Multifocal myeloid sarcomas: a rare presentation of AML

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DESCRIPTION

A 29-year-old previously healthy man presented to the hospital with a 2-month history of generalised weakness and swelling of the right eyelid. Physical examination showed ptosis of the right eye (figure 1A). In addition, he was anaemic and had had a decreased breath sound on the left lung. White cell count was 78 000 cells/mm^3, haemoglobin was 5.3 g/dL and platelet was 63 000 cells/mm^3. Peripheral blood smear revealed immature circulating cells (figure 1B). Flow cytometry was positive for CD34, CD117 and CD13, compatible with acute myeloid leukaemia (AML). Whole body CAT scan demonstrated infiltrative orbital mass at right eyelid and the superior aspect of right orbit (figure 1C,D) and a pleural-based left lower lung mass with associated left pleural effusion. Cytology of pleural fluid was positive for numerous blast cells. Biopsy of retro-orbital mass was deferred due to the concern of bleeding complication. He was immediately started on an induction chemotherapy regimen consisting of idarubicin and cytarabine. Circulating blasts rapidly declined and, correspondingly, eyelid swelling and pleural effusion were diminished.

Myeloid sarcoma (MS), also called extramedullary disease of AML, or chloroma, is a rare disease, occurring in only 2%–8% of patients with AML. It can present as an isolated extramedullary leukaemic tumour, concurrently with AML, or at relapse. Pathobiology of MS could be related to an aberrant homing signal for the leukaemic blasts allowing for clonal expansion outside of the more common bone marrow localisation. MS is most commonly reported in the skin, bone and lymph nodes. Owing to the rarity of MS, prognostic factors in these patients remain unknown. While it has been correlated with poor prognosis in adult, the superior outcome has been observed in paediatric AML with orbital and central nervous system MS.[1] Currently, recommended treatment is systemic therapy with induction regimen used in AML (combination of cytarabine and anthracyclines) in patients presenting with isolated MS or MS presenting concomitantly with AML, because of high failure rate associated with local treatment alone (88%–100% vs 42%).[2] Radiotherapy does not prolong overall survival but is commonly used to palliate symptoms. Orbital granulocytic sarcoma

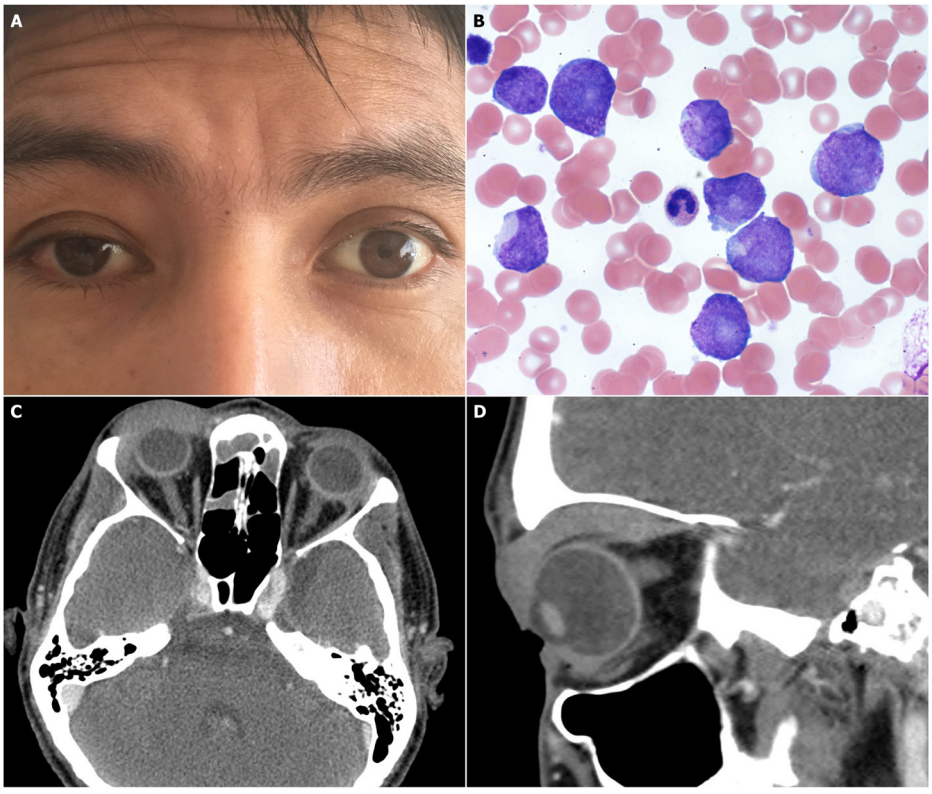

Figure 1 (A) A photograph at presentation demonstrating right eyelid swelling with ptosis. (B) Peripheral blood smear showing multiple blast cells. Axial (C) and sagittal (D) CT images demonstrating a right orbital mass.

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Learning points

- Myeloid sarcoma, a rare presentation of acute myeloid leukaemia (AML), can occur as an isolated extramedullary leukaemic tumour, concurrently with, or at relapse.
- Recommended treatment is systemic therapy with induction regimen used in AML (combination of cytarabine and anthracyclines).
- Orbital granulocytic sarcoma can progress rapidly resulting in permanent visual loss and is an oncological emergency that may require decompressive surgery or radiation.

occurs more commonly in paediatric rather than adult AML. Rapid visual loss from orbitopathy leading to compression of the optic nerve is an oncological emergency.[3] Decompressive surgery and radiation have both been used successfully.

Contributors Both authors were involved in the management of the case and the manuscript preparation.

Competing interests None declared.

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