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CASE REPORT

Unusual Location for Langerhans Cell Histiocytosis: Basisphenoid extended to Parapharyngeal Space

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disease with unknown etiology involving abnormal proliferation of histiocytes. We hereby describe an LCH that has a rare location. A 4-year-old female patient was referred to our clinic with headache lasting for 2 months. Magnetic resonance imaging (MRI) showed an expansile mass on the level of basisphenoid extended to the right parapharyngeal space with dense contrast enhancement. The patient underwent endoscopic endonasal transsphenoidal surgery for biopsy of the mass. Immunohistochemical and pathological studies confirmed LCH diagnosis. This is the first case report of LCH extended to the parapharyngeal space in the current literature.

Keywords: Basisphenoid, Langerhans cell histiocytosis, Parapharyngeal space.

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disease with unknown etiology involving abnormal proliferation of histiocytes. It occurs most often between 1 and 3 years of age and may appear as a single lesion or can affect many body systems, such as skin, bone, lymph glands, liver, lungs, spleen, brain, pituitary gland, and bone marrow.[1] Bone involvement mostly occurs in LCH and often includes the skull. When calvarial lesions extend into the nervous system, a variety of neurologic manifestations may be seen. Also, the skull base involvement may cause neurologic disorders.

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CASE REPORT

A 4-year-old female patient referred to a pediatric clinic with headache lasting for 2 months. Radiological imagings showed a mass located at the skull base and the patient was sent to our clinic for futher investigations. Physical examination revealed a swelling of approximately 1 cm diameter on the frontal convexity. No other system or site was involved. Magnetic resonance imaging (MRI) showed an expansile mass on the level of basisphenoid extended to the right parapharyngeal space with dense contrast enhancement (Figs 1 and 2). Another mass was seen on the right frontal calvarium convexity

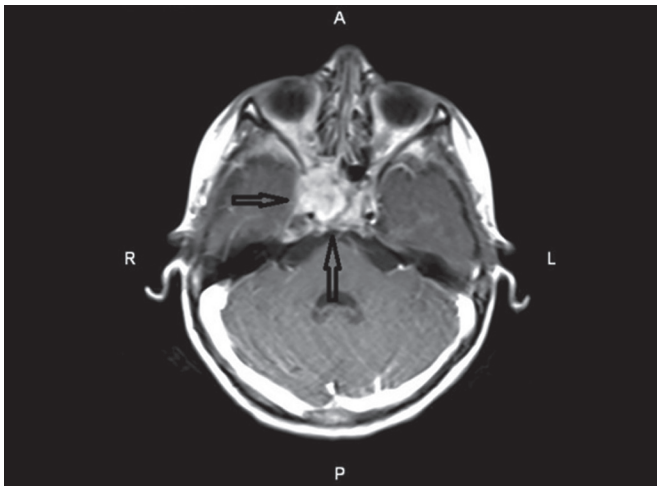

Fig. 1: Axial T1 postcontrast MRI showing lesion mainly involving the right side of the basisphenoid

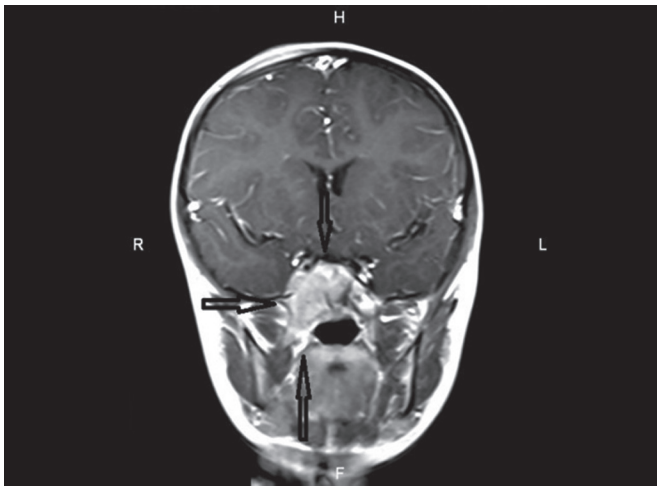

Fig. 2: Coronal T1 postcontrast MRI showing lesion extended to the parapharyngeal space

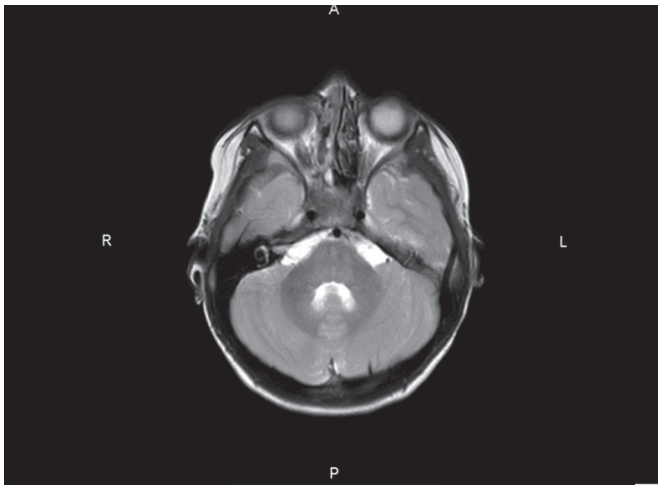

Fig. 3: Axial T2 MRI (after 24-week treatment) showing marked regression of the lesion

with dense contrast enhancement and subperiosteal and epidural soft tissue thickening. The patient underwent endoscopic endonasal transsphenoidal surgery for biopsy of the mass. Immunohistochemical and pathological studies confirmed LCH diagnosis. The patient was consultated to the pediatric oncology department for further treatment. The treatment started with vinblastine and prednisolone chemotherapy. After 24 weeks of chemotherapy treatment, an MRI showed marked regression of the lesion (Fig. 3). The patient is now following with remission for 4 months.

DISCUSSION

Masses of the sphenoid bone and sphenoid sinus in the pediatric population are uncommon; however, a wide range of pathologies can involve this central skull base structure, including benign and malignant lesions. Due to its central location, abnormalities of the sphenoid bone can affect many critical skull base structures.[2] Levine described 13 sensitive structures that are at risk in the setting of sphenoid disease: The dura, the cavernous sinus, the internal carotid artery, the oculomotor nerve, the pituitary gland, the optic nerve and chiasm, the trochlear nerve, the ophthalmic nerve, the abducens nerve, the maxillary division of the trigeminal nerve, the sphenopalatine ganglion, the sphenopalatine artery, and the pterygoid canal and nerve.[3] In our case, interestingly, there was no other neurologic symptom except for headache although the expansile mass extended from the basisphenoid to the parapharyngeal space. In the literature, there are several case reports about sphenoid bone LCH, and nearly all of them have different neurological symptoms. Stromberg et al[4] presented a 16-year-old boy with a 4-week history of impaired vision in his left eye associated with light perception. An MRI demonstrated a mass filling the sphenoid sinus on the left extending into the orbital apex. Binning and Brockmeyer[5] presented a 12-year-old boy with a headache, swelling around his left eye, and blood discharge from his nose. Radiological imagings were performed, which showed a mass within the greater wing of the left sphenoid bone and the lateral wall of the left sphenoid sinus. The mass abutted the left lateral rectus muscle, left temporal lobe, and cavernous sinus and extended into the left pterygopalatine fossa. Krishna et al[6] presented a 15-year-old boy with raised intracranial pressure, decreased visual acuity, bilateral abducent nerve palsy, and 25% hypoesthesia in all three divisions of the right trigeminal nerve. The cranial MRI of the patient revealed a lesion of the clivus and the sphenoid sinus, extending to the right cavernous sinus to encase the right cavernous internal carotid artery segment, and also involving the right petrous apex and the extradural space in the prepontine region. Yu et al[7] presented a 22-year-old boy with isolated sphenoid sinus LCH, and the only symptom in the patient was headache. Also, there are reports with orbital pain[8] and optic neuorpathy.[9]

Tissue sampling is an important step in diagnosis as in our case and all similar cases, and it should be performed endoscopically. With the endoscopic approach, adequate tissue can be sampled for pathological analysis and anatomical structures better defined compared to an open approach.

In the differential diagnosis of sphenoid bone masses in the pediatric population, we should keep in mind leukemia/lymphoma infiltration, rhabdomyosarcoma, meningioma, fibrous dysplasia, aneurysmal bone cyst, giant cell tumor, chordoma, and craniopharyngioma. And LCH should not be forgotten within these rare pathologies.

Our case is the first case report of LCH extended to the parapharyngeal space in the current literature. We should consider LCH in differential diagnosis of the tumors invading the sphenoid bone–skull base and even in the parapharyngeal space.

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