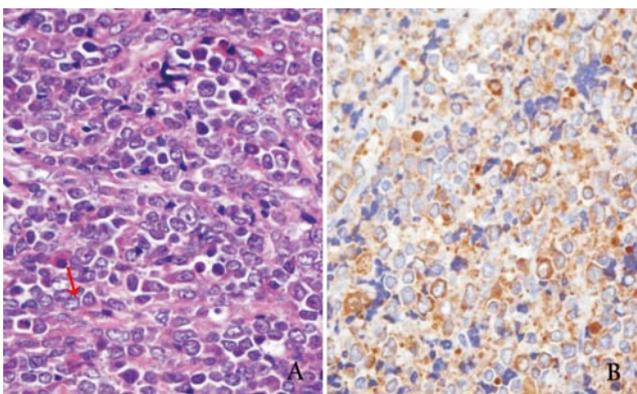


Figure 2: (A) Histopathological examination shows diffuse sheets of immature cells with large, vesicular nuclei (H&E stain). Note the immature eosinophils (arrow). (B) Immunohistochemistry shows tumour cells diffusely positive for myeloperoxidase stain.

Our patient presented again three months later with right eye massive proptosis and total loss of vision. A repeated CT scan of the brain and orbit showed a large lobulated mass measuring 104.5mm x 87.21mm x 77.25mm, occupying the right orbit (Figure 3). The tumour also extended into the right frontal sinus, right ethmoidal air cells, right sphenoid sinus, right maxillary sinus, right nasal cavity right infero-temporal fossa, and infiltrated the right pterygoid muscle. There was destruction of the bony orbital walls, with intracranial extension into the right temporal and frontal region, causing a mass effect to adjacent brain (Figure 4). CT scans of the thorax/abdomen did not reveal any other sites of involvement.

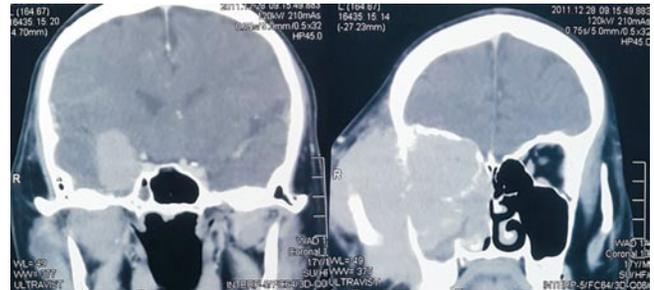


Figure 3: Coronal view of computed tomography of brain showing a large, lobulated mass occupying the orbit and involving the right frontal sinus, right ethmoidal air cells, right sphenoid sinus, right maxillary sinus, right nasal cavity and right inferotemporal fossa.

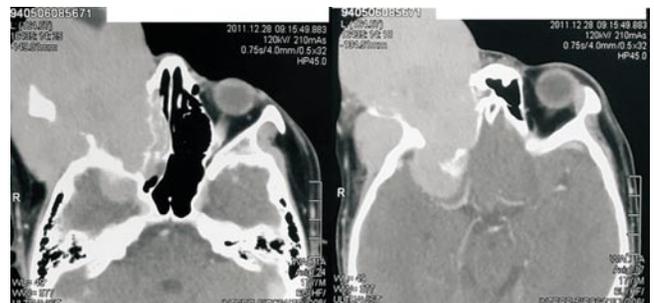


Figure 4: Axial view of computed tomography of the brain showing the mass extending into the intracranial region.

A repeat full blood picture and bone marrow aspiration showed 30% blast cells, which was consistent with the diagnosis of acute myeloid leukemia. Patient was started on chemotherapy and radiotherapy. Unfortunately, despite a reduction in tumour size, the patient passed away after the fifth cycle of treatment due to brain metastases.

3. Discussion

Myeloid sarcoma, also known as granulocytic sarcoma, is a rare presentation of acute myeloid leukemia which tends to affect bones (especially those of the skull, vertebral, paravertebral, and orbit) and soft tissue[4,5]. It has a slight male predominance and is more

commonly reported in the Middle East, Asia, and Africa[4,6]. As this condition is relatively more common in children, our patient had an unusual age of presentation.

Although myeloid sarcoma generally presents with bilateral proptosis (60%), our patient had only unilateral involvement[7,8]. The tumour may develop before, during or after occurrence of systemic leukemia[9]. In cases where myeloid sarcoma presents in non-leukemic patients, acute myeloid leukaemia usually develops within 10.5 months from the time of initial diagnosis[7]. The importance of continued surveillance cannot be overemphasized, as the haematological features of the disease may not be apparent on initial presentation.

In this case, the differential diagnoses included rhabdomyosarcoma, orbital lymphoma, neuroblastoma and eosinophilic granuloma. The most useful diagnostic test to differentiate these conditions is histopathology. Peripheral blood smear is essential in cases of sudden onset of proptosis or an orbital mass in children[10]. Besides that, bone marrow biopsy can be complementary, especially in cases where the blood smear is inconclusive.

Although there is currently no standard criteria for the treatment of patients with myeloid sarcoma, chemotherapy is generally the mainstay of treatment. In patients with CNS involvement, radiation therapy should be used in combination with chemotherapy[10].

The clinical outcome of myeloid sarcoma is poor and the mortality rate high[11]. The prognosis for life is especially dismal when the condition is associated with acute myeloid leukemia, with a 5-year survival rate of 20-30%[2].

Learning points:

- 1) Myeloid sarcoma is a rare, aggressive malignant neoplasm which tends to involve soft tissue and bones, especially those of the skull, vertebrae and orbit.
- 2) Myeloid sarcoma is usually seen in association with acute leukaemia; even in apparently healthy patients, acute myeloid leukaemia tends to develop within months of the initial diagnosis.
- 3) In young patients presenting with proptosis, a high index of suspicion for malignancy should prompt investigations like a peripheral blood smear and a biopsy to obtain a histopathological diagnosis.
- 4) As the haematological features of the disease may not be apparent on initial presentation, continued surveillance with

peripheral blood smear is essential, because delay in treatment may easily prove fatal.

Conflict of interest statement

The authors report no conflict of interest.

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