images and diagnosis

Orbital myeloid sarcoma presenting as massive proptosis

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A 10-year-old boy presented with right proptosis for 8 months. The eyeball was grossly pushed down, with diffuse corneal haze and non-reactive pupil. Systemic examination was normal. Previous investigations in another centre included a computerized tomography scan, which showed a well-defined enhancing retro-bulbar mass, a non-contributory fine needle aspiration cytology and a biopsy showing fibrocollagenous tissue with moderate lympho-monocytic infiltrate suggestive of non-specific inflammation. PET-CT scan revealed the presence of enlarged fluoro-deoxyglucose-avid cervical and mesenteric lymph nodes. Biopsy of the retro-bulbar mass was repeated in our centre. It showed fibrocollagenous and skeletal muscle tissue infiltrated by lymphoid follicles, dispersely lying lymphocytes and plasma cells, and admixed large atypical cells with vesicular nuclei, prominent nucleoli and scanty cytoplasm, strongly positive for myeloperoxidase, CD43 and CD99 immunohistochemistry. Hemogram was normal. Bone marrow aspiration/biopsy and CSF showed no evidence of acute myeloid leukemia. The child received chemotherapy in another centre and is in complete remission 6 months after completion.

A 10-year-old boy presented with right proptosis which had persisted for eight months. The eyeball was grossly pushed down, with diffuse corneal haze and non-reactive pupil. Systemic examination was normal. Previous investigations at another center included a computerized tomography scan, which showed a well-defined enhancing retro-bulbar mass (Figure 1), a non-contributory fine needle aspiration cytology and a biopsy showing fibrocollagenous tissue with moderate lympho-monocytic infiltrate suggestive of non-specific inflammation.

A PET-CT scan revealed the presence of enlarged fluoro-deoxyglucose (FDG)-avid cervical and mesenteric lymph nodes. Biopsy of the retro-bulbar mass was repeated at our center. It showed fibrocollagenous and skeletal muscle tissue infiltrated by lymphoid follicles, dispersed lymphocytes and plasma cells, and admixed large atypical cells with vesicular nuclei, prominent nucleoli and scanty cytoplasm, strongly positive for myeloperoxidase, CD43 and CD99 immunohistochemistry (Figure 2).

Hemogram was normal. Bone marrow aspiration/biopsy and CSF showed no evidence of acute myeloid leukemia (AML). The child received chemotherapy at another center and is in complete remission six months after completion.

DISCUSSION

Myeloid sarcomas are rare extramedullary manifestations of AML, with an estimated incidence of 2.5–8% in AML.[1] Orbital location is the most common.[2] Myeloid sarcoma may be metachronous or synchronous to leukemia. In the case of orbital location, bilateral proptosis is slightly more frequent than unilateral proptosis.[3]

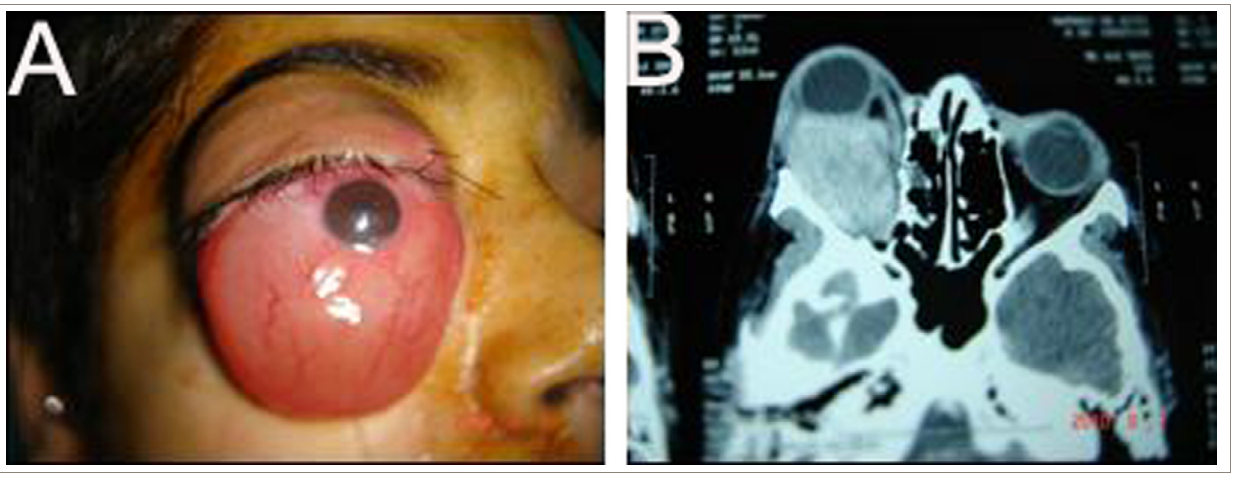

Figure 1. (A) Massive proptosis with eyeball pushed down grossly with restriction of motility, diffuse corneal haze and non reacting pupil. (B) CT scan of the orbit showing a well defined enhancing mass in the right retrobulbar region.

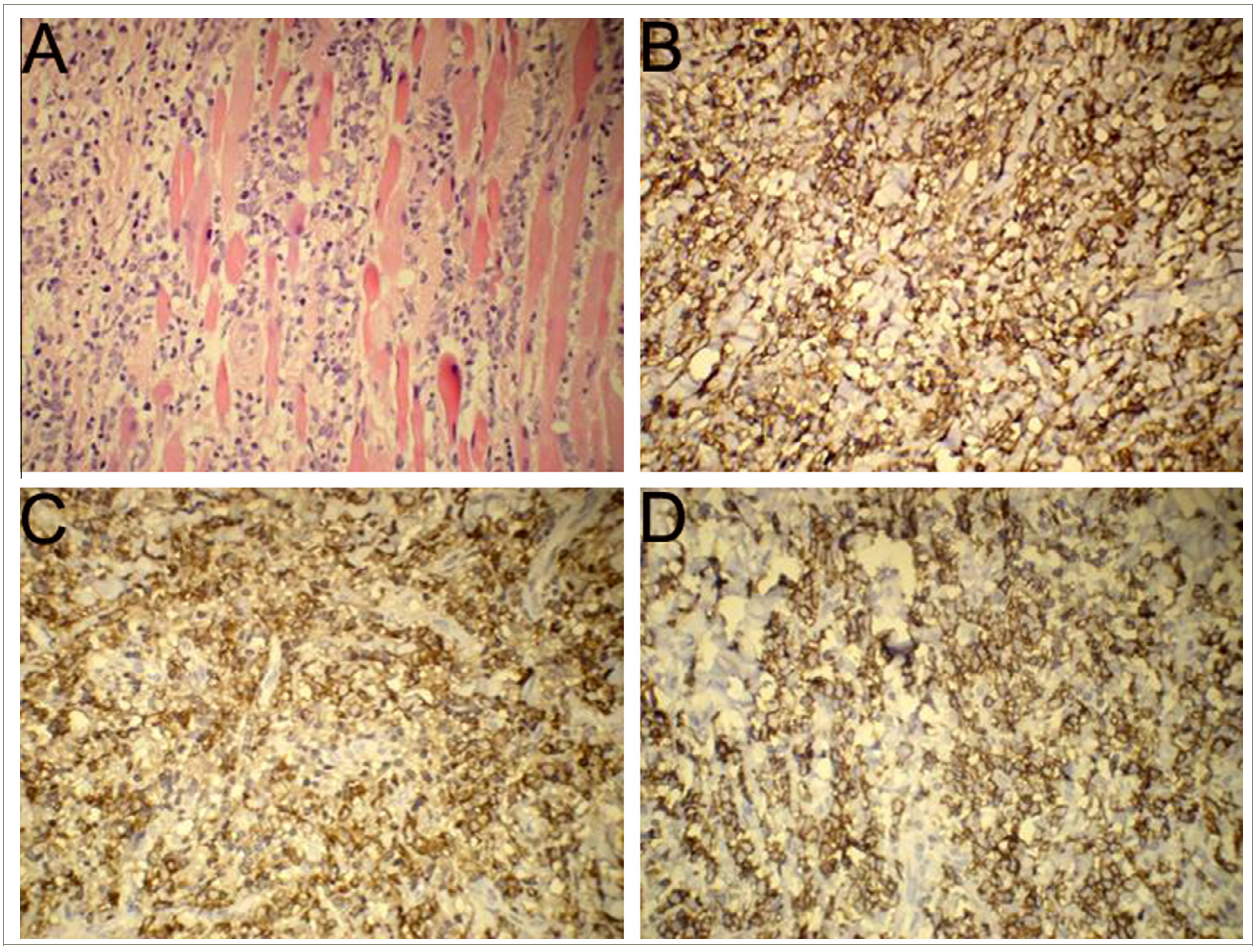

Figure 2. Orbital mass biopsy (A) Hematoxylin and eosin stain. (B) MPO, (C) CD 99 and (D) CD 43 immunostains are positive.

The prognostic implication of myeloid sarcoma is unclear. Turkish reports show that AML patients have poorer survival when orbital myeloid sarcoma is present.[4] However, US data showed that AML children with orbital myeloid sarcoma have significantly better survival than those with isolated

AML.[5] Possibly, orbital swelling leads to patients seeking medical attention and referral to specialized centers earlier, thus enabling early therapy and better outcome. In lower income countries, late referrals are common, leading to adverse outcome in most cancers, including AML.[6] Despite delayed diagnosis, our patient is alive and disease-free.

AML should be considered in the differential diagnosis of an orbital mass, even in the absence of typical leukemic symptoms.

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