When it's not a complicated frontal sinusitis - A Langerhans cell histiocytosis case-report

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ABSTRACT
Langerhans cell histiocytosis (LCH) is a rare and heterogeneous disease that appears mainly in children and frequently affects bone structures such as the skull, with possible involvement of the orbit. In this paper, we present the case of an 11-year-old boy who came to the emergency department with left periorbital edema and erythema with about 10 days of progressive worsening. Computed tomography was compatible with complicated left frontal sinusitis with bone erosion into the orbit and intracranial compartment, and magnetic resonance imaging corroborated these findings. Due to the lack of improvement with medical therapy, surgical exploration was performed through an external approach, identifying a lesion in which the anatomopathological study confirmed the diagnosis of LCH.
Keywords: Langerhans cell Histiocytosis; Pre-septal cellulitis; Frontal bone; Orbit; Pediatrics

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INTRODUCTION

Langerhans cell histiocytosis (LCH; formerly known as eosinophilic granuloma or histiocytosis X), is a rare, heterogeneous disease characterized by the clonal proliferation of pathological Langerhans cells (histiocytes)[1]. Although it can manifest at any age, its incidence (3–5/million) is highest among children[2].

Langerhans cell histiocytosis can affect any organ or physiological system, but the most common site is the axial skeleton (the skull, jaw, spine, pelvis, ribs, and long bones). The most commonly affected bone in children is the skull, especially the frontal bone, and the orbits can be involved[3].

Orbital LCH is rare, accounting for < 1% of orbital tumors, and can be the only manifestation of LCH. The signs and symptoms include proptosis, diffuse periorbital edema with or without erythema, eyelid edema and pain, and differential diagnoses include preseptal cellulitis[4], which is a complication of acute sinusitis.

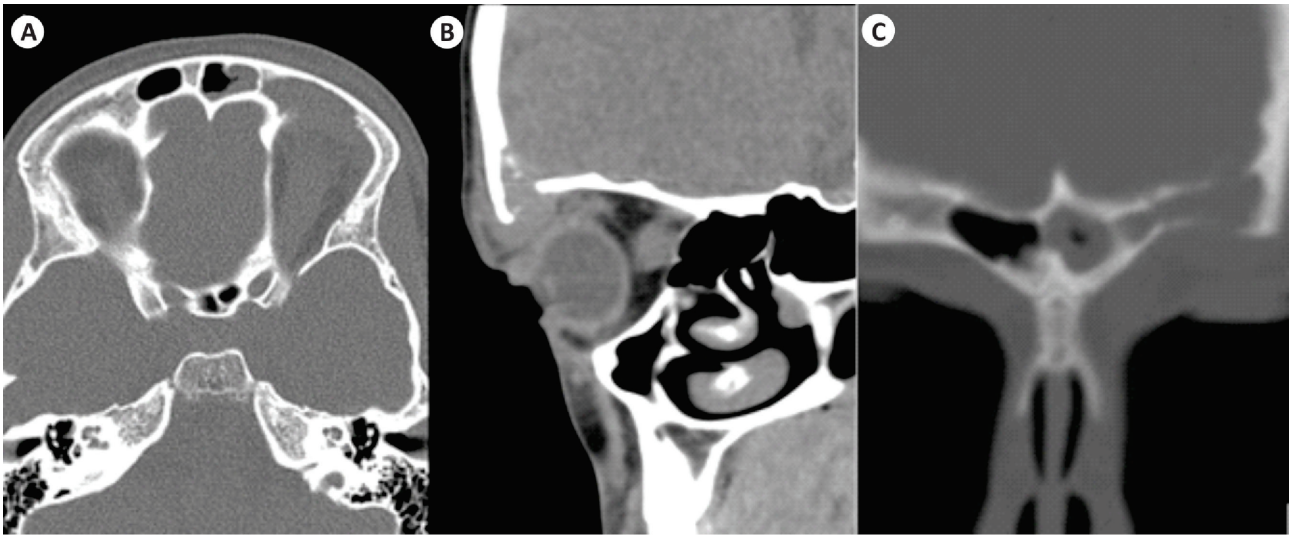
Here we describe the case of a child who initially presented with apparent frontal sinusitis complicated by preseptal cellulitis and was finally diagnosed with LCH.

CASE DESCRIPTION

An 11-year-old boy with a history of allergic rhinitis treated with topical nasal corticosteroids, and no other relevant personal or family history, presented at the emergency department with edema in the upper left eyelid, which had been present for 10 days. An ophthalmologist evaluated him on the day after symptom onset and prescribed topical ofloxacin. However, inflammatory signs progressively worsened, with extension of the edema to the lower eyelid, mild erythema, and pain on palpation in the left frontal region, but without limitation or pain during eye movement or changes in visual acuity. He experienced left facial pressure but denied recent worsening of allergic rhinitis as well as significant nasal obstruction or rhinorrhea. Other than the periorbital changes, the general condition of the patient was good while in the emergency department. He had no fever or meningeal signs, and nasal endoscopy revealed mild hyperemia and hypertrophy of the inferior turbinates, without lesions or mucopurulent rhinorrhea.

Computed tomography (CT) imaging showed findings compatible with sinusitis, with the epicenter in the left

FIGURE 1
Computed tomography on admission. A – axial plane; B – sagittal plane; C – coronal plane.

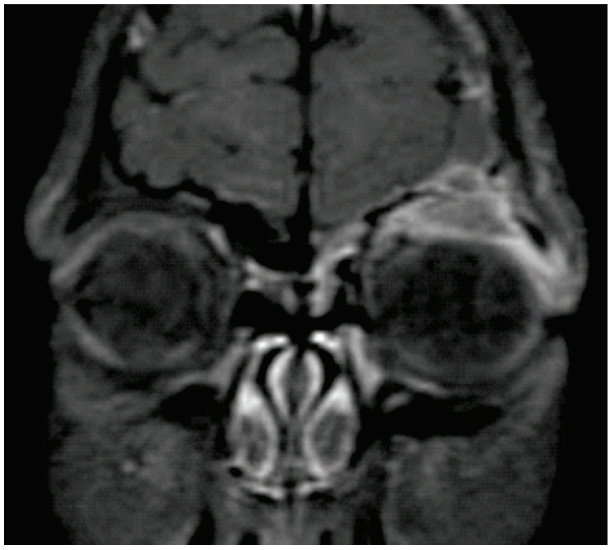


frontal sinus and extension of the infectious process to the preseptal and intracranial orbital cavity. Solution continuity was found in the bone at the level of the floor and posterior wall in the left frontal sinus (Figure 1). Magnetic resonance imaging (MRI) of the head on the same day revealed an inflammatory/infectious process centered in the left frontal sinus, with a central area of low uptake with peripheral enhancement corresponding to a possible abscess, preseptal extension to the orbit, and intracranially with slight adjacent meningeal enhancement (Figure 2).

The patient was admitted for endovenous treatment with amoxicillin-clavulanic acid (50 mg/kg/dose 8/8h) and methylprednisolone (1 mg/kg/day) and to monitor disease progression. Although his clinical status did not worsen during hospitalization, the initial improvement of the inflammatory signs at the start of treatment was not significant and MRI one week later revealed a slightly worsened state. Laboratory findings did not show anemia, leukocytosis, or neutrophilia, and C-reactive protein was slightly elevated (0.30 mg/dL).

The possibility of a neoformation process ,rather than a complication of frontal sinusitis, was considered. Hence the left frontal sinus with an extemporaneous anatomopathological examination was surgically explored through a supraciliary incision (Figure 3). The findings revealed no purulent content, but a preliminary anatomopathological examination revealed a friable and hemorrhagic lesion that was compatible with a histiocytic tumor, namely LCH. Considering the location and preoperatively extended bone erosion likely requiring complementary systemic treatment, we excised the lesion by partial curettage of the bone walls. No postoperative complications developed. The increased supraorbital edema during the immediate postoperative period was expected and gradually subsided within a few days. The diagnosis of LHC was

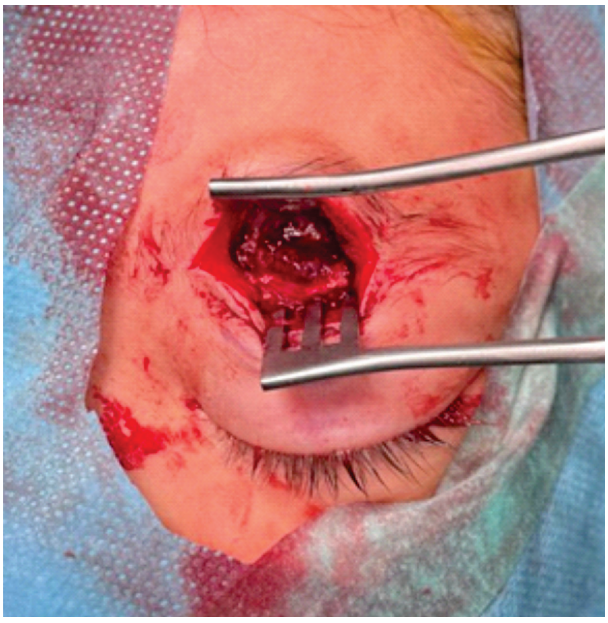
FIGURE 2
Magnetic resonance imaging on admission. T1-weighted coronal plane with gadolinium enhancement.



confirmed by the final anatomopathological findings of diffuse medium-to-large cell (CD68+, CD1a+, S100+, CD4+, CD45+/-, and CKAE1/AE3) proliferation associated with occasional multinucleated giant cells and some neutrophils and eosinophils.

Abdominal ultrasound, bone and chest X-rays, bone scintigraphy, and CT of the spine revealed no additional lesions; therefore, the osteolytic lesion of the frontal bone was framed within the diagnosis of LCH of a single system, with a unifocal lesion “of risk” to the central nervous system (CNS). The patient was referred to a specialized pediatric center to undergo systemic treatment with vinblastine and prednisolone. Imaging assessment six weeks after treatment revealed disease regression, but not complete resolution. Treatment was thus continued for a further six weeks.

FIGURE 3
Surgical approach. Supraciliary incision with visualization of the lesion



DISCUSSION

Multisystem LCH has a diverse presentation, as clinical manifestations vary according to the affected bone and surrounding structures. The initial symptoms of periorbital lesions like that presented herein, can easily be confused with those of an inflammatory/infectious eye disease[5] and the progression to periorbital edema and erythema can mimic cellulitis which does not improve with antibiotic therapy[3].

The current classification of LCH distinguishes between multisystem and single-system diseases and bone involvement alone is further divided into unifocal/isolated and multifocal types. The objective of this classification is to guide the diagnosis and treatment of patients, because although this disease is histologically benign, the clinical course can vary from self-limiting, to rapidly progressive (which is potentially fatal), and 30–40% of patients develop permanent sequelae[6].

The therapeutic options depend on the extension and severity of the disease. However, some LCH presents as individual lesions located in regions that present a risk for the CNS, with orbital involvement being an example. The best treatment for such lesions has not been standardized. Most ophthalmologists and surgeons advocate a more conservative approach that includes subtotal curettage with intralesional corticoid therapy, and pediatric oncologists recommend systemic therapy to prevent permanent debilitating sequelae[4].

The patient described herein might have been categorized into the latter group. However, systemic treatment was the immediate choice due to the significant bone destruction in the orbital and intracranial compartments and the MRI findings that strengthened the suspicion of intracranial extension. The treatment protocol followed the current guidelines of vinblastine and prednisolone for six weeks, followed by imaging assessment to evaluate the response. In the absence of complete resolution, the treatment can be continued for a further six weeks and maintenance therapy can be extended for up to 12 months. Although LCH is considered predominantly benign, it can be associated with several sequelae that can manifest at any time after diagnosis, up to several years later. Therefore, children with LCH should be followed-up regularly for at least 5 years after the end of treatment or until the end of growth and puberty6.

In conclusion, although orbital LCH is rare, it should be included in the differential diagnoses of preseptal cellulitis. Otorhinolaryngologists, who frequently encounter complications of sinusitis, should be aware of the different manifestations of LCH, aim for a timely diagnosis and the provision of adequate guidance.

Conflict of Interest

The authors declare no conflict of interest regarding this article.

Privacy policy, informed consent and approval by the ethics committee

The authors declare having obtained written informed consent for the use of patients' photographs in this article.

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