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Eosinophilic granuloma at the cerebellopontine angle in an adult; a rare case report and literature review

Arash Safarian^a, Nima Derakhshan^{a,*}, Mousa Taghipour^a, Amirreza Dehghanian^b^a Neurosurgery Department, Shiraz University of Medical Sciences, Shiraz, Iran^b Pathology Department, Shiraz University of Medical Sciences, Shiraz, Iran

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ABSTRACT

INTRODUCTION: Langerhans cell histiocytosis (LCH) is a rare immunologic disorder, identified by immature proliferation of histiocytes which may present as systemic or focal lesions. Eosinophilic granuloma (EG) is localized form of LCH mainly involving bones such as skull, femur, spine, ribs, mandible and pelvis. Cerebello-pontine (CP) angle is a rare anatomic location for involvement by EG.

PRESENTATION OF CASE: A 32 year old man was being evaluated in our neuro-oncology clinic due to diplopia since 4 months ago. On physical examination he had left sided abducens paresis, hyposthesia over left half of his face and a decreased corneal reflex on left side. A magnetic resonance imaging (MRI) study revealed a lesion at left CP angle measuring 30 × 25 × 25 mm in size which was isointense in T1, hypointense on T2 with homogenous enhancement in post-contrast study. A standard retrosigmoid approach was carried out for resection of this lesion. Pathology report of the frozen section depicted infiltration of eosinophils and large mono-nuclear cells. The infiltrative nature of the lesion encouraged us not to attempt further resection. Permanent pathology report was in favor of EG. Patient was referred to an oncology clinic for proceeding with the steroid therapy.

DISCUSSION: To the best of authors' knowledge, this is the first report of EG at CP angle in an adult, in the literature. Infiltration of eosinophils and positivity for CD1a and S-100 renders the diagnosis unmistakable.

CONCLUSION: When the diagnosis is suggestive of EG, incompletely excised lesions can be further managed by steroid therapy.

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1. Introduction

Langerhans cell histiocytosis (LCH) is defined as neoplastic proliferation of eosinophils and histiocytes. Three major subtypes of LCH include eosinophilic granuloma (EG), Hand-Schuller-Christian disease and Letterer-Siwe disease. EG is described as the focal form of LCH mostly affecting the skull, femur, pelvis, ribs, spine and mandible. In the skull base, it has been reported to affect temporal bone, petrous apex and clivus, often in children and young adults. However, intracranial presentation is extremely rare, especially in adults. Here in, we present an adult with left abducens palsy due to EG at left CP angle mimicking a cp angle meningioma.

2. Case report

A 32-year old man was referred to our neuro-oncology clinic with chief complaint of diplopia and hyposthesia over the left half of his face since 4 months prior to this visit. His physical examination revealed left abducens paresis and hyposthesia over left half of his face particularly over V1 and V2 territory plus a diminished corneal reflex on left side. History and physical examination were normal for facial and vestibule-cochlear nerves. MRI revealed a left CP angle lesion measuring about 30 × 25 × 25 mm in size with compression over anterolateral aspect of the pontomesencephalic junction of brain stem. The lesion was isointense on T1-, hypointense on T2-weighted images with homogenous enhancement in post-contrast images (Fig. 1).

A retrosigmoid approach was attempted for resection of the lesion and after exposure of the tumor sample was obtained for frozen section. Pathology report of the frozen section was in favor of infiltration with large mononuclear cells and abundant eosinophils. Due to infiltrative nature of the lesion attempt for further resection was avoided. Permanent pathology was confirmed with immunohistochemical staining, which demonstrated immune-reactivity

* Corresponding author at: Shiraz University of Medical Sciences, Neurosurgery Office, Namazi Hospital, Namazi Square, Shiraz, Postal Code: 71973-11351, Iran.
E-mail address: nima.med83@yahoo.com (N. Derakhshan).

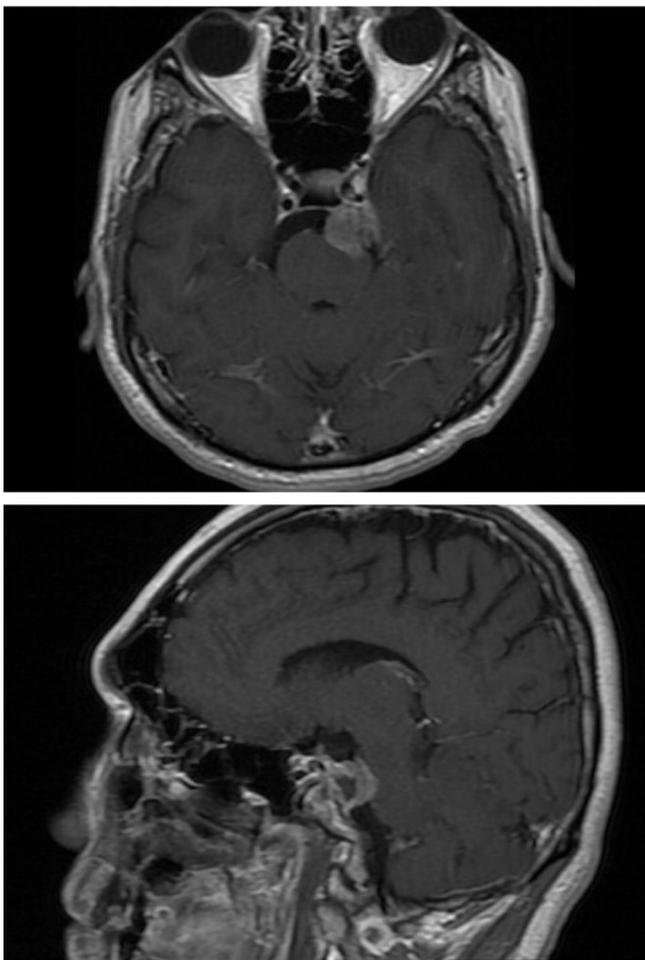


Fig. 1. Contrast-enhanced T1-weighted axial and sagittal magnetic resonance image of the lesion in CP angle with homogenous enhancement, with compression on anterolateral aspect of pontomesencephalic junction.

for S100, vimentin and CD1a which established the final diagnosis for EG (Fig. 2).

3. Discussion

Langerhans cell histiocytosis (LCH) is a group of disorders described as neoplastic proliferation of eosinophils, multinucleated giant cells, mononuclear cells, and Langerhans cells. This clinical entity is comprised as three clinical syndromes: Eosinophilic granuloma (EG), Hand-Schuller-Christian disease and Letterer-Siwe disease. EG is referred to the focal form of LCH with primary involvement of bones, particularly skull, pelvis, mandible, spine and ribs [1].

A few reports are available on presentation of EG at skull base (lesser wing of sphenoid [2], infra-temporal fossa [3], Sphenopetro-clival [4], clivus [5,6]) and orbit [7,8] and [9] in the literature which mostly affects the pediatric population.

Sampson [10] reported an adult male with EG of clivus which underwent transsphenoidal resection. Recently Öğrenci et al. [11] reported the first case of EG at CP angle in a 17-year old boy.

Schwannoma, meningioma and epidermoid cyst comprise the majority of CP angle lesions. Unusual pathologies which may arise at CP angle include arachnoid cyst, choroid plexus papilloma, medulloblastoma, ependymoma, atypical teratoid-rhabdoid tumor, chordoma, chondrosarcoma, lymphoma, exophytic brain stem glioma, lipoma, posterior fossa aneurysm and hydatid cyst [12,13].

Öğrenci et al. [11] analogized it to Pandora's box which we are not determinate whether to open it or not. Following incomplete resection of the CP angle EG, they achieved tumor regression by a trial of adjuvant steroids. They suggested that such intracranial lesions must be addressed with a trial of steroid therapy, prior to surgery, in children and young adults with high rates of eosinophils and lymphocyte in peripheral blood smear.

Here in, we report the first case of EG of CP angle in an adult patient. What has made our case further interesting is the hypointense appearance on T2-weighted MRI.

Albeit rare, this entity must be considered among the differential diagnosis of CP angle lesions. A trial of steroid is considered to have dramatic effects on tumor size and clinical recovery; how-

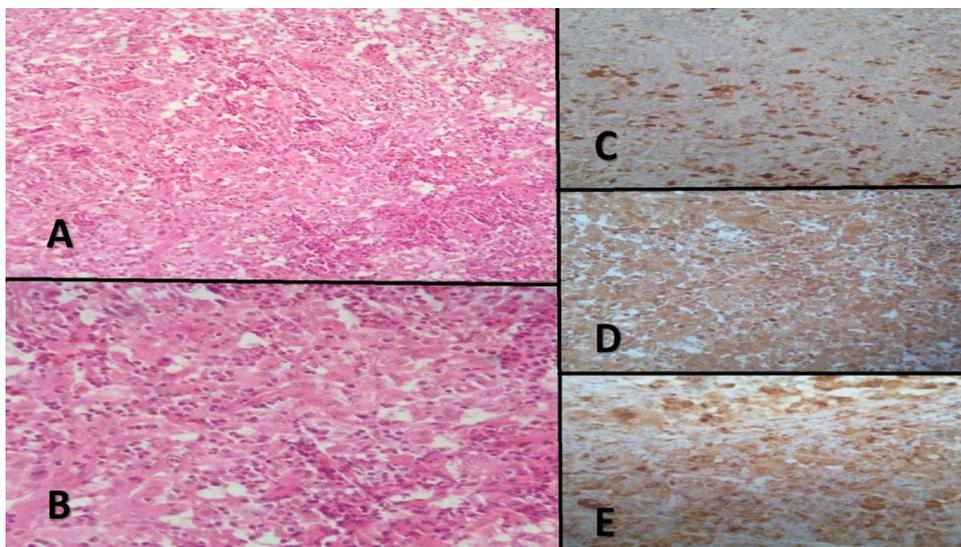


Fig. 2. A, B. Histopathological sections of the tumor show cellular sheet of cells with infiltration of large mononuclear cells (Arrow) with central nucleolus and vesicular chromatin and abundant eosinophilic cytoplasm and good number of eosinophils. (A. H&E, X40. B. H&E, X400.) C–E. Immunohistochemical analysis of the tumor shows immunoreactivity for S-100 (C), CD1a (D), Vimentin (E).

ever non-responders should undergo radiotherapy with or without chemotherapy for providing local control.

4. Conclusion

Eosinophilic granuloma (EG) should be considered as a rare differential diagnosis of CP angle lesions. Although the radiologic appearance is quite similar to meningiomas in this region, abundant eosinophils and positivity for CD1a, S-100 and vimentin renders the diagnosis unmistakable. To our knowledge this is the first report of EG at CP angle in an adult. Whenever total resection is impossible, tumor residues can be successfully managed by administration of low dose radiotherapy [14,15] and chemoradiotherapy.

This case report was adjusted according to SCARCE guidelines [16].

Conflict of interest

None declared.

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Ethical approval

N.A.

Consent

Informed consent has been taken from the patient.

Author contribution

AS and MT were the Surgeons and attending physicians of this Patient. ND prepared the manuscript. AD and ST provided the pathologic informations and slides.

Guarantor

Nima Derakhshan.

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