Article published online: 2022-09-20

Case Report

Intradural Eosinophilic Granuloma Invading Skull: Case Report and Review of the Literature

Abstract

Eosinophilic granuloma is a localized form of Langerhans cell histiocytosis, most commonly involving the skeletal system. Their origin from the dura is rare with only a handful of cases on record. We present one such rare case of an eosinophilic granuloma originating from the dura mater with secondary osseous invasion in an 11-year-old female child who presented with a swelling in the right parietal region. Magnetic resonance imaging demonstrated an enhancing mass with a wide dural attachment with a lytic lesion in the overlying skull. Right parietal extended craniotomy was done with the excision of mass from the dura. Histopathological features of mass were characteristic of eosinophilic granuloma which was confirmed by positive immunohistochemical staining for CD1a.

Keywords: *Dura, eosinophilic granuloma, immunohistochemical, Langerhans cell histiocytosis*

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Introduction

Langerhans cell histiocytosis (LCH) is a group of idiopathic disorders which are characterized by an abnormal proliferation of histiocytes. Eosinophilic granuloma is an older term for unifocal LCH. It is a slowly progressing disease characterized by an expanding proliferation of Langerhans cells most commonly involving the skeletal system.[1] Many reports of eosinophilic granuloma of the skull and the brain have been reported. However, there are only a few reports of intradural LCH in the literature.[2,3] We report a rare case of eosinophilic granuloma of the dura mater in an 11-year-old female child.

Case Report

An 11-year-old female child presented to Neurosurgery OPD with a swelling in the right frontoparietal region. She was neurologically intact, and there was no history of diabetes insipidus, exophthalmos, otitis media, skin lesion, or lymphadenopathy. An osteolytic lesion was detected on X-ray in the right frontal area. Magnetic resonance imaging demonstrated a well-defined enhancing mass measuring 3.5 cm × 3 cm × 2.5 cm with a wide dural attachment and an expansile lytic lesion in the frontal bone [Figure 1]. In chest X-ray and other radiological examinations, there were no other osseous or soft tissue lesions. The patient underwent right parietal extended craniotomy. Intraoperatively, there was a soft fleshy mass originating from the dura mater in the right parietal region and protruding out of bony defect. The lesion was completely removed together with a margin of grossly uninvolved dura and bone. A dural patch was formed with periosteum. The cranial defect was repaired with acrylic resin. The excised mass was sent to us for histopathological examination. Microscopically, there were sheets and clusters of Langerhans cells with typically grooved and irregularly contorted nuclei with a thin nuclear membrane, delicate chromatin, and inconspicuous nucleoli. The cytoplasm was abundant and lightly eosinophilic. Abundant eosinophils and frequent multinucleated giant cells were also present [Figure 2]. The Langerhans cells were immunoreactive for CD-1a [Figure 3]. The postoperative course was uneventful, and the patient was discharged 5 days after the operation.

Discussion

LCH (histiocytosis X) is a disease complex that includes Letterer–Siwe disease, Hand–Schüller–Christian disease, and eosinophilic granuloma. While the former two are systemic diseases, the latter is a localized form of histiocytosis. It is a nonneoplastic chronic disease of a granulomatous nature and unknown cause.

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How to cite this article: Mardi K, Thakur RC, Negi L. Intradural eosinophilic granuloma invading skull: Case report and review of the literature. Asian J Neurosurg 2017;12:698-700.

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Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_47_15

Quick Response Code:

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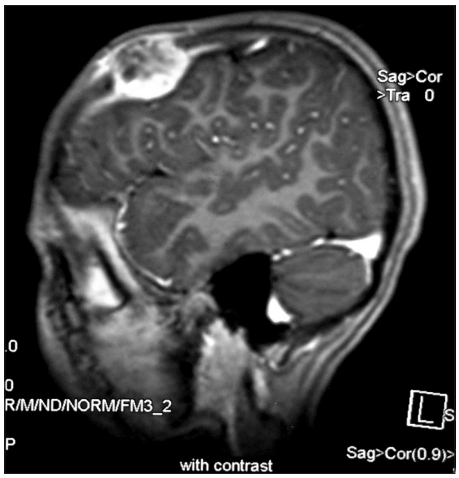

Figure 1: Magnetic resonance imaging showing an enhancing mass with a wide dural attachment and a lytic lesion in the overlying skull

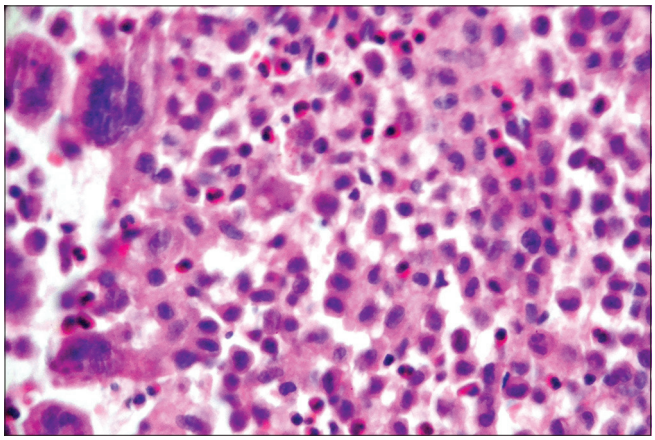

Figure 2: Histological examination showing a mixture of Langerhans cells and eosinophils along with multinucleated giant cells (H and E, ×40)

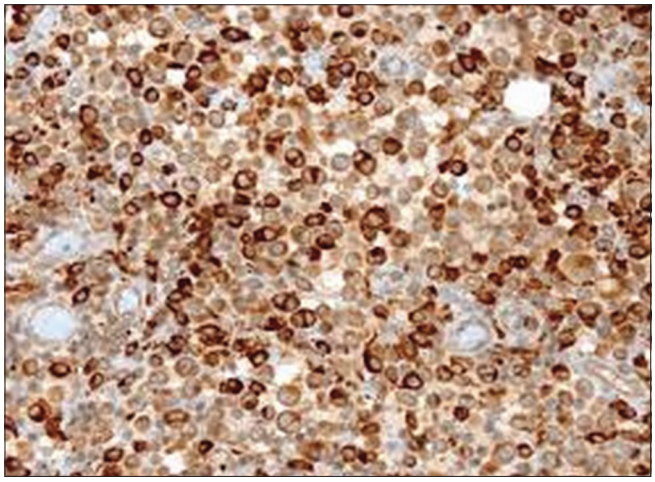

Figure 3: Langerhans' cells showing diffuse immunoreactivity for CD-1a (IHC, ×40)

Eosinophilic granuloma is the mildest form of LCH most commonly involving the skeletal system and rarely may show aggressive features. Central nervous system (CNS) involvement is unusual and frequently associated with bone lesions. Most of the patients with eosinophilic granuloma presented with local tenderness caused by involvement of calvarial bone.[4,5] CNS lesions are classified into two forms cerebral and meningeal.[6] Intradural development is a possible growth pattern of eosinophilic granuloma. An inflammatory process of the dural membrane with migration of Langerhans cells could be the physiopathological basis for the formation of intradural eosinophilic granuloma.[3]

In a literature review, only a few cases of eosinophilic granuloma originating from dura mater was found.[2,3] These case were similar to the present case and originated from the dura. Superficially, the tumor extended through the skull bone and deeply invaded the arachnoid plane and cerebral parenchyma. Carangelo *et al*. reported a case of LCH of skull with dural invasion, and they also showed Ki-67 nuclear antigen expression.[7]

The assumption that the LCH in our case was originated from the dura mater and invaded the overlying skull bone is supported by some observations. The rounded mass had a wide dural attachment which was the main factor leading us to think about dural origin. Clear implantation and greater extension of an eosinophilic granuloma on the dural plane also indicated its dural origin. After craniectomy was done and the bone was removed, eosinophilic granuloma remained attached to dura mater which also supported the dural but not cranial origin of LCH.

The differential diagnosis for intradural LCH includes several other diseases such as meningioma, meningosarcoma, metastases, tuberculosis, sarcoidosis, bacterial and fungal abscesses, and intracranial germinoma.[4,8] Radiological examination may be confusing, but pathological examination provides the basis for the definitive diagnosis of LCH.

Total surgical removal with normal tissue margins for a localized lesion is curative.[2,8,9] Radiotherapy, chemotherapy and radiosurgery may be used in the management of the LCH in aggressive and multifocal cases.[2,4,5,8,9]

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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