

Giant Occipital Intradiploic Epidermoid Cyst

Abstract

Intraparenchymal or intradiploic epidermoid cysts are very rare. Most of these cysts, when present, tend to involve the frontal and temporal lobes, and occasionally, the pineal gland or the brain stem. Here, we report a 45-year-old female, who presented with localized occipital headache and a tender occipital swelling, gradually increasing in size. She was hemodynamically and neurologically stable and did not have any focal neurological deficits. Whole skull and brain imaging revealed a well-demarcated expansile lytic lesion in the right occipital bone, which was hypointense on T1-weighted and hyperintense on both T2-weighted imaging and diffusion-weighted imaging without any contrast enhancement. The patient underwent a right occipital craniotomy and total excision of the intradiploic space occupying lesion. Histopathological examination confirmed the lytic bone lesion over occipital bone as intradiploic epidermoid cyst.

Keywords: Ectodermal inclusion cyst, epidermoid tumor, intradiploic epidermoid cyst, occipital epidermoid cyst, pearly tumor

Introduction

Intradiploic epidermoid cysts derived from the ectodermal cells of the cranium account for 1% of all intracranial tumors.^[1] Stratified squamous epithelium lines these cysts. Although rare, their common locations include the frontal, parietal, and occipital bones. These cysts grow very slowly. They usually present as painless bony swelling under the scalp. Headaches may occur due to erosion of the calvarium, and seizures due to local pressure. They may perforate the dura, rupture into the subarachnoid space, or involve the brain parenchyma. Microscopically, squamous epithelium lines the wall of the cystic tumor and lamellated keratin content fills the cyst.

Intraparenchymal or intradiploic epidermoid cysts are very rare, accounting for <5% of all intracranial epidermoid cysts.^[1] Conversely, atypical epidermoid cysts are rare, with intra-axial epidermoid cysts accounting for <1.5% of all intracranial epidermoid cysts^[2] and intradiploic epidermoid cysts accounting for ~3%.^[3] Eighty percent of reported intraparenchymal epidermoid cysts involve the frontal and temporal lobes, and occasionally, the pineal gland or the brainstem.^[4]

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Case Report

A 46-year-old female presented with a localized headache, in the occipital region, which had recently increased in severity. There was a history of a small tender swelling in the occipital region, which she noticed few months back. The swelling has been gradually increasing in size. The patient did not have any episodes of fever, seizures, double vision, or loss of consciousness. There was no significant past illness.

General examination did not show pallor, icterus, cyanosis, clubbing, edema, and lymphadenopathy. The patient was hemodynamically stable. There was a soft, mildly tender occipital lump, which had no mobility over the underlying bone. Skin over the swelling could not be retracted.

She was conscious and oriented with a Glasgow coma scale of 15/15 (E4V5M6). The pupils were equal and reactive to light. There were no gross motor/sensory deficits. All the deep tendon reflexes were normal. The plantar responses were flexor bilaterally. She did not have any cerebellar/meningeal signs. All other systems were within normal limits.

Ophthalmological evaluation showed visual acuity of 6/6 in both eyes. Eye movements were equal in all directions.

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There was no nystagmus. She had normal color vision. Visual field evaluation was normal. Fundus examination was normal.

Imaging

Magnetic resonance imaging (MRI) of the brain revealed a well-demarcated expansile lytic lesion (4.0 cm × 7.0 cm × 6.7 cm) in right occipital bone with thinning and disruption of inner and outer tables. The lesion was hypointense on T1-weighted and brightly hyperintense on T2-weighted imaging [Figure 1a and b]. There was no postcontrast enhancement [Figure 1c]. The lesion exhibited hyperintense signal on diffusion-weighted imaging (DWI) [Figure 1d]. MR spectroscopy did not show any intralésional choline. The differential diagnoses included plasmacytoma, giant cell tumor, or metastasis.

Management

In view of her symptoms, she underwent a right occipital craniotomy and total excision of the extradural/intradiploic space occupying lesion through a right paramedian incision in prone position under general anesthesia.

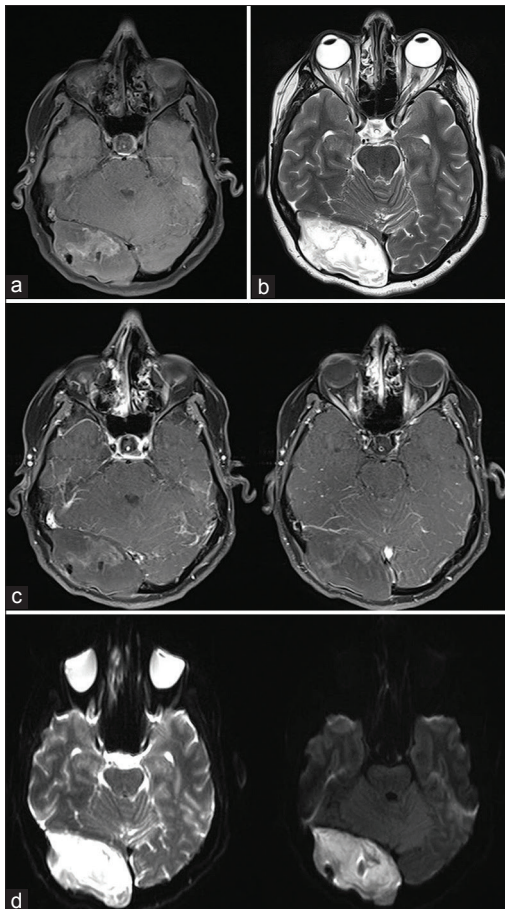


Figure 1: (a) T1-weighted sequence showing hypointense expansile lytic occipital lesion. (b) T2-weighted sequence showing hyperintense occipital lesion. (c) Postcontrast sequences showing occipital lesion without any contrast enhancement. (d) Diffusion-weighted sequence showing occipital lesion with hyperintense signals

Intraoperatively, the space occupying lesion was partly eroding the outer plate and almost completely eroding the inner plate. Dura mater was intact. The cyst capsule could be clearly separated from the dura and the involved bone. Cyst contained grayish white flaky puttylike material. The patient underwent a total excision of the entire cystic tumor with complete excision of the eroded bone and cranioplasty of the defect using titanium mesh.

Histopathological sections from the bony tissue showed a cyst lined by nonneoplastic stratified squamous epithelium with granular layer. Luminal aspect showed anucleate squamous and keratinous flakes without any skin appendages. The periphery of the cyst showed fibrosis with adjacent bone showing osteoclastic activity [Figure 2a and b].

Follow-up

Postoperative computed tomography (CT) brain showed total excision of the entire cystic tumor [Figure 3]. The patient was symptomatically better without any focal neurological deficits at 2 months of follow-up.

Discussion

In 1838, Muller reported the first primary intradiploic epidermoid cyst.^[5] In 1922, Cushing first described an intradiploic epidermoid cyst.^[6] Ciappetta *et al.* cited a total of 223 cases of intradiploic extradural epidermoids, reported in the literature by 1990.^[7] Intracranial epidermoid cysts are congenital lesions and also known as epidermoid tumor or “pearly tumor,” or ectodermal inclusion cysts, epidermoid cyst is a very slow-growing benign cyst. They arise from ectodermal cellular remnants, during 3–5 weeks of gestation, as a result of incomplete cleavage of neural ectoderm from the cutaneous ectoderm.^[7]

Epidermoid tumor commonly occurs within the third or fourth decade of life. They are painless subcutaneous scalp swelling covered with normal skin. These cysts are more commonly seen in men than women. Intracranial epidermoid cysts are well-circumscribed lesions, which most often occur within the diploe, in the parasellar region, Sylvian fissure, cerebellopontine angle, posterior fossa, and fourth ventricle.^[8] Intradiploic epidermoid

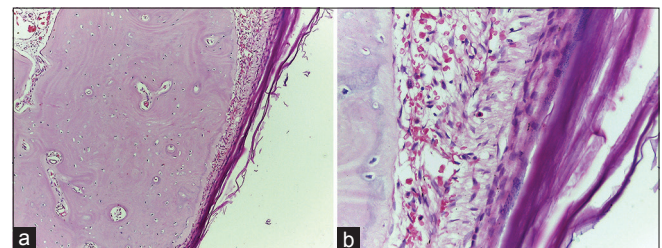


Figure 2: (a) Low power (×10) view showing cortical bone with cyst wall. (b) High power view (×40) showing cyst wall lined by stratified squamous epithelium with granular layer, lumen showing anucleate squames of lamellated keratin

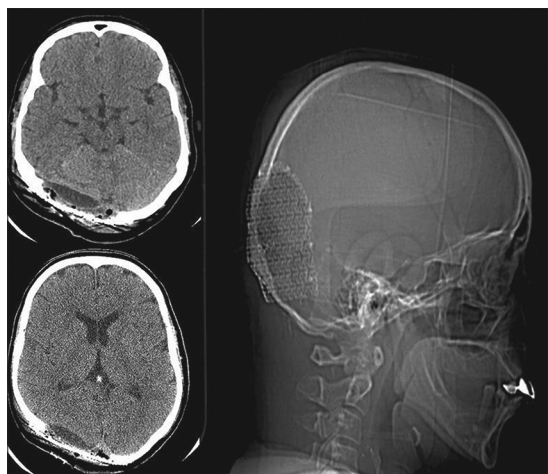


Figure 3: Postoperative computed tomography brain showing total excision of the entire cystic tumor and cranioplasty with titanium mesh

cysts are much less common than intradural lesions. Ectodermal inclusion cysts have a thin capsule of stratified squamous epithelium filled by keratin, cellular debris, and cholesterol. They do not contain hair or other dermal elements. This differentiates them from the dermoid tumor.^[9]

Intradiploic epidermoid cysts are hypoattenuating lesions on CT scans.^[4] They appear as low density, hypodense, nonenhancing lesions, similar to the fat density. MR images show inhomogeneous hypointense T1 signal and inhomogeneous hyperintense T2-fluid-attenuated inversion recovery (FLAIR) signal intensities.^[4] These lesions are often nonenhancing but may present with minimal peripheral contrast enhancement. Hyperdense cyst contents often mimic a hemorrhage. DWI is the best imaging sequence in diagnosing epidermoid cysts.^[7] They show restricted diffusion with higher signal intensity than that of cerebrospinal fluid (CSF) on DWI.

Spontaneous cyst rupture is a possible complication of intradiploic epidermoid tumors. This is often accompanied with the secondary discharge of the cystic content (keratin, cellular debris, and cholesterol) into the subarachnoid space, producing aseptic meningitis, leading to chronic granulomatous arachnoiditis.^[10]

The common differential diagnosis includes^[11] dermoid cyst, sebaceous cyst, eosinophilic granuloma, cavernous hemangioma, giant cell reparative granuloma, arachnoid cyst, neurocysticercosis, cystic tumors, plasmacytomas, and metastasis.

Dermoid cysts are often associated with the suture lines. They commonly occur in the orbital region and in the midline. They usually present in childhood and often contain fat. Sebaceous cysts present with fat density due to sebum. Eosinophilic granulomas are less heterogeneous. They are often tender. They are characterized by “beveled edge” appearance.

Cavernous hemangiomas have honeycomb or radiating sunburst appearance. Giant cell reparative granulomas are lytic lesions without sclerosis. On MRI, they are isointense to hypointense. They enhance intensely after gadolinium. Arachnoid cyst or mega cisterna magna is less lobulated. They follow CSF on all sequences including FLAIR sequences and DWI sequences.

Neurocysticercosis is an inflammatory cyst. They are smaller and multiple. They may enhance peripherally and may have associated edema. It does not show diffusion restriction.

Acoustic schwannoma and craniopharyngioma are cystic tumors. They usually have an identifiable solid enhancing part. Plasmacytomas are tumor of plasma cells. They may be solitary or a component of systemic multiple myeloma. Plasmacytomas are hyperdense on noncontrast CT and hyperenhancing on contrast CT. Sclerotic margins are absent, but they may have peripheral bone fragments.

Breast, lung, thyroid, and kidney cancers as well as neuroblastoma metastasize to calvarium. They have a variable appearance. They may be single or multiple masses with regular or irregular margins. They are seen as hyperintense lesions in CT due to increased vascularity.

The other less common differentials include calcified cephalohematomas, occult meningocele, and osteolytic intradiploic metastasis.

The only living and growing part of the intradiploic epidermoid cyst is its capsule. Hence, surgical removal of the tumor together with its capsule is the recommended treatment modality for intracranial epidermoid tumors.

Conclusion

Atypical epidermoids are very rare lesions with atypical imaging features including reversal of the typical T1 and T2 signal, as well as a lack of restricted diffusion. In spite of their slow growth, they can erode and invade adjacent structures and hence requiring meticulous neurosurgical intervention.

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Conflicts of interest

There are no conflicts of interest.

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