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Occipital intradiploic epidermoid cyst. Case report

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ABSTRACT

Intradiploic epidermoid cysts are rare intracranial tumours, with an estimated prevalence of less than 1% of all cranial tumours. Most of the intradiploic cysts are asymptomatic; however, clinical findings consist of headache and tenderness. We presented a case of a 21-year-old female who consulted the neurosurgery department for a period of three years about a mass in the occipital region, which was growing gradually. The patient was treated with surgical removal without complications. Intradiploic epidermoid cysts are infrequent intracranial tumours that primarily affect adults. This tumour has a wide spectrum of symptoms. Radiologic findings on computed tomography (CT) and magnetic resonance imaging (MR) are the cornerstone for the diagnosis, which is finally confirmed by histopathological studies. Surgical management with a complete resection of the cyst and capsule is the most commonly used treatment.

Keywords
epideroidl cyst,
occipital,
posterior cranial fossa,
bone tumo

INTRODUCTION

Intradiploic epidermoid cysts are rare intracranial tumors, with an estimated prevalence of less than 1% of all cranial tumors. (1) The origin is controversial but generally attributed to congenital origin, with defects in neural tube closure that give rise to inclusions of ectodermal cells. The area's most frequently affected are the frontal, parietal, cranial sutures, and cranial bases (2, 3). These defects are more common in men than women. (2, 4) In addition, epidermoid tumors occur most likely in the third and fourth decades of life. (4)

Most of the intradiploic cysts are asymptomatic; however, clinical findings consist of headache and tenderness. (3) Less common symptoms include traumatic rupture, focal neurological signs, intracranial hypertension, and seizures; furthermore, these findings are most likely to be present in large cysts. (3) The definitive treatment is a complete surgical resection, including the capsule to prevent recurrence. (1, 3)

CASE REPORT

A 21-year-old female with no prior medical history consulted the neurosurgery department for a period of three years of gradual enlargement of a tumor in the occipital region. On a physical exam, a

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mass on the right occipital region was palpable and had characteristics of soft texture and non-mobile consistency.

Cranial computed tomography (CT) and brain magnetic resonance imaging (MRI) conducted in other health institutions revealed an osteolytic lesion in the right occipital cranial diploe. This lesion, located near the junction of the transverse sinus with the sigmoid sinus at the ponto-cerebellar angle, did not exhibit contrast enhancement. Additionally, there is a potential presence of a fistulous trajectory. The patient was treated with surgical removal. The surgery commenced with making an incision in the occipital region at the level of the superior nuchal line, careful desperiotization, and decortication. Subsequently, an intradiploic lesion was identified and dissected until a complete lesion was achieved. Various fistulous tracts were discovered along the midline, which are necessary to be reamed and resected. The cessation and occlusion of venous bleeding originating from a tributary branch is necessary. Ultimately, the bone deficiency is remedied through the utilization of a circular plate that is securely affixed in place using screws. The treatment was successfully concluded without any difficulties.

The patient received analgesic treatment, and a control cranial CT reported post-surgical changes in the right occipital region with osteosynthesis material, edema, and emphysema of soft tissues in the right occipital region (Figure 1).

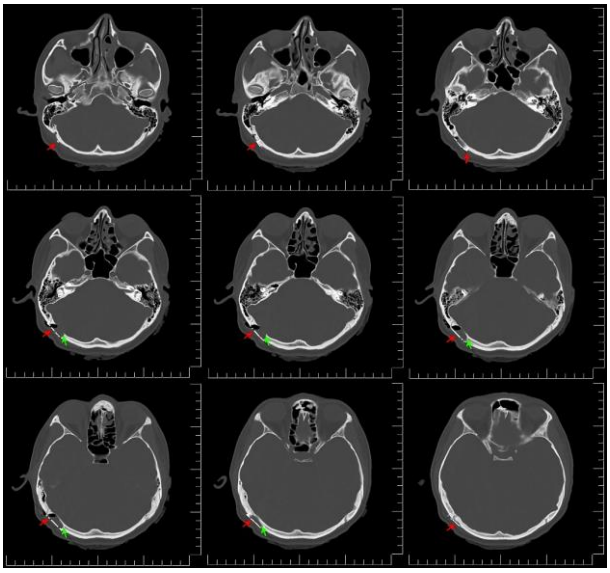

Figure 1. A CT of the head showing a rounded area with well-delineated sclerotic margins of bone destruction (green arrows) and post-surgical changes with osteosynthesis material covering the defect (red arrows).

A histopathological test confirmed the diagnosis of an intradiploic epidermoid cyst (Figure 2). Finally, the patient was discharged to continue outpatient care.

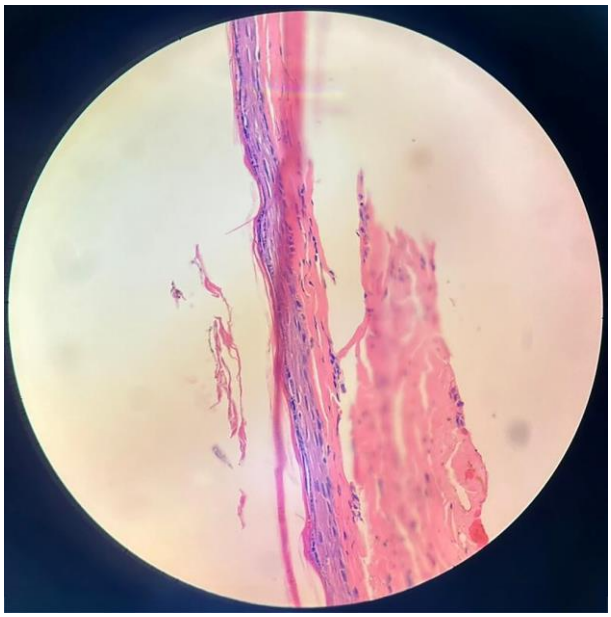

Figure 2. Hematoxylin-eosin staining of the tissue specimen removed during the surgical procedure shows the presence of a cyst with epithelial cells without atypia and containing lamellar keratin compatible with an intradiploic epidermoid cyst.

Discussion

Since the first description of an intradiploic cyst in 1922, there have been little more than two hundred cases reported in the literature. (4, 5) The ectodermal cysts have a capsule of stratified squamous epithelium with keratin, cholesterol, and cellular debris, and they do not include any dermal elements, which is the main difference with dermoid tumors. (5)

Regarding diagnosis, a CT scan or MR images are the cornerstone to detecting this rare anomaly. In a CT scan, they have low density, so they are hypodense without any enhancement. (5) The MRI can report hypointense lesions in T1, hyperintense lesions in T2, and fluid-attenuated inversion recovery (FLAIR) without enhancement upon application of intravenous contrast. According to Law EK et al. (2015), epidermoid and cysts can have atypical imaging findings, being able to be T1 hyperintense and T2 hypointense with a lack of restricted

diffusion. (6) The opposite pattern of a typical epidermoid cyst can be present due to the T1 and T2 weighted MRI signals being strongly influenced by protein content, which also explains the lack of restricted diffusion. (6)

The radiological differential diagnoses include primarily dermoid cysts and arachnoid cysts; abscesses and metastasis are less frequent. (6, 7) Classically epidermoid cysts are described as soft lessons that are conforming brain structures or insinuating between them with the classic T1-hypointense and T2-hyperintense patterns. (7) On the other hand, arachnoid cysts are isointense with the cerebrospinal fluid (CSF) in all sequences, and they do not restrict on diffusion-weighted images. The dermoid cysts are usually located along the midline and appear like fat, not CSF. (7) The definitive diagnosis is by histopathological study, which reported stratified epithelium that it is desquamative on keratin. (1) Our patient´s lesion showed these histopathological findings, confirming the diagnosis.

The optimal course of treatment is a surgical approach that prioritizes the excision of the cyst and full tumour capsule. This procedure aims to alleviate brain congestion and facilitate subsequent histopathological examinations. Certain tumors have intracranial involvement affecting various intraparenchymal structures, resulting in incomplete resection. (8,9,10) The recurrence rate ranges from 1% to 54% and can potentially be mitigated by devitalizing the remaining capsule fragments during the surgical procedure. (11)

Conclusion

Intradiploic epidermoid cysts are infrequent intracranial tumours that primarily affect adults within the age range of the second to fourth decades of life. This tumour has a wide spectrum of symptoms. Radiologic findings on CT and MR are the cornerstone for the diagnosis, which is finally confirmed by histopathological studies. Surgical management with a complete resection of the cyst and capsule is the most commonly used treatment.

Abbreviations

CSF: cerebrospinal fluid
CT: computed tomography
FLAIR: fluid-attenuated inversion recovery
MRI: magnetic resonance imaging

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