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A pitfall diagnosis of orbital tumor

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A 42-year-old Moroccan man presented with a 9-month history of progressive right orbital proptosis. Although the patient did not complain of pain in the eye, he had discomfort due to proptosis.

No vision disturbance, photophobia, or chemosis was observed. The ocular motility was intact. The remainder of the ocular examination, including slit lamp biomicroscopy and ophthalmoscopy showed nothing abnormal. His physical examination was unremarkable without evidence of lymphadenopathy or organomegaly. The orbital computed tomography (CT) scan revealed a homogeneous mass of 20×30×40 mm in the right orbit (**Figure 1**). The lesion showed enhancement with contrast agents. Mild destruction of the lesser wing of the sphenoid was seen with brain involvement.

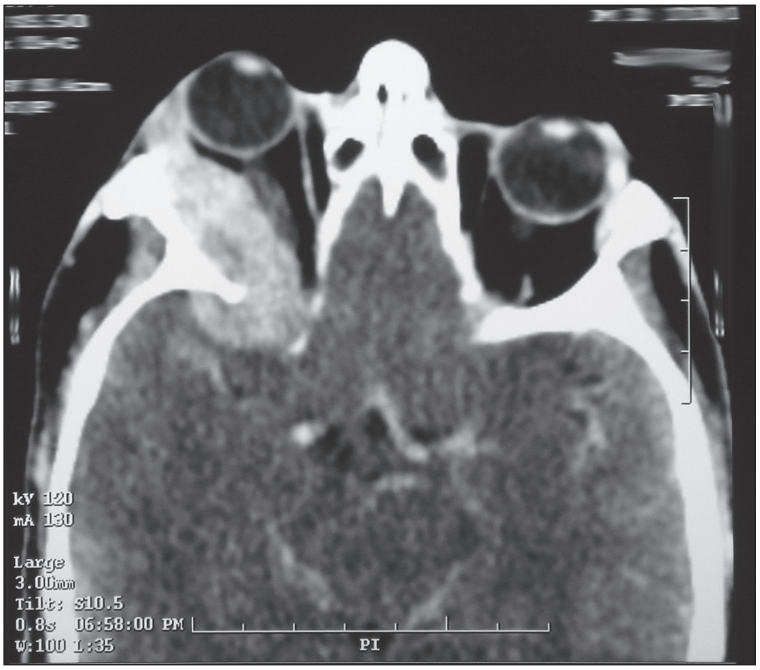

Figure 1. Orbital computed tomography (CT) scan revealing a homogeneous mass in the right orbit.

The differential diagnosis included lymphoma, leukemia, metastatic lesion, and diopathic orbital inflammation on the basis of clinical findings and imaging study results. A complete blood count was normal. Serum levels of antinuclear antibody were normal. The histopathological examination of the tumor biopsy showed a dense fibrous proliferation, with collagen deposition having numerous inflammatory cells. Idiopathic orbital inflammation was suspected, and the patient was treated with steroids. A follow-up examination of 4 months revealed worsening in his vision due to exophtalmous.

He was later transferred to our medical department for further investigations and management. On admission, the patient was experiencing general malaise and fever. At the time of admission, the medical examination revealed a febrile patient without peripheral lymphadenopathy.

A laboratory evaluation revealed the following results: hemoglobin level of 10 g/dL, platelet count of 95 000/L, and white blood count of 3800/L with 70% neutrophils, 25% lymphocytes, 4% monocytes, and 1% eosinophils. The peripheral smear study was normal without leukemic blasts or Auer rods. Hematologic evaluation including the cytologic analysis of bone marrow aspiration revealed M2 acute myelogneous leukemia (WHO Classification AML with t[8;21][q22;q22]).

According to our institutional protocol, the patient was treated with cytosine arabinoside (200 mg/[m^2.d] for 7 days) and daunorubicin (50 mg/[m^2.day] for 3 days) and achieved remission. The orbital tumor regressed significantly 10 days after the initiation of chemotherapy.

Granulocytic sarcoma (GS) is a rare tumor composed of immature granulocytic cells at various levels of differentiation in extramedullary sites.[1] In published reports, it is known under a variety of names including chloroma, extramedullary myeloid tumor, myeloblastoma, and monocytic sarcoma. GS may occur in a variety of tissues. The leukemic infiltrates develop preferen-

tially in bones; however, orbital involvement has been reported in 24% of cases.[2] GS occurs most often with acute myeloid leukemia (AML) or chronic myeloid leukemia in the accelerated phase. The majority of GS cases were associated with AML-M2.

The prognosis of GS is not optimistic. Primary GS may transform to acute leukemia in 8 days to 28 months.[3] GS is difficult to diagnose and is often initially misdiagnosed, particularly, if the tumor does not occur in the context of AML, such as in our patient. Pathological, radiological, and clinical confusion with lymphoma may occur. Most of the cases reported with GS being the presenting sign of leukemia are described in the pediatrics or young adults.[4] Radiologically, the CT findings are not sufficiently characteristic. The orbital lesions are usually localized soft tissue masses with homogenous enhancement.[5] GS are seen to respond very well to chemotherapy, radiotherapy, or both.

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