Short communication

Conjunctival myeloid sarcoma in acute myeloblastic leukemia-M1

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ABSTRACT

Clinical case: A 69-year-old man with a history of acute myeloblastic leukemia-M1 presented with bilateral conjunctival injection. Ophthalmological examination revealed lesions located at the upper tarsal conjunctiva of the right eye and lower tarsal conjunctiva of both eyes. Histological and immunohistochemical studies confirmed conjunctival myeloid sarcoma (MS). The patient died due to multiorgan failure three months later.

Discussion: Extramedullary recurrence of leukemia can appear as an ocular manifestation that brings about a recurrence of the leukemia, leading to a poor prognosis.

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Sarcoma granulocítico conjuntival en leucemia mieloblástica aguda M1

RESUMEN

Caso clínico: Varón de 69 años diagnosticado de leucemia mieloblástica aguda M1 en remisión completa, remitido a Oftalmología por cuadro de ojo rojo no doloroso. Biomicroscópicamente se observan lesiones sobrelevadas asalmonadas en conjuntiva tarsal superior de ojo derecho e inferior de ambos ojos. Mediante estudios histológicos e inmunohistoquímicos se confirma recidiva extramedular en forma de sarcoma mieloide en conjuntiva. El paciente fallece a los 3 meses de un fallo multiorgánico.

Discusión: La recidiva extramedular de una leucemia puede presentarse como una manifestación ocular que conlleva una recurrencia de la leucemia, siendo esta de mal pronóstico.

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Introduction

Acute myeloblastic leukemia (AML) is an aggressive hematological neoplasia with a mean survival rate of 19.5% after 5 years. There are 8 types of AML described according to the Franco-American British Classification (FAB), from M₀ to M₇. Autopsy studies refer ophthalmic involvement of between 50% and 90% in leukemia patients. The most frequent leukemia infiltration is in the retina and choroids.

M5 is characterized by the presence of one or more tumor masses comprised by immature myeloid masses at extramedullar locations. It can predict or express together with acute or chronic myeloblastic leukemia or with myelodysplastic syndromes.

Clinic case

A male, aged 69, presented with a personal history of AML M1 (myeloblastic without differentiation) in complete remission (CR) which started 11 months earlier after chemotherapy treatment according to the PETHEMA-LAM 99 protocol. In February 2008 the patient was admitted due to breathing condition with catarrh and significant nasal congestion without response to antibiotics. In addition, the patient exhibited eritematous papules in the back, supraclavicular and submaxillary adenopathies.

The patient was referred by the hematologist due to red eye with 2 days evolution and without any other ocular symptoms. The exploration gave a corrected visual acuity of 20/20 in right eye (RE) and 18/20 in left eye (LE). Ocular reflexes, motility and tonometry were within normal ranges. Anterior pole biomicroscopy showed in the RE a fleshy, salmon-colored raised lesion in the upper (Fig. 1) and lower tarsus. In the LE we found a similar lesion in the lower tarsal conjunctiva (Fig. 2). The ocular fundus assessment did not reveal alterations.

The possibility of leukemic involvement of the conjunctiva was considered and a biopsy of the lesion was carried out, the histological study of which revealed a diffuse proliferation of myeloid habit cells arranged under the conjunctival epithelium which were positive at the immunohistochemical level for myeloperoxidase and CD43 (Figs. 3 and 4), and negative for B and T cell markers (CD20, CD3, CD5) as well as for...
TdT, CD34 and CD117. Accordingly, the case was diagnosed as granulocytic sarcoma.

New biopsies confirmed the involvement of the nasal mucosa and the skin. With these findings, the diagnosis was of MS involving the conjunctiva, the skin and the nasal mucosa. However, in a peripheral blood and a medulla ossea sample relapse was not observed and therefore it was concluded that at this level the CR persisted. Lumbar puncture as well as the rest of explorations gave normal results.

After the diagnosis of extra-medullar relapse in the form of AML MS, second-line treatment was initiated according to the FLAG-IDA protocol with a good response by the infiltrating lesions. The patient died in May 2008, 3 months after the MS diagnosis due to a multiorgan failure.

**Discussion**

Ocular involvement by leukemia is well known, with the most frequent being retinal hemorrhage usually due to the systemic effects of the disease such as leukocytosis, hyperviscosity or anemia. In turn, the retina is the most frequently involved ocular structure due to it being directly affected by leukemia cells, up to 30%.

In AML, the infiltration of the conjunctiva rarely occurs. It is more frequent in the myelomonocytic M4 and monocytic M5 forms. In the other types of AML, only 4 cases of conjunctival infiltration have been described, all of them located in the bulbar conjunctiva. Two cases occurred in adults and a further 2 in children, with 50% of these cases being bilateral. The appearance of the lesion was nodular although one was described as a "salmon-colored fleshy mass." As far as we know, this is the first case in which MS is diagnosed as a salmon-colored mass in the tarsal conjunctiva with bilateral expression in an adult in CR of M1 AML, even though our patients exhibited MS simultaneously in other locations, with the most frequent being the periosteum and the skin including the subcutaneous cellular tissue according to other studies. Up to 25% of MS are found in the orbit, which is the most frequent location in children with AML. Occasionally, it appears in several areas at the same time as in the case presented here. It usually indicates that the disease is at an advanced stage.

By way of conclusion, we emphasize that leukemic infiltration of the conjunctiva in the context of acute leukemia is an ominous prognostic sign. In our case, matching the mean descriptions of the literature, survival did not exceed 3 months. Said infiltration can be a sign of presentation of leukemia or relapse thereof and usually does not involve the eyesight.

**Conflict of interests**

The authors have not declared any conflict of interests.

**REFERENCES**