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.

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. 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.

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 further 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 involving the right side of the basisphenoid

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

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A mass filling the sphenoid sinus on the left extending into the orbital apex. Binning and Brockmeyer 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 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 internal carotid artery segment, and also involving the right petrous apex and the extradural space in the preoptic region. Yu et al 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 and optic neuropathy.

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.

REFERENCES


