



Case report

Orbital myeloid sarcoma in adult mimicking nasolacrimal duct obstruction: A case report

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ARTICLE INFO

Article history:

Received 14 March 2016

Accepted 9 August 2016

Available online 12 August 2016

Keywords:

Myeloid sarcoma

Nasolacrimal duct obstruction

Orbital mass

Orbit

Granulocytic sarcoma

Chloroma

ABSTRACT

Purpose: To describe an orbital myeloid sarcoma in adult presenting with a swollen mass at inferomedial canthal area and epiphora which was misdiagnosed as nasolacrimal duct obstruction.**Observations:** A 45-year-old male presented with a swollen right lower eyelid around medial canthal area for 2 months with tearing for 6 month-period earlier. Eye examination demonstrated a high tear meniscus, slightly erythematous eyelid with palpable mass closed to the lacrimal sac along the inferior orbital rim. Computed tomography scan depicted infiltrative mass at the inferomedial aspect of right orbit with bony erosion, extended to adjacent paranasal sinuses. An incisional biopsy was performed. Histopathological study revealed soft tissue which was diffusely infiltrated by monotonous medium-sized round cells resembling blasts with lymphoglandular bodies, focally positive myeloperoxidase and negative lymphoid markers. The findings were consistent with myeloid sarcoma. No systemic involvement was found. The patient underwent chemotherapy and radiation without systemic leukemic disease progression.**Conclusions and importance:** Although orbital myeloid sarcoma is rare and difficult to diagnose, it can mimic nasolacrimal duct obstruction. We should consider this condition in our differential diagnosis.© 2016 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Myeloid sarcoma (MS) is an uncommon extramedullary manifestation of acute myeloid leukemia (AML). It can be found at any sites of body, particularly the skin, gastrointestinal tract, lymph nodes and bone.^{1,2} Orbital involvement is extremely rare,³ especially near lacrimal drainage area as the current report.^{4,5}

MS is often found concurrently in a patient with recognized AML, but it may also occur prior to the appearance of blood or bone marrow disease.¹ Neither systemic AML detected at the initial presentation nor progression occurred after 11-month follow-up period. So we herein reported a rare case of preceding orbital MS mimicking nasolacrimal duct obstruction.

2. Case report

A 45-year-old Thai male presented with history of tearing for 6 months followed by a swollen right lower eyelid around medial canthal area for 2 months. The patient had been diagnosed elsewhere with nasolacrimal duct obstruction. He denied pain, double vision, abnormal rhinorrhea, and nasal congestion. He had no previous history of trauma, surgery or systemic diseases.

Along ocular and adnexal examination disclosed right medial eyelid swelling and erythema, increased tear meniscus, and a palpable mass closed to lacrimal sac area without proptosis (Fig. 1A.). Full extra ocular motilities, normal visual acuity and fundoscopy of both eyes were demonstrated. Left eye was unremarkable. Lacrimal sac irrigation can definitely pass through to the nose without fluid reflux of the right eye. Somatic examination was unremarkable.

Orbital computed tomography (CT) depicted large, isodense with respect to extra ocular muscle, infiltrative mass at the inferomedial aspect of the right orbit with bony erosion at lamina papyracea of the medial orbital wall extended to adjacent paranasal sinuses (Fig. 1B). Based on the clinical findings and imaging results,

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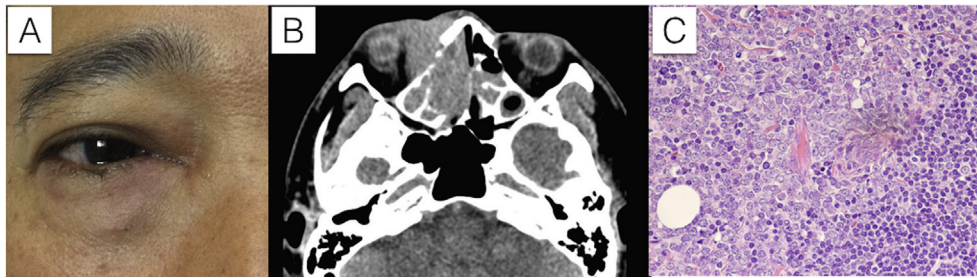


Fig. 1. Clinical photo compared with computed tomography scan and histopathologic findings. A: Right lower eyelid swelling and inferomedial mass pushing eyelid upward. B: Axial view of computed tomography scan showed an infiltrative mass at the inferomedial aspect of right orbit with bone erosion and extended to adjacent paranasal sinus. C: Histopathology showed diffuse infiltration by monotonous medium-sized round cells with blastic features.

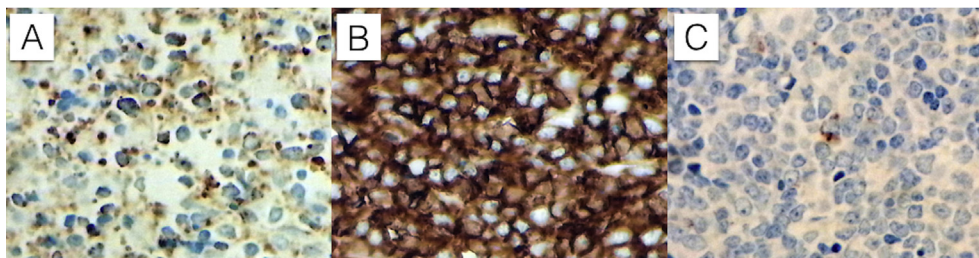


Fig. 2. Immunohistochemistry. A: Focal positivity for CD 34. B: Positivity for CD 43. C: Scattered positivity for myeloperoxidase.

the differential diagnoses included infiltrative tumors such as leukemia or lymphoma, metastatic tumors, and idiopathic orbital inflammation. Peripheral blood count revealed normal hemoglobin level with normal white blood cell count and differentiation. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were within normal range.

An incisional biopsy was performed via transconjunctival anterior orbitotomy of right lower eyelid. Histopathological study revealed a soft mass composed of monotonous medium-sized round neoplastic cells resembling blasts with lymphoglandular bodies (Fig. 1C). Immunohistochemistry of tumor cells showed diffuse positivity for CD43 and CD45 with focally positive myeloperoxidase (MPO) and CD34 (Fig. 2). The tumor cells are negative for CD3, CD20, CD68 or CD117. These findings were consistent with myeloid sarcoma. Bone marrow biopsy and CSF showed no evidence of acute myeloid leukemia. The patient underwent chemotherapy with cytarabine (Ara-C) and Idarubicin followed by totally 2400-cGy target orbital radiation with 12-month free of systemic leukemic disease.

3. Discussion

Myeloid sarcoma (MS) or granulocytic sarcoma or chloroma, an additional form of AML, is an extramedullary form of myeloid blasts.^{1,2} It has mostly been reported in children but rare in adulthood.^{6,7}

Orbital MS is uncommon but has been reported.^{1–3,8} To our knowledge, after reviewing the English literatures from Scopus, Pubmed database, we have found many cases of orbital MS presenting with several symptoms such as bilateral orbital tumors of myeloid sarcoma in a child by Shields JA. Et al⁹ and the AML patient with recurrent right upper eyelid mass by Phelps PO. Et al.¹⁰ The current case showed the inferomedial orbital mass with tear duct blockage-like symptom. Tearing in this case might be from the swollen of right lower eyelid which moved the eyelid upward and interfered the normal physiological pumping mechanism or might be from the mass effect over the lacrimal sac. Without syringing,

many ophthalmologists may jump to misdiagnose patients as nasolacrimal duct obstruction from the presenting signs and symptoms.

Computed tomography of MS showed homogenous, isodensity to muscle on non-enhancement, ill-defined mass which molds to the bones and surrounding structures.⁴ These findings can be found in several diseases, not specific for MS. Histopathology and immunohistochemistry study are necessary to confirm the diagnosis particularly in the cases without previous history of systemic leukemia. The specimen displayed diffuse infiltration of blasts with fine chromatin, and positive immunohistochemistry CD 34, 43, 45, MPO staining. Comparison with the results of Aggarwal E. Et al,³ there were three similar reliable markers. Minimal variation in immunohistochemistry markers^{7,11,12} might be from the differentiation of blast cells.

MS has been found in 2.5–9.1% of the patients with AML.³ Twelve-month free of systemic leukemic disease in this case might be from the immediate treatment and completion of the chemotherapy and radiotherapy protocol.

4. Conclusions

Orbital myeloid sarcoma without bone marrow involvement was uncommon in adult. It is important not to overlook this condition with an unusual presentation.

4.1. Patient consent

The study adhered to the principles of the Declaration of Helsinki and was exempted by the Institutional Review Board of the Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand (IRB No. 095/59). Patient provided written consent for publication of photographs and other potentially identifying information.

Conflicts of interest

No funding has been provided for this study.

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