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BRIEF COMMUNICATION

Orbital Tumour as Initial Manifestation of Acute Myeloid Leukemia: Granulocytic Sarcoma: Case Report

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Abstract We report orbital involvement as an initial manifestation of acute myeloid leukemia in a 57-year-old woman. The patient presented with painful proptosis and limited ocular motility. Orbital computed tomography revealed bilateral homogeneous masses. Orbital biopsy was performed on the right side; and histopathology disclosed a myelocytic tumour. Despite treatment using irradiation and chemotherapy, the patient died eleven months after presentation. There appear to be only a few previous reports of acute myeloid leukemia cases presenting with orbital involvement, and most cases occurred in children. This very rare condition has a poor survival prognosis, even with radiation treatment and chemotherapy.

Keywords Orbital granulocytic sarcoma · Acute myeloid leukemia

Introduction

Granulocytic sarcoma is a solid tumour composed of immature granulocytes.[1] Acute myeloid leukemia (AML) presenting with orbital involvement is unusual in older patients.[2–6] Histopathology and immunhistochemistry are required for diagnosis of the condition. Orbital involvement by AML carries a very poor survival prognosis, even with radiation treatment and chemotherapy.

Case Report

A 57-year-old deaf-mute woman presented in September 2003 after 3 days of painful proptosis (Fig. 1), with limited ocular motility, and conjunctival chemosis of the right eye. Previous medical history included arterial hypertension and hypothyreosis.

Best-corrected visual acuity was 0.2 (right) and 0.9 (left); intraocular pressure was 26 and 55 mmHg respectively. Hertel exophthalmometry readings were 31 mm and 20 mm (base 110 mm). On fundoscopy, choroidal folds were observed in the right eye. The left globe was normal.

Orbital computed tomography revealed a right sided well-defined, 2.9×2.4 cm, intraconal tumour of soft-tissue density and minimal contrast enhancement, with impression on the globe. A similar but smaller mass was found medially to the left eye. No bone destruction or extraocular muscles involvement were observed. Abdominal CT scan was normal. Peripheral blood abnormalities were not demonstrated.

Tentative diagnosis was orbital pseudotumour, or orbital lymphoma and chronic open-angle glaucoma. Systemic corticosteroid (prednisolone) therapy was started, with topical antiglaucoma drops (betaxolol, dorzolamide).

One week later, since proptosis was unchanged, a right orbital biopsy was performed through a superior eyelid-crease incision. Histolopathology showed tumourous infil-

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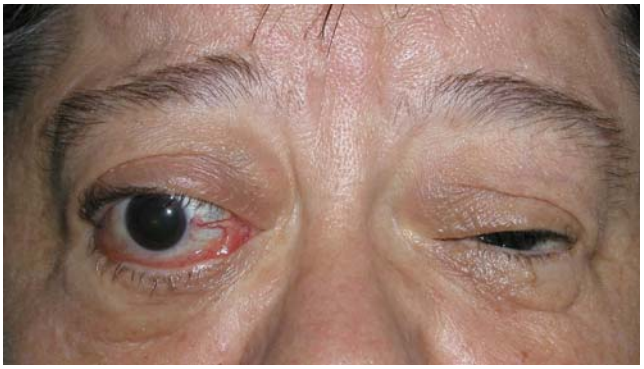

Fig. 1 Proptosis of the right eye

tration by undifferentiated, large cells with kidney-shaped nucleus, finely dispersed nuclear chromatin and clear cytoplasm, the mitotic activity was conspicuous. Immunohistochemistry was positive for CD13, CD33, CD34 (Fig. 2), CD45, CD117, cytoplasmatic myeloperoxidase; and negative for CD3, CD4, CD8, CD23, TdT (terminal deoxynucleoitidyl transferase), and cytokeratin. These findings indicated a myelocytic origin for tumour cells.

Bone marrow aspiration disclosed hypercellularity and elevated level of blast cells. No chromosomal changes were found.

Protrusion of the right eye and conjunctival chemosis progressed, disc swelling with venous dilatation developed. Two months after presentation irradiation of the right orbit and chemotherapy (idarubicin, cytosine arabinoside) were performed. Regression of proptosis was achieved; but a large, non-reacting pupil developed with white optic disc and occlusion of fundus vessels.

Relapse developed six months after initial presentation. Peripheral blood examination showed leukocytosis, monocytosis, anaemia, and thrombocytopenia, with elevated serum lactate dehydrogenase level. A chest CT scan revealed a mediastinal mass with compression of the trachea. Tumours developed bilaterally in the submandibular regions, and in the right breast and thigh. Final diagnosis was AML of FAB-type M4 (myelomonocytic AML) with extramedullar involvement. After chemotherapy (cytosine arabinoside, daunorubicin) and supportive therapy, regression was again achieved.

Ten months after presentation sudden visual loss developed in the left eye, with headache, nausea, skin rash and slight fever. Myeloblasts were observed in the peripheral blood. Cranial CT scan showed no evidence of an intracranial tumour. Fundus examination revealed papilloedema with haemorrhages and venous statis on the left side.

Eleven months after presentation, the patient died following an epileptic attack related to hypertensive crisis.

Comment

Granulocytic sarcoma (GS) is a solid tumour of immature granulocytes that can involve any part of the body: the head and neck region (e.g. orbit), body cavities (e.g. mediastinum), the trunk and limbs and subcutaneous tissue.[1]

The mass of immature cells formed in myeloid leukemia has been called chloroma, describing the typical greenish appearance (Greek *chloros*) caused by presence of the pigmented enzyme myeloperoxidase. The first published report of the tumour was in 1811 by Allen Burns who described a green tumour involving the orbit. Rappaport in 1966 renamed these tumours granulocytic sarcoma, since many tumours are colourless.

GS may present at any time during the course of the disease: concurrently with the leukemic phase, during remission or relapse, or occasionally, as in our case, it can precede the peripheral blood manifestation. If so, diagnosis may be difficult.[2] All types of leukemia may involve the orbit; however, orbital involvement is more common in acute than in chronic leukemia, and it occurs more often with the lymphoid than with the myeloid type.[3] There are only a few reports of AML presenting with orbital involvement, which is relatively scarce among orbital tumours. The majority of cases of orbital GS occur in young children; it is highly unusual in older patients.[4–6] Certain morphological subtypes of leukemia (FAB types M2, M4, and M5) are associated with a higher incidence of extramedullary myeloid tumours. Orbital infiltration in leukemia presents with proptosis, lid oedema and chemosis. Histopathology and immunhistochemistry are required for diagnosis. Orbital involvement by AML carries a very poor prognosis, even with radiation treatment and chemotherapy.

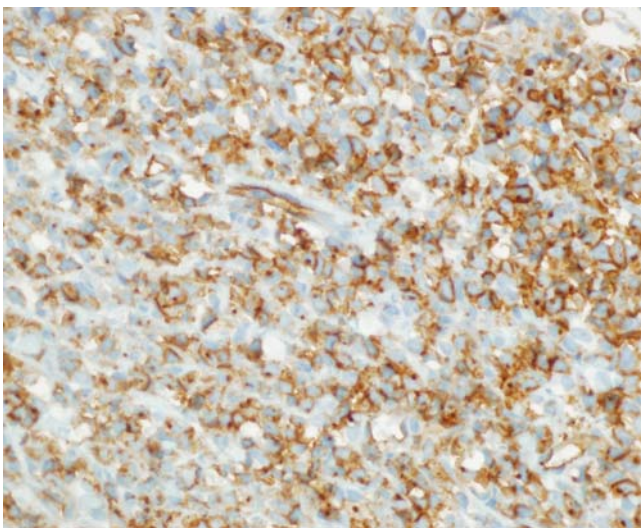

Fig. 2 Positive immunhistochemical staining for CD34

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