

Isolated Conjunctival Myeloid Sarcoma as A Presenting Sign of Acute Leukemia

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Myeloid sarcoma is known as a tumor mass of myeloblasts or immature myeloid cells occurring in an extramedullary site. When ophthalmic areas are involved, it is usually located in the orbits and noted at or after the diagnosis of an underlying leukemia. We report a 38 year-old woman who had isolated conjunctival myeloid sarcoma without any other clinical signs and symptoms. Acute myeloid leukemia (AML) was diagnosed after a thorough examination. The image studies revealed no orbital or subcutaneous involvement. The patient had complete remission of AML after systemic chemotherapy. We reported this case to emphasize the unusual presentation of a conjunctival nodule of uncertain origin, particularly if it is salmon-pink and grows rapidly. The patient should undergo prompt evaluation for underlying hematological disease even if there are no ocular or systemic symptoms. (*Chang Gung Med J* 2010;33:334-7)

Key words: conjunctival myeloid sarcoma, acute myeloid leukemia

Myeloid sarcoma, known as granulocytic sarcoma, is an extramedullary solid tumor composed of immature myeloblasts and other granulocytic precursors, with or without an associated hematological malignancy.⁽¹⁾ A myeloid sarcoma most commonly arises in the setting of an underlying leukemia.⁽²⁾ Orbital granulocytic sarcoma usually arises from adjacent bone and less commonly, from the lacrimal gland or intraorbital muscles. The conjunctiva is occasionally involved.⁽³⁾ We herein describe a patient with a unilateral, isolated conjunctival nodule without any other ocular symptoms and signs which was the initial manifestation of acute myeloid leukemia (AML).

CASE REPORT

A 38-year-old healthy woman developed a mild tender, pinkish conjunctival nodule on the left eye

with progressive enlargement for 3 weeks. One week before referral to our hospital, she received a treatment under the impression of a hordeolum at a private clinic. At presentation, a 18 mm x 10 mm salmon-pink conjunctival nodule was noted in the inferior fornix of the left eye (Fig. 1). There was no vision disturbance, photophobia, chemosis, proptosis, or lid swelling. The ocular motility was intact. The remainder of the ocular examination, including slitlamp biomicroscopy and ophthalmoscopy showed nothing abnormal. She denied any other ocular or systemic disease. An excision biopsy was performed smoothly, although hemostasis was a problem during the operation. One week later, the histopathologic report showed myeloid sarcoma (Fig. 2). She was referred to oncology specialists for further management. On examination, a few petechiae on the central trunk and four limbs were noted. No other orbital nodule was found on computed tomography images.

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Fig. 1 A 18 mm x 10 mm salmon-pink conjunctival nodule is noted in the inferior fornix of the left eye.

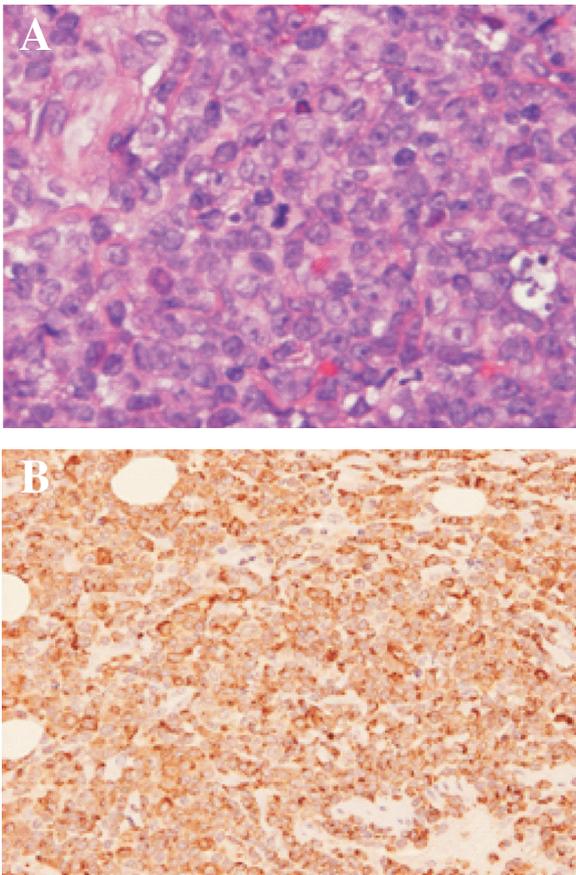


Fig. 2 The histopathologic section. (A) Biopsy of the nodule shows myeloid (granulocytic) sarcoma. Normal tissue is replaced by sheets of abnormal leukemia cells. (stains, hematoxylin and eosin; original magnification, x 400) (B) Myeloperoxidase immunostaining shows a positive reaction (intense brownish red reaction product), confirming that the cells are leukemic myeloblasts. (original magnification, x 200)

The initial peripheral blood cell count revealed leukocytosis with 76% blast cells, anemia and thrombocytopenia. The results of bone marrow aspiration were compatible with AML myeloblastic with maturation (M2) (Fig. 3). The bone marrow karyotypic study revealed no 8; 21 translocation. The patient received chemotherapy with cytarabine (ara-C) and an anthracycline drug. The induction treatment resulted in complete haematological remission and bone marrow transplantation had not been performed as of the last follow-up at 10 months. After biopsy the conjunctival nodule resolved and the lesion was stable at the follow-up till now.

DISCUSSION

Myeloid sarcoma is relatively uncommon in the western hemisphere, but is more prevalent in the Middle East, Asia, and Africa.⁽⁴⁾ No significant sex predominance is apparent. The tumor is found more than twice as often in children as in adults. Myeloid sarcoma can involve any part of the body, but the most common sites of occurrence are the orbits and subcutaneous soft tissues. Other locations that have been described include the paranasal sinuses, lymph nodes, bone, spine, brain, pleural and peritoneal cavities, breasts, thyroid gland, salivary glands, small bowel, lungs, and testes.⁽⁵⁾ In most instances, orbital myeloid sarcoma occurs in young children.⁽⁴⁾ An orbital myeloid sarcoma usually arises from adjacent

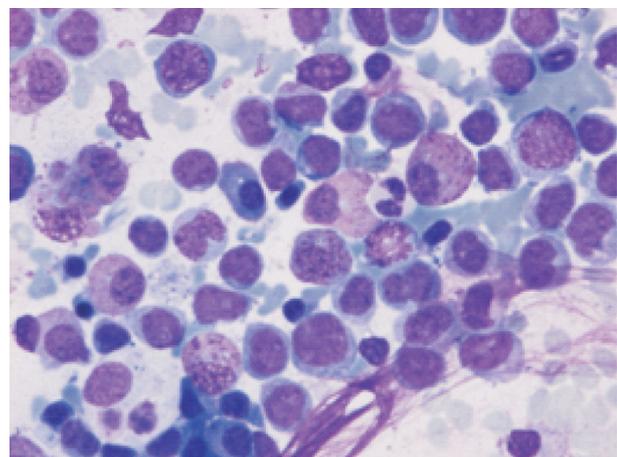


Fig. 3 The bone marrow aspirates show acute myeloid leukemia with maturation. (stain, Liu; original magnification, x 1000)

bone and, less commonly, from the lacrimal gland or intraorbital muscles. The conjunctiva is rarely involved.⁽³⁾ However, Lee et al. and Hon et al. reported a tumor located at the conjunctival fornix or found as an isolated conjunctival mass in patients with a history of AML.^(6,7) Fleckenstein et al. reported a patient with a history of myeloproliferative syndrome who presented with bilateral chemosis, markedly distended conjunctiva, as an initial symptom of AML.⁽⁸⁾ Tumors in the conjunctiva, muscle, testes, and bladder tend to cause diffuse thickening rather than discrete masses.⁽³⁾ However, our patient presented with an isolated nodule in her left conjunctiva before the diagnosis of underlying leukemia.

Myeloid sarcoma is usually found in patients with both acute and chronic myelogenous leukemia. However, it also can be found in association with other myeloproliferative disorders including myeloid metaplasia, myelofibrosis, polycythemia vera, and chronic eosinophilic leukemia.^(5,8) A single myeloid sarcoma is present in about 2% of patients with AML. A myeloid sarcoma associated with AML usually affects no more than two sites.⁽²⁾ It is well known that AML can present initially with orbital involvement, before the diagnosis of the underlying leukemia.^(4,9) The rate of occurrence is approximately 3-9% in patients with AML.⁽⁵⁾ Myeloid sarcomas are most common in certain subtypes of AML, in particular M5a (monoblastic), M5b (monocytic), M4 (myelomonocytic), and M2 (myeloblastic with maturation).⁽⁴⁾ Our patient's case was compatible with AML M2. On autopsy, approximately 30% of eyes in patients with fatal leukemia show ocular involvement, mainly leukemia infiltrates in the choroids. Also, 42% of newly diagnosed cases of acute leukemia show ocular findings, especially intraretinal hemorrhage, white-centered retinal hemorrhage, and cotton-wool spots. Retinal hemorrhage is most likely to occur in patients who have combined anemia and thrombocytopenia.⁽¹⁰⁾ Interestingly, our patient did not have retinal hemorrhage or ocular symptoms, although she had both anemia and thrombocytopenia.

In summary, we reported a case of AML presenting as a unilateral, isolated conjunctival myeloid sarcoma. Neither hematological study nor image sur-

vey was done before an assumed simple lesion excision biopsy. We could find no other reports of conjunctival myeloid sarcoma presenting before diagnosis of acute hematological diseases. We must be alert to this rare neoplasm as a presenting sign of acute leukemia. As the lesion was first misdiagnosed as a hordeolum, we should consider the possible differential diagnosis, including benign lesions such as phlyctenulosis and pyogenic granuloma, or malignant lesions such as lymphoma or squamous cell carcinoma.

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急性骨髓性白血病： 以單一結膜骨髓樣肉瘤爲先發症狀之病例報告

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骨髓外區域的急性骨髓性白血病細胞形成瘤狀物，稱之爲骨髓樣肉瘤。該腫瘤若在眼部被發現，通常是以侵犯眼窩爲表徵，極少數伴有結膜病灶，且其被發現的時間點大多在已知患有血液疾病的同時或之後。本文報告一位 38 歲女性病患，在診斷爲急性骨髓性白血病之前，以不尋常之單眼單一結膜病灶爲先發症狀，且該病患電腦斷層檢查顯示無併發常見的眼窩或皮下組織侵犯。該患者經全身性化學治療後，急性骨髓性白血病達完全緩解。本文旨在強調臨床上，對一生長急速的結膜病灶，除進行簡單地局部麻醉、切除併送病理診斷，並應仔細全身理學、血液等檢查，及電腦斷層檢查，尤應特別注意是否有其他血液疾病存在之可能。(長庚醫誌 2010;33:334-7)

關鍵詞：結膜骨髓樣肉瘤，急性骨髓性白血病

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