

Orbital Mass Secondary to Precursor T-Cell Acute Lymphoblastic Leukemia

A Rare Presentation

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We describe a 40-year-old woman with a history of precursor T-cell acute lymphoblastic leukemia who developed an orbital mass associated with diffuse infiltration of the paranasal sinuses. The clinical and radiologic findings suggested an orbital abscess. Examination of orbital and ethmoid sinus biopsy specimens revealed relapse of precursor T-cell acute lymphoblastic leukemia. Although orbital involvement by granulocytic sarcoma (also known as extramedullary myeloid cell tumor and chloroma) with or without concurrent acute myeloid leukemia is well described in the literature, similar presence of acute lymphoblastic leukemia of either precursor T-cell or B-cell lineage is rare.

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Orbital disease secondary to acute lymphoblastic leukemia (ALL) is rare, particularly in the adult population. Despite several reports that describe the clinical and radiologic features of orbital extramedullary myeloid cell tumor (also known as granulocytic sarcoma or chloroma) secondary to myeloid leukemias,¹⁻³ we were able to find only 1 case report of orbital and choroidal disease secondary to ALL in an infant.⁴ We report the case of a 40-year-old woman with an orbital mass and diffuse infiltration of paranasal sinuses secondary to precursor T-cell ALL. The radiologic findings in this patient were not typical for the previously described features of orbital chloroma,⁵⁻⁸ but were more consistent with an orbital abscess. The clinical, radiologic, as well as histopathologic features of this neoplastic process are described in the present article.

REPORT OF A CASE

A 40-year-old woman with a history of precursor T-cell ALL with mediastinal involvement was admitted to the hospital

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with a fever. She had a computed axial tomography scan of the head and neck, which showed opacification of the maxillary sinuses and ethmoidal sinuses, as well as a subperiosteal orbital "abscess." An ophthalmological consult was requested.

MEDICAL HISTORY

The patient was diagnosed with precursor T-cell ALL approximately 1 year prior to this hospitalization. She had experienced multiple relapses of her leukemia in the preceding year, and had been treated with various chemotherapeutic regimens. She was being considered for allogeneic bone marrow transplantation. Four weeks prior to this hospitalization, she underwent uneventful sinus surgery for pansinusitis. The cultures from the ethmoidal sinuses at that time grew coagulase-negative *Staphylococcus*. She was treated with systemic antibiotics and responded well.

CLINICAL FEATURES

The patient was found to be alert, oriented, and in no distress. She denied any ocular or periocular symptoms or headaches. Her visual acuity was 20/20 OU. The

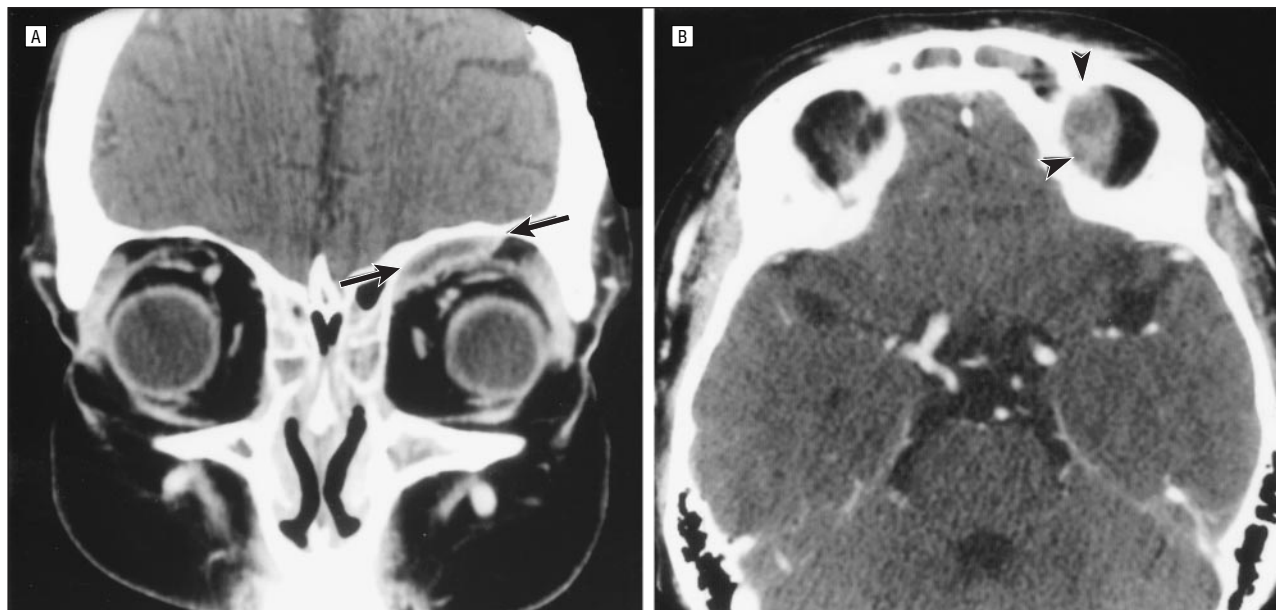


Figure 1. Enhanced computed tomography images reveal the left superior medial orbital mass. A, Coronal computed tomography image shows the left extraconal, marginally enhancing mass (arrows). B, An axial computed tomography image also demonstrates the superior orbital mass (arrowheads).

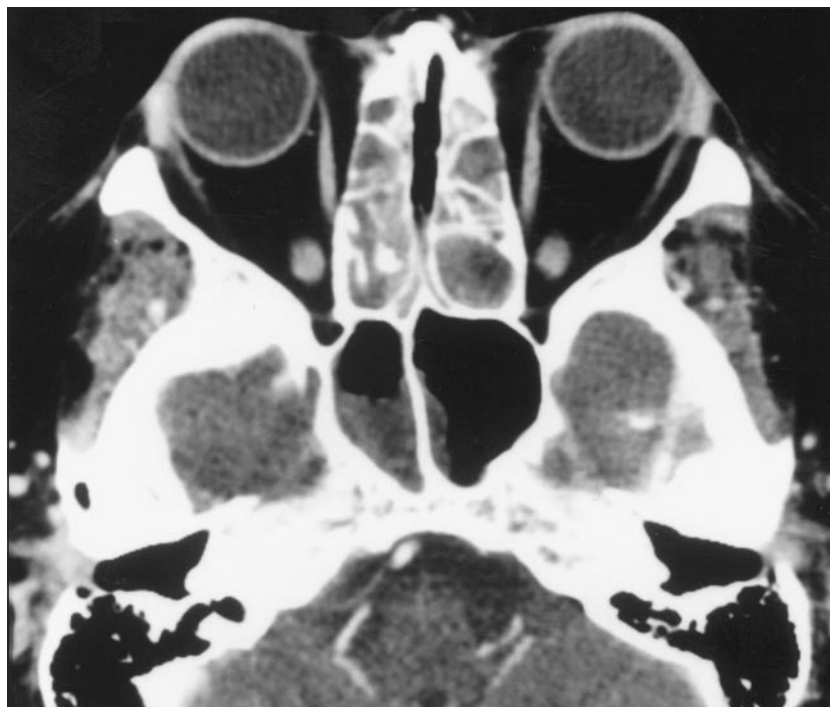


Figure 2. Axial enhanced computed tomography images demonstrate sinus disease. Mucosal thickening is seen in the sphenoid sinuses, and complete opacification of the ethmoid air cells is present.

external examination revealed quiet globes. The ocular adnexal examination was positive for mild edema and echymosis of the left upper eyelid. The results of the extraocular motility examination were normal with no diplopia. There was minimal to no proptosis, although Hertel exophthalmometry measurements were not done during the bedside examination. Confrontation visual fields were

full. The pupils reacted normally to light and accommodation, and there was no afferent pupillary defect. A penlight examination revealed a normal cornea and anterior chamber in each eye, with no evidence of inflammatory signs. Intraocular pressures with a TonoPen (Mentor, Norwell, Mass) measured 18 mm Hg in each eye. A dilated fundus examination revealed cup-disc ratios of 0.2 bilater-

ally with sharp disc margins. The central and peripheral retina appeared normal in each eye.

The patient's white blood cell count was found to be $63 \times 10^9/L$ on admission to the hospital.

COMPUTED TOMOGRAPHIC SCAN FEATURES

A low-density, extraconal mass was found in the superior medial left orbit with marginal enhancement (peripheral enhancement with central nonenhancement) consistent with an orbital abscess. There was no bone destruction (**Figure 1**). There was considerable opacification of the frontal, maxillary, and ethmoid sinuses (**Figure 2**).

HOSPITAL COURSE

The patient was taken to the operating room where explorations of the left orbit and endoscopic sinus surgery were undertaken simultaneously. A superonasal orbitotomy (Lynch incision) was performed, and a collection of brownish gray material was found in the subperiosteal space. This material was friable and difficult to excise, but as much of this material as possible was removed. Bacterial (anaerobic and aerobic) and fungal cultures were obtained. The tissue was also sent for histopathologic diagnosis.

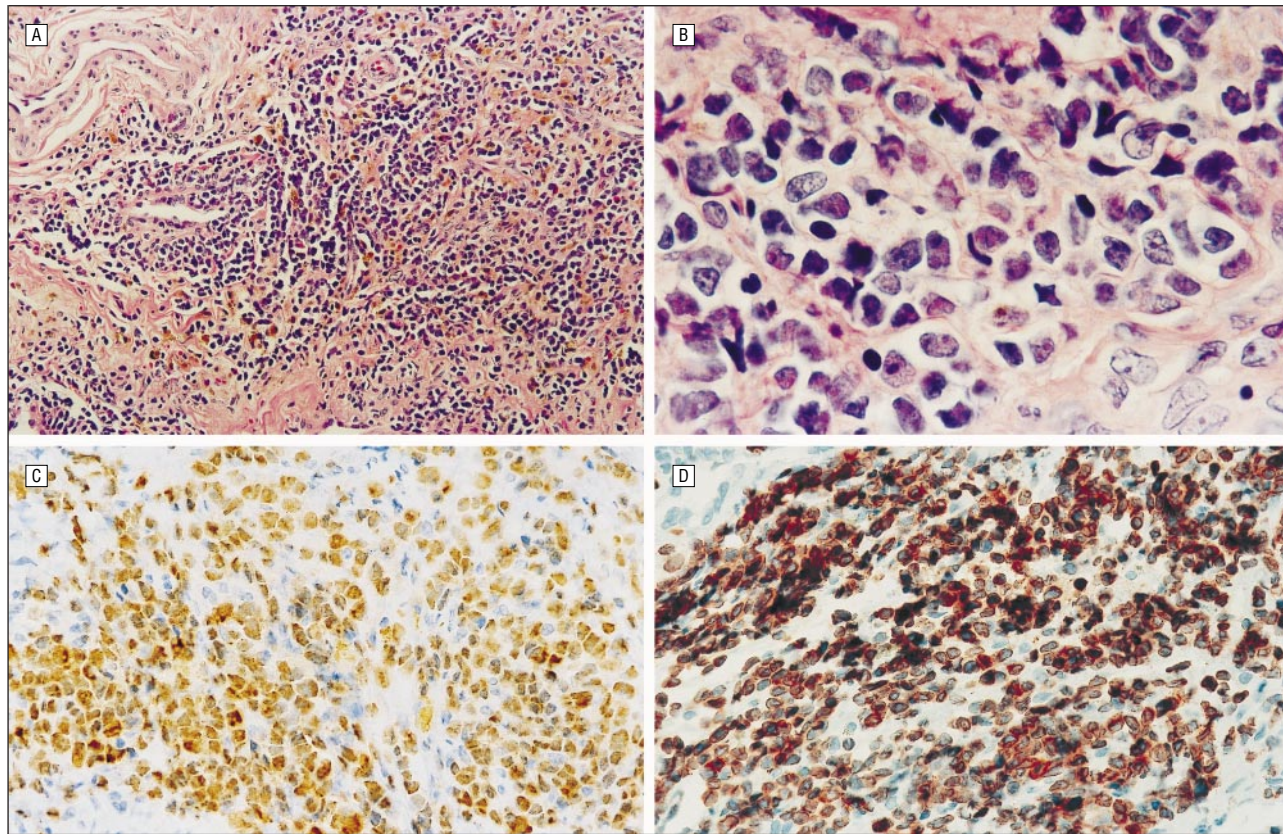


Figure 3. Precursor T-cell acute lymphoblastic leukemia involving the orbit and ethmoid sinuses. A, Low-power view of lymphoblasts intersecting between collagen bundles in the orbit. The pigment in the field is hemosiderin secondary to hemorrhage (hematoxylin-eosin, original magnification $\times 200$). B, High-power field of lymphoblasts with finely granular blastic chromatin and irregular nuclear contours (hematoxylin-eosin, original magnification $\times 1000$). C and D, The lymphoblasts are strongly positive for TdT (C) and CD3 (D) and were negative for CD20 (not shown) (immunoperoxidase with hematoxylin counterstain, original magnification $\times 400$).

The cultures from both orbital and sinus specimens were negative for bacteria and fungus.

The patient's high white blood cell count was treated with plasmapheresis. She then received systemic chemotherapy as recommended by her hematologist. She subsequently underwent allogeneic bone marrow transplantation. At the time of this report, she remains asymptomatic from an ocular standpoint, and remained free of disease for 3 months following her bone marrow transplantation.

HISTOPATHOLOGIC FEATURES

Biopsy specimens from the left orbit, left ethmoid sinus, and right ethmoid sinus were obtained for examination. Histologically, all biopsy specimens were similar, and the orbital specimen is shown in **Figure 3A** and Figure 3B. Within the orbital connective tissue and sinusoidal mucosa, aggre-

gates of lymphoblasts were present intersecting between collagen bundles (Figure 3A). At high power, the lymphoblasts had irregular nuclear contours and finely granular, blastic nuclear chromatin without prominent nucleoli (Figure 3B). Mitotic figures were easily identified.

Immunohistochemical studies were performed using fixed, paraffin-embedded tissue sections of the left ethmoid sinus tissue. The lymphoblasts were positive for terminal deoxynucleotidyl transferase (Figure 3C) and the T-cell antigen CD3 (Figure 3D) and were negative for the B-cell antigen CD20 (not shown). These results supported the diagnosis of precursor T-cell ALL.

COMMENT

We report an unusual orbital and paranasal sinus presentation of ALL, which appeared radiologically as an abscess. To our knowl-

edge, there is only one other report of a leukemic infiltration of the orbit secondary to ALL.⁴ That case was in a 6-month-old boy, and the orbital involvement was diffuse and also associated with intraocular (choroidal) infiltration. Radiologically, the orbital lesion in our patient was a well-circumscribed, hypodense mass with peripheral enhancement. This is in contrast to granulocytic sarcomas, which are usually homogeneously enhancing masses.^{5,8} Given our patient's previous history of sinus infection and the radiologic appearance consistent with an orbital abscess, the most likely diagnosis was an infectious process in the sinuses and in the orbit. However, histopathologic evaluation of the biopsy specimens confirmed the diagnosis of leukemic infiltration secondary to precursor T-cell ALL.

Unlike the orbital mass described in this patient, the majority of head and neck or orbital masses secondary to leukemia are of the my-

eloid lineage. Extramedullary myeloid cell tumors (also known as granulocytic sarcoma, chloroma, or myeloblastoma) are the most common form of leukemic infiltration involving the orbit.⁹ Orbital chloromas may occur in various forms, especially in children. Focal masses may arise intraconally or extraconally and may be bilateral.¹⁰ Generally, the diagnosis of leukemia is known, but as with leukemic masses elsewhere, they may precede the onset of systemic disease.

Before modern chemotherapy, central nervous system involvement by leukemia was common, affecting approximately 75% of patients with ALL and up to 50% of those with acute myelogenous leukemia.⁹ Extramedullary disease secondary to ALL has been reduced with chemoprophylaxis and more advanced therapy for this disease. Our patient had several relapses of her ALL prior to this presentation and was in blast

crisis while she developed the leukemic infiltration of the orbit and paranasal sinuses. Localized leukemic nodules in the orbit may respond to external beam radiation therapy, but more commonly, since they are associated with systemic blast crisis, chemotherapy or bone marrow transplantation may be necessary.

Leukemic infiltration should be considered on the differential diagnosis of a marginally enhancing mass of the orbit, even in the presence of diffuse paranasal sinus disease, which may look suspicious for an infectious process radiologically.

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