

An Unexpected Presentation of Pediatric Acute Lymphoblastic Leukemia: Maxillary Sinus Mass

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Dear Editor;

Extramedullary (EM) involvement of acute lymphoblastic leukemia (ALL) other than the central nervous system (CNS) and testis is an extremely rare condition. Breast, skin, kidney and parotis involvement have rarely been reported [1–6]. Regarding the pathologic mechanism yielding EM tumor development, both a local transformation of histiocytes to blasts particularly in myeloid leukemia and a ‘homing’ phenomenon of leukemic cells in determined tissues have been discussed [7–8]. Extramedullary involvement usually points to poorer prognosis [9]. In the treatment; radiotherapy and surgery is an option either separately or as a combination besides chemotherapy, particularly in resistant cases [10].

A previously healthy three-year-old boy presented to the Ophthalmology Outpatient Clinic with complaints of excessive tearing and photosensitivity of the right eye for 2 weeks. The right eye was apparently smaller than the left eye. There was no sign of injection in both conjunctiva and periorbital soft tissue indicative for inflammation or infection. Orbital MR imaging yielded a 36 × 32 × 38 mm mass fulfilling the right maxillary sinus and protruding into the infraorbital extrachanoal region (Figure 1). Complete blood count showed hemoglobin; 10 g/dL, thrombocyte; 77 × 10⁹/L and leukocyte 4.4 × 10⁹/L with no blasts on peripheral

blood smear. Serum biochemistry for liver, kidney functions and electrolytes were within normal limits. Both bone marrow aspiration and mass biopsy were performed at the same time under general anesthesia. Light microscopy revealed FAB L1 type of lymphoblasts. Flow cytometric analysis of the bone marrow aspirate demonstrated of the blast gate of 56% with a positivity of CD10, CD19 and TdT but negativity of myeloperoxidase. Mass biopsy also revealed lymphoblastic infiltration with CD10 and CD19 positive cells (Figure 2). ALL-IC



Figure 1A. A huge mass fulfilling the right maxillary sinus protruding into extrachanoal region.

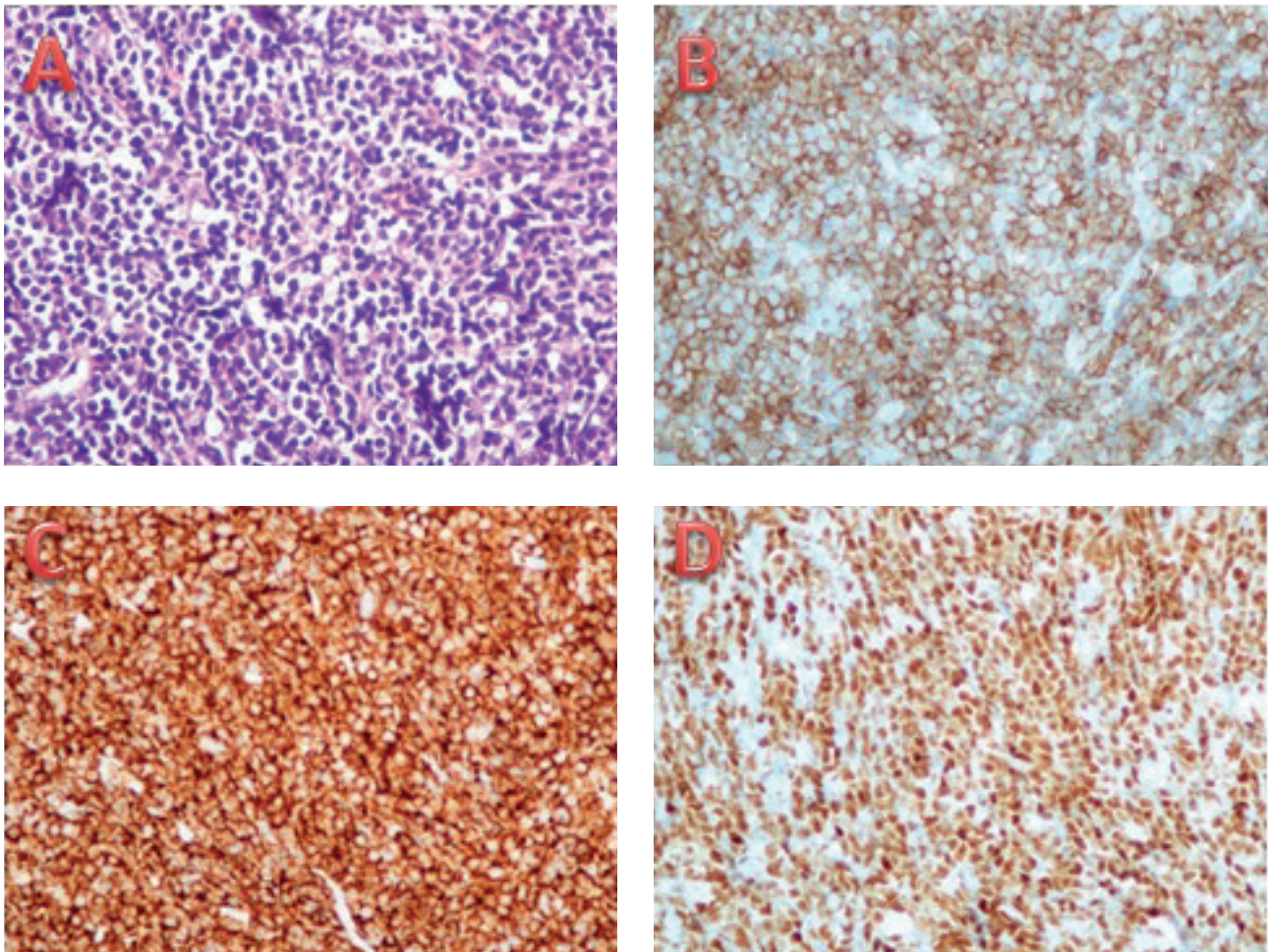


Figure 2. a) Hematoxylin-eosin staining, b) tdt positivity, c) CD10 positivity, d) CD19 positivity of the mass.

2009 protocol was started and facial asymmetry dramatically improved by the end of a week course of steroid treatment. Cerebrospinal fluid analysis showed no blasts. He was defined as high risk group due to minimal residual disease (MRD) assay on the 15th day. After completion of the high risk blocks of the protocol, no residual mass was detected in the right orbital cavity with imaging.

Maxillofacial mass at the presentation of ALL has rarely been reported [11]. The vague ophtalmological symptoms and facial asymmetry may urge physicians to consider a possible mass in the differential diagnosis and imaging might be very instructive. Unusual presentations of EM masses should be kept in mind in atypical cases.

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