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Pediatric acute lymphoblastic leukemia presenting with periorbital edema

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ABSTRACT

Objective: To describe a rare presentation of acute lymphoblastic leukemia (ALL) that is important to the field of pediatric otolaryngology.
Methods: The case of a nine-year old female who presented with left periorbital edema as her initial symptom of ALL was reviewed along with pertinent literature.
Results: At the time of patient's initial presentation, a computed tomography scan of the sinuses showed sphenoid sinus opacification and left lateral rectus muscle enlargement. She was started on antibiotics without improvement. Subsequently, magnetic resonance imaging of the brain demonstrated an infiltrative process involving the sphenoid sinus marrow space, lateral rectus muscles and dura. Her peripheral blood count revealed 40% blasts, and a bone marrow biopsy demonstrated B-cell ALL. She immediately underwent induction chemotherapy, and her post-induction bone marrow biopsy showed no evidence of residual disease.
Conclusion: This case highlights the importance of understanding and considering rare, aggressive diseases that can masquerade as simple periorbital edema.

1. Introduction

Acute lymphoblastic leukemia (ALL) represents the most common childhood cancer, comprising about 26% of cancers diagnosed in patients aged birth to 14 years [1]. Typical presenting symptoms can be vague and include fever, pallor and fatigue, which are also prominent features of more common, self-limited viral illnesses. Therefore, it is important to evaluate for other features that are more specific to ALL such as hepatomegaly, splenomegaly, lymphadenopathy and petechiae [2]. Extramedullary sites can also be involved with ALL including the testicles, central nervous system and orbit [3]. Orbital or ocular involvement as the presenting symptom for ALL is very rare with only five case reports identified in the literature [3–7]. This is an important entity to recognize as it can portend a poorer prognosis and more aggressive underlying disease [8,9].

In this article, we detail the clinical course of a nine-year old girl who developed unilateral periorbital edema as her initial symptom of ALL. Surprisingly, there are not any reports of ALL presenting with initial orbital findings in the otolaryngology literature. Pediatric otolaryngologists are routinely involved in the care of children with periorbital swelling, often when there is concern for periorbital or orbital cellulitis as a complication of sinusitis. Thus, it is important for pediatric otolaryngologists to be aware of and consider rare, aggressive diseases that can present with periorbital edema.

2. Clinical case

MC is a nine-year old girl with no past medical history who presented to her pediatrician with progressive left eyelid and periorbital edema over the preceding days. She denied other systemic symptoms. A computed tomography (CT) scan of the sinuses demonstrated opacification of the sphenoid sinuses and enlargement of the left lateral rectus muscle (Fig. 1). She was treated empirically with cefdinir for acute sinusitis with associated periorbital cellulitis.

Her symptoms worsened despite antibiotic therapy. She was admitted to the hospital for further evaluation, and at this point the pediatric otolaryngology team became involved in her care. On admission, the patient endorsed progressive fatigue, lower back pain and left frontal headaches for three weeks. More recently she developed double vision on left lateral gaze. She denied any nasal symptoms including congestion, post-nasal drip or rhinorrhea. On physical examination, she had mild proptosis of the left eye with associated periorbital edema as well as incomplete abduction of the left eye on lateral gaze. Her periorbital region was not erythematous, warm nor was there any purulent drainage to suggest underlying infection. Thus, her presentation did not appear consistent with an infectious etiology.

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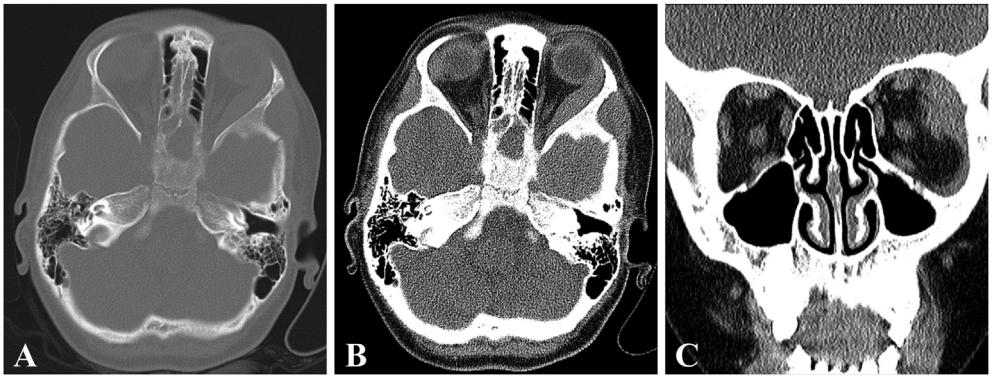

Fig. 1. Initial CT scan of the sinuses. Non-contrast CT scan of the sinuses shows isolated opacification of the bilateral sphenoid sinuses on bone window (A) as well as enlargement of the left lateral rectus muscle on axial and coronal soft tissue windows (B, C).

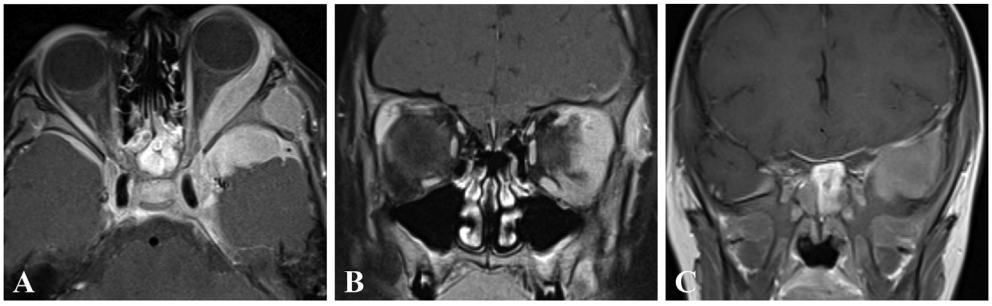

Fig. 2. Initial MRI scan of the orbits. T1 post-gadolinium, fat suppressed MRI of the orbits shows significant soft tissue enlargement and enhancement of the left lateral rectus muscle (A, B) as well as along the dura (A, C). There is also abnormal soft tissue and marrow replacement within the bilateral sphenoid sinuses (A, C).

Magnetic resonance imaging (MRI) of the brain demonstrated abnormal signal within the bilateral sphenoid sinuses, sphenoid wings, petrous apices and bony walls of the orbit, suggestive of a neoplastic bone marrow replacement process (Fig. 2). It also highlighted abnormal enhancing soft tissue extending along the lateral aspect of the orbits, dura and perineurium, consistent with either lymphoma or leukemia. Her labs showed a normal white blood cell count of 6.3, but she had 40% blasts in the peripheral smear as well as thrombocytopenia and anemia. At this time there was suspicion for acute myeloid or lymphoblastic leukemia.

A bone marrow biopsy demonstrated B-cell lymphoblastic leukemia with a hypercellular bone marrow and decreased hematopoiesis. Lumbar puncture did not show any blasts in her spinal fluid. Based on the dural and perineural involvement on the MRI, she was determined to have central nervous system disease. She was started on induction chemotherapy per the AALL1131 regimen consisting of dexamethasone, vincristine, PEGAsparaginase, daunorubicin and intrathecal therapy with cytarabine and methotrexate. Her induction chemotherapy finished during this admission, and a post-induction bone marrow biopsy showed no evidence of minimal residual disease. A repeat MRI of the brain showed significant improvement in the soft tissue and marrow abnormalities noted on her admission MRI (Fig. 3).

She proceeded with consolidation chemotherapy, and a MRI of the brain following this treatment showed continued improvement. Her post-treatment bone marrow biopsy revealed a hypocellular bone marrow with trilineage hematopoiesis and no morphologic or immunophenotypic evidence of B-cell ALL. Subsequently, she started an interim maintenance chemotherapy regimen consisting of high-dose methotrexate. Currently, she is twenty months out from her initial diagnosis and remains without evidence of clinical disease. Her course of therapy will be approximately three years and will include cranial radiation.

3. Discussion

3.1. Review of current literature

In this article, we outline a rare cause of unilateral periorbital edema and proptosis in a pediatric patient: ALL. Our literature review revealed five other case reports from the ophthalmology and oncology literature detailing ALL presenting with orbital or ocular symptoms (Table 1) [3–7]. Chaudry et al. and Hossain et al. described patients

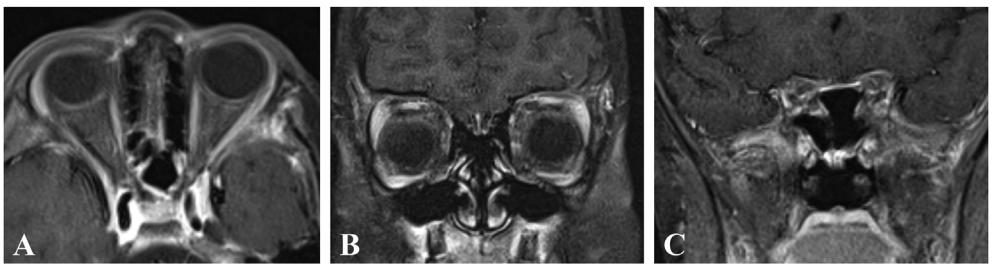

Fig. 3. Post-induction chemotherapy MRI scan of the orbits. T1 post-gadolinium, fat suppressed MRI of the orbits shows marked improvement of the previously seen soft tissue enlargement/enhancement of the left lateral rectus muscle (A, B), sphenoid sinuses/marrow space (A, C) and along the dura (A, B, C).