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Case report

Langerhans cell histiocytosis in the jugular foramen

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ABSTRACT

Langerhans cell histiocytosis (LCH) is a rare disease of neoplastic proliferation of monocyte–macrophage system. Although LCH can affect almost any organ, solitary involvement of jugular foramen is extremely rare and can present a diagnostic dilemma because of its rarity at this location. Here, we present the case of an adult patient with LCH affecting the jugular foramen, and review the relevant literature.

1. Introduction

Langerhans cell histiocytosis (LCH), previously referred to as histiocytosis X, is a rare disorder characterized by clonal proliferation and excess accumulation of pathologic Langerhans cells causing local or systemic effects [1,2]. The exact etiology of LCH is still unknown. Clinical syndromes within this entity include eosinophilic granuloma, Hand–Schüller–Christian disease, and Abt–Letterer–Siwe disease [2]. Langerhans cell histiocytosis typically occurs in childhood and adolescence as solitary osteolytic lesions. The most frequent sites of the bony lesions are the skull, femur, mandible, pelvis and spine [3]. A variety of treatment modalities have been reported [4,5]. Here, we present an adult female patient with LCH of the jugular foramen.

2. Case report

2.1. History

A 23-year-old female patient presented with a 6-week history of occipital pain. There was no history of trauma or neoplasm. More recently, the patient complained of progressive stiffness and weakness of neck, which impaired her range of neck motion and caused torticollis. She must use cervical gear to complete the daily activities. In the month prior to her admission, her occipital pain increased with hoarseness of voice and difficulty swallowing.

2.2. Examination

Neurologic examination was remarkable for marked impairment of cervical flexion, extension and rotation. The left palate was mildly weak with diminution of the gag reflex. Exceptionally, physical examination revealed a 3-cm, firm, and regular lesion with normal overlaying skin in the left mastoid process.

2.3. Investigation

Magnetic resonance imaging (MRI) demonstrated a homogeneous, 5.5 cm × 3.5 cm solid mass involving the left jugular foramen and lateral mass of atlas. The mass showed low signal intensity on both T1- and T2-weighted images, and intense heterogeneous enhancement following intrave-

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nous gadolinium administration. Magnetic resonance venography revealed occlusion of transverse and sigmoid sinus. Computed tomography showed an irregular osteolytic lesion of jugular foramen extending downwards lateral mass of atlas (Fig. 1). The remainder of the examination was unremarkable.

2.4. *Surgery*

A left far lateral approach was utilized to excise the lesion. A surgical corridor was created by separating suboccipital muscles and paravertebral muscle, drilling the left partial occipital condyle. Exposure of the tumor demonstrated a gray, yellow mass in the jugular foramen and lateral mass of atlas. The lesion was easily separated, blood supply was moderate, and finally tumor was partially resected. In order to restore stability of the cervical spine, occipitocervical fusion was performed.

2.5. *Histology*

Gross examination of the surgical specimen revealed multiple, irregular fragments of pale and tan soft tissue measuring in aggregate 3 cm × 2.1 cm × 1.7 cm. Histological sections revealed a granulomatous reaction pattern, with extensive aggregates of histiocytes proliferation, which showed broad cytoplasm cells and a kidney-shape nucleus, along with clusters of eosinophils. Immunohistochemical stain by CD1a antibody and S-100 immunoperoxidase stain were positive only in the histiocytic cells. Because of the immunoexpression of S-100 and CD1a by lesional cells, the diagnosis of LCH was made (Fig. 2).

2.6. *Postoperative course*

The patient tolerated surgery well, without neurological deficit and with good recovery. In the first month, MRI demonstrated

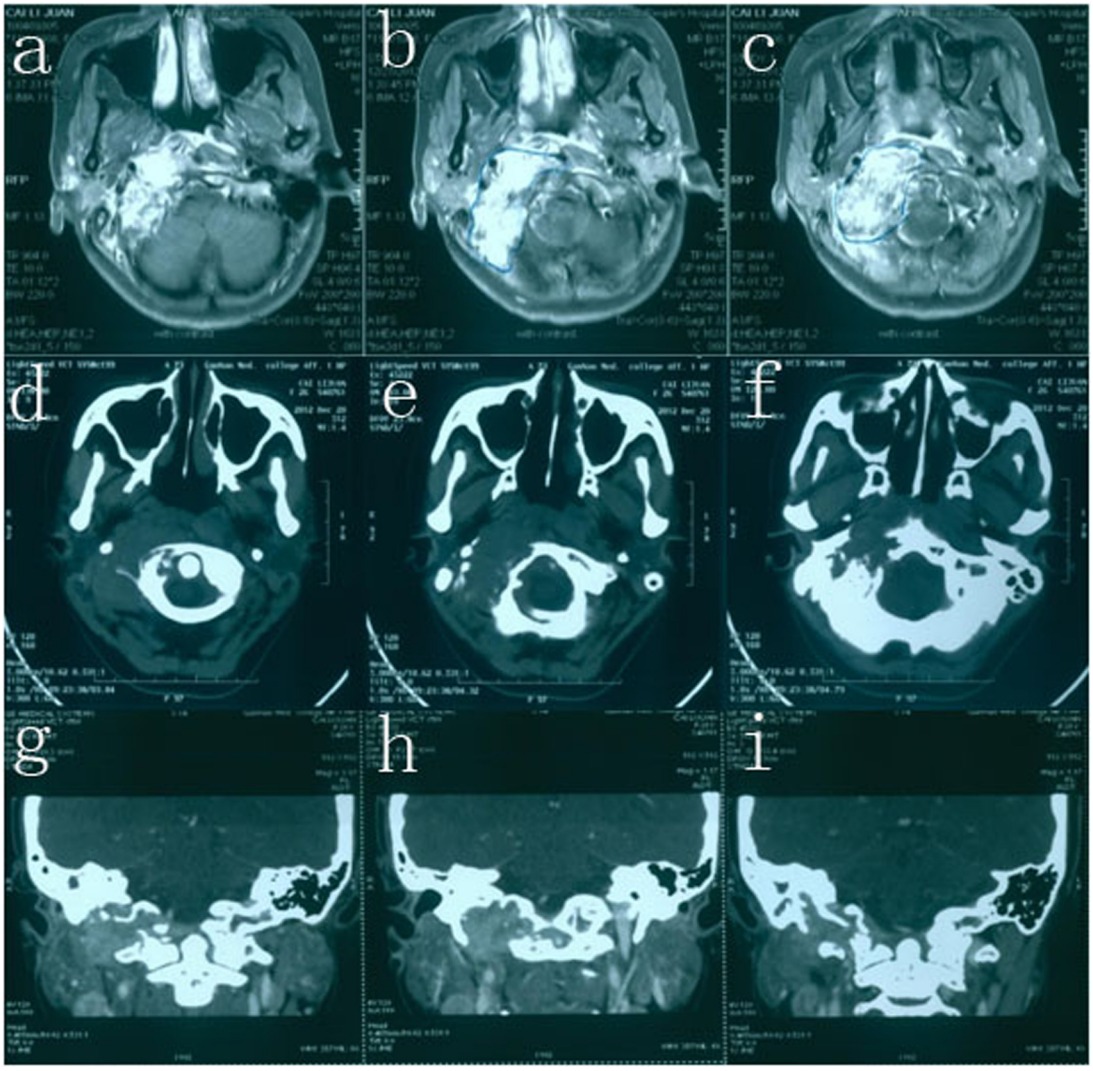

Fig. 1 – Post-gadolinium axial T1-weighted MR images (a–c) show an intensive heterogeneously enhancing mass affecting the left jugular foramen with extension to the lateral mass of atlas. CT images (d–i) show bone destruction of the left jugular foramen and lateral mass of atlas.