Eosinophilic granuloma of the clivus

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The working group of the Histiocyte Society has divided histocytic disorders into 3 groups: (1) dendritic cell histiocytosis, (2) macrophage-related disorders, and (3) malignant histiocytosis.

Langerhans cell histiocytosis LCH belongs in group 1 and encompasses a number of diseases. On one end, the clinical spectrum includes an acute, fulminant, disseminated disease called Letterer-Siwe disease, and, on the other end, solitary or few, indolent and chronic lesions of bone or other organs called eosinophilic granulomas.

Eosinophilic granuloma of the skull is the most common presentation of the disease, and the associated symptoms depend on the location of the lesion.

In the skull base, petrous ridge of the temporal bone is the most common site and solitary involvement of the clivus is extremely rare [1) 2) 3)].

Lederman first reported the case of a eosinophilic granuloma, or unifocal Langerhans' cell histiocytosis, in the clivus of a child [4)].

Clinical features

Clinical presentation of clival LCH depends on the size and extent of the lesion. The commonest presentation is with abducens nerve palsy with or without retro-orbital, post-auricular or neck pain, decreased vision, and raised intracranial pressure.

Diagnosis

They present a diagnostic dilemma because of its rarity at this location.

It is usually hyperdense on plain computed tomography (CT).

MRI

In MRI it is usually a heterogeneous destructive mass with areas of hemorrhages, cystic changes, and calcifications. It shows little or no enhancement on post-contrast study. [5)].

It includes abnormal marrow signal with homogeneously or heterogeneously enhancing destructive soft tissue mass and variable local extension. It is hyperintense on T2 and hypointense on T1-weighted images [6)].

Diffusion restriction is probably due to the increased cellularity of the lesion and may also correlate with the aggressive nature of the LCH [7)].

Differential diagnosis

LCH is a rare but possible cause of a clival mass in the pediatric population. Since the imaging features are non-specific, it is often difficult to differentiate this from other similar masses. Biopsy is often required to confirm the diagnosis [8)].

MRI features of rhabdomyosarcoma are not specific [9)] and may be difficult to differentiate from LCH in the absence of characteristic lytic skeletal lesions at other sites.

Imaging differentials of an aggressive lesion involving the clivus or central skull base in a pediatric patient would include chordoma, lymphoma, metastatic neuroblastoma, and rhabdomyosarcoma.

Calcifications and hemorrhage are uncommon features in LCH and synchronous characteristic lytic lesions in other areas of the skeleton would favor LCH. Lymphoma of central skull base is another uncommon tumor in pediatric patients, but the most common malignancy in the head and neck region. It presents characteristically as T1-/T2-weighted isointense or hypointense mass with homogeneous enhancement and lack of calcifications and hemorrhage [10)].

The presence of enlarged lymph nodes in the neck would favor lymphoma. Neuroblastoma can present for the first time with features related to extra-abdominal metastases and cross-sectional imaging of the abdomen may reveal the primary lesion. MRI features of metastatic neuroblastoma are nonspecific, and it presents as a homogenously or heterogeneously enhancing mass which is hypointense on T1 and hyperintense on T2-weighted MRI, with or without calcifications and cystic changes. [11)].

Lymphoma and neuroblastoma can be associated with multiple metastatic bone lesions however lesions are mottled or permeative unlike the characteristic punched-out or bevelled lesions of LCH. Rhabdomyosarcoma arising in the nasopharynx and invading the sphenoid and clivus can present as a mass in the central skull base [12)].

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Case reports

2012

A 6-year-old girl presented with a 1½ month history of hoarseness of voice and deviation of the angle of mouth to the left side, and slurring of speech, difficulty in swallowing, and decreased hearing for 20 days. Examination revealed right 6 th, 7 th, 8 th, 9 th, 10 th, and 12 th cranial nerve palsy. Magnetic resonance imaging (MRI) revealed a large homogeneously enhancing mass involving the clivus with extension into the prepontine cistern, petrous apex, sphenoid sinus, and the right nasopharynx

Biopsy of the friable mass seen in the nasopharynx was done under endoscopic guidance. Histopathology showed a tumor composed of large cells with grooved nuclei and eosinophilic cytoplasm, which were CD1a positive.

Admixed with them were lymphocytes, plasma cells, and eosinophils. A diagnosis of LCH was made. Bone scans did not reveal any other lesion in the rest of the body. She was given chemotherapy for 12 weeks and showed a good response with reduction in the size of the mass and clinical improvement.

T1-weighted sagittal MR image (a) shows a hypointense mass involving the clivus with extension into the nasopharynx and sphenoid sinus. Post-gadolinium T1W sagittal image shows intense enhancement in the mass. T2-weighted axial image shows hyperintense mass extending into the right petrous and prepontine cistern. DWI (d) and ADC map image (e) show restricted diffusion in the mass (arrow). (f) Photomicrograph illustrating the histiocytes with grooved nuclei (black arrows) (H and E, ×100). (g) Immunohistochemistry for CD1a (white arrows) (avidin peroxidase, ×400). (h) Axial post-gadolinium T1W image shows reduction in the size of the lesion.

Multiple cranial nerve palsy is a rare presentation as seen in the present case [14)].

2004

A 15-year-old boy presented with raised intracranial pressure, decreased visual acuity, bilateral abducent nerve palsy, and 25% hypoesthesia in all three divisions of the right trigeminal nerve. He had normal strength but with a left-sided upper and lower limb hypertonia and hyper-reflexia and an upgoing plantar reflex. The magnetic resonance (MR) imaging revealed a heterogeneously iso-to hyperintense 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. The lesion was brilliantly enhancing on contrast. An extended frontobasal approach was used to excise the lesion. Following surgery, the residual parasellar and the right petrous apex tumor was treated with a low dose radiation therapy. At follow-up after 1.5 years, there was significant clinical improvement and the computed tomographic scan showed no residual lesion.

A review of the literature reveals that this is only the second reported case of a spheno-clival LCH. An additional feature includes extensions into the parasellar as well as the petrous apex regions. Despite the extensive spread, the surgical excision with low dose radiation therapy was successful in providing complete resolution of the tumor [15)].

A 5-year-old girl with Langerhans cell histiocytosis (LCH) of the clivus.

In 2004 only five patients, have been described with LCH at this site.

The patient of Hurley et al. differs from those previously reported by her atypical clinical presentation with torticollis, but without a abducens nerve palsy. In addition, she is the first patient to present with concomitant disease elsewhere at the time of diagnosis, i.e. both femoral necks and left proximal humerus.

This patient presents unique features and underlines the importance of including LCH in the differential diagnosis of erosive lesions of the clivus [16)].

1998

Lederman first reported the case of a eosinophilic granuloma, or unifocal Langerhans' cell histiocytosis, in the clivus of a child [17)].

1997

a 4.5-year-old boy who presented with a complete abducens palsy on the right with an associated head turn. A computed tomographic scan of his head revealed a lytic lesion on that side, and magnetic resonance imaging showed the mass to be of low intensity on T1-weighted images and of high intensity on T2-weighted images with heterogeneous enhancement. INTERVENTION: A transnasal stereotactic biopsy was performed, revealing an EG. The patient was treated with stereotactic radiotherapy, and he became symptom-free with radiographic resolution of his lesion. Reviewing the literature, we found 13 series with 87 cases of EG in the petrous portion of the temporal bone. EG in the cranial base occurring outside of the temporal bone or in the temporal bone and extending intracranially is, however, quite rare, with only nine other cases reported, two of them clival. CONCLUSION: These findings suggest a classification schema in which cranial base EG lesions be grouped with either the more common extracranial petrous temporal bone lesions or the very rare intracranial lesions. Although there are few cases in the literature, treatment results indicate that clival EG, and perhaps all intracranial cranial base EGs, be treated by a biopsy alone, followed by surgery or stereotactic radiotherapy if there is an incomplete resolution of the symptoms or if there is a recurrence [18)].

1992

A 41-year-old white man with facial pain and diplopia was found to have an invasive lesion of the clivus. The final pathological diagnosis was eosinophilic granuloma. The patient's symptoms resolved completely after transsphenoidal resection of the lesion [19)].

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