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JSM Neurosurgery and Spine

Case Report

Intradiploic Epidermoid Cyst of the Parietal Bone: Case Report with Literature Review

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Submitted: 11 June 2023

Accepted: 03 July 2023

Published: 06 July 2023

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OPEN ACCESS

Keywords

- Epidermoid cysts
- Skull tumor
- Calvarial defect
- Radical removal

Abstract

Authors describe an intradiploic epidermoid cyst of the parietal bone with intra and extracranial extension. A 23-years old male patient with a history of subcutaneous swelling 3 months ago on the right parietal scalp. CT and MRI evaluation revealed a well defined extracerebral mass in the parietal scalp with destruction of both the inner and outer tables of the calvaria. The patient underwent a total resection of the tumor and the histological examination suggesting a epidermoid cyst. The patient post-operative course was uneventful, without any neurological deficit. Intradiploic epidermoid cysts of the skull are rare, slow growing, mostly benign tumors. The prognosis is good when the cyst is totally resected with its capsule.

ABBREVIATIONS

EC: Epidermoid Cyst; CT: Computed Tomography; MRI: Magnetic Resonance Imaging

INTRODUCTION

The epidermoid cyst (EC) is a benign dysembryoplastic lesion that accounts for less than 0.3 to 2% of all primary intracranial tumors [1]. Intradiploic forms represent 25% of all intracranial ECs [2]. They may appear as small isolated and perfectly benign swelling of the scalp typically well and long tolerated [3]. Modern imaging including Computed tomography (CT) and Magnetic resonance imaging (MRI) plays an important role in the diagnostic and surgical approach. The prognosis is generally good after complete excision. We report a new case of parietal bone cyst with extracranial and intracranial "iceberg" extension. The diagnosis was suspected on radiological imaging data and confirmed by pathological examination with good follow-up after surgical excision.

CASE REPORT

A 23-year-old man with no medical history presented with a subcutaneous lesion on his right parietal scalp for 3 month ago. At observation, we revealed a mobile unpainful mass, 5 cm in diameter, elastic and soft. It was also adherent to the surface. The bone around seemed to be intact at the limit of the clinical observation. Neurological examination was entirely normal and no cognitive deficits were found. Routine biochemical and hematologic parameters were also normal without imflammatory biological syndrome.

CT demonstrated a cystoid lesion of the parietal bone, with calvarial defects on the right parietal bone. The outer and the inner table of the skull bone were widely destroyed; in some areas, the inner table was thinned out (Figure 1).

Cerebral MRI showed the lesion with hypointense signal characteristic on T1-weighted images without enhancement after gadolinium administration. MRI demonstrating a light bulb bright appearance on diffusion-weighted imaging and characteristics consistent with an epidermoid. The lesion demonstrated a mild mass effect on the adjacent brain parenchyma; however, no signal abnormalities or pathologic post-contrast enhancement were noted. There was smooth remodeling of the calvarial margins abutting the lesion consistent with a benign, slow-growing process. Ventricles and cisterns appeared of normal size, shape, and configuration. The underlying brain parenchyma appeared unremarkable, with no other mass, bleed, acute infarct, or midline shift (Figure 2).

The patient underwent a right parietal craniotomy. When the scalp flap was reflected, a white encapsulated, mostly intradiploic large tumor was identified. The 5 cm tumor was herniating through bone defect. The tumor was resected within the apparently normal bone followed by the excision of the markedly thickened dura. A duroplasty with a galeal flap and a reconstruction of the skull bone were performed (Figure 3). His postoperative course was uneventful.

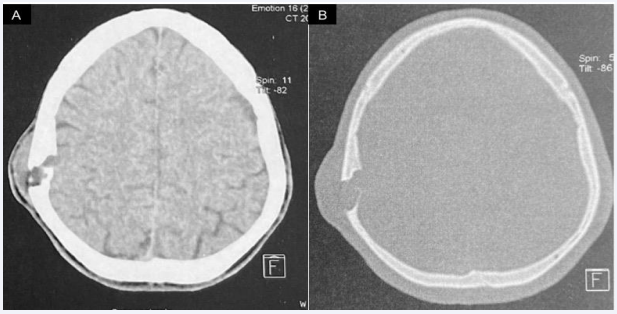

Figure 1 (A) Axial CT scan in soft-tissue window showed an inhomogeneous intradiploic solid mass in the right parietal region with areas of calcification. (B) Axial CT scan in bone window demonstrated osteolytic changes with erosion of both tables of the vault.

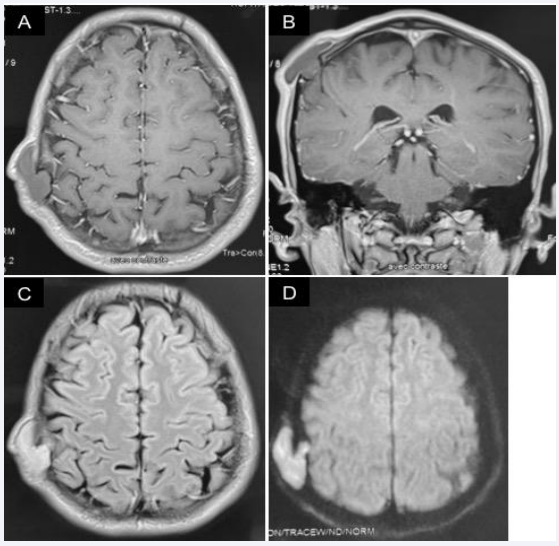

Figure 2 T1 weighted Magnetic Resonance Imaging (MRI), axial view (A) and coronal view (B) showing a hypointense lesion in the right parietal region without enhancement after gadolinium administration with mild thickening of the dura. FLAIR: Fluid-attenuated inversion recovery (C) and Diffusion Weighted Imaging (D) showing hyperintense lesion.

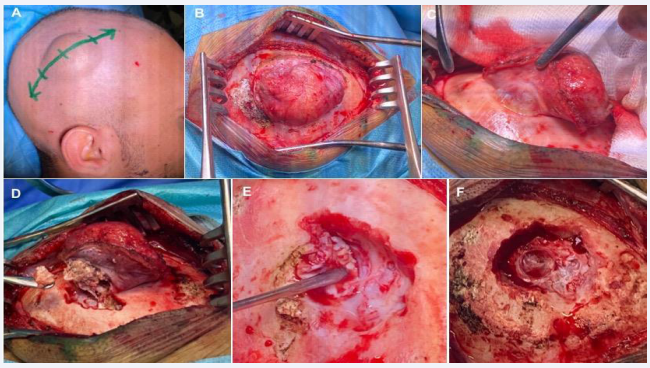

Figure 3 Intraoperative photograph showing:
(A) Mark of the incision. (B) Exposure of the scalp lesion. (C) Bone defect with intracranial extension of the lesion. (D) Widening of the craniotomy, thus revealing the intracranial part of the lesion « pearly contents ». (E) Capsule of lesion being dissected off the thinned dura. (F) Final aspect after complete excision, arachnoïd membrane herniated throught circular defect of the dura.

Histological examination through Hematoxylin-and-eosin–stained sections revealed lamellar keratinous material, cholesterol crystals, and cellular debris. No malignant features were observed. The cyst wall showed many forms of keratinization. Bone invasion was not seen. Theses pathological findings were consistent with the diagnosis of intradiploic epidermoid cyst.

DISCUSSION

In 1838, Müller was the first to describe an intradiploic EC of the bone [4]. All ages may be involved, even cases discovered prenatally [5], however it is mostly children and youngs adult who are interested at the time of diagnosis with an average age of 34 years [6], with no clear sex predominance.

The pathogenesis of these cysts is the subject of several controversies, mainly between congenital theory and acquired post-traumatic theory. result mainly from entrapped ectodermic embryonal remnants (ectodermic cell rests between third and fifth gestational weeks) because of incomplete cleavage of neural crest and a sequestration of skin elements within skull bones; some authors imply an acquired state due to trauma and implantation of epidermal cells in the connective tissue of the diploe [7]. Whether post-traumatic or dysembryoplastic, these lesions are benign and slow-growing that grow linearly at variable rate between 0.16 and 1.3 cell generations per month [8].

Clinically, these lesions may appear as benign swelling of the scalp well and long tolerated. However, the giant cysts, evolving towards the deep part of the vault, may be able to compress the underlying brain and cause certain neurological deficit, which can range from a few headaches to a true intracranial hypertension, Seizures and focal neurological deficits [7].

For the radiological diagnosis, CT and MRI allow for the good assessment of both skull involvement and intracranial extension. The typical CT aspect is a large homogenous hypodense mass, with or without calcifications [9]. The characteristic MRI findings of intradiploic epidermoid cysts include well-demarcated osteolysis, high signal intensity on T2-weighted images, and varied signal intensity on T1-weighted images [10]. Some observations have reported heterogeneous appearance of some EC, explained by the variability of the proportion between the fat, fluid and, calcium component [11].

For pathologic findings, epidermoid cysts have a thin capsule of stratified squamous epithelium filled by keratin, cellular debris and cholesterol, and do not contain hair or other dermal elements [12]. The differential diagnosis includes dermoid cysts, eosinophilic granuloma, fibrous dysplasia, osteomyelitis, and metastatic lesions.

A possible complication of intradiploic epidermoid tumors is the spontaneous cyst rupture with secondary discharge of the cystic content (keratin, cellular debris and cholesterol) into the subarachnoid space, producing aseptic meningitis which frequently leads to chronic granulomatous arachnoiditis [13]. The goal of surgery in the treatment of epidermoid tumors is total surgical removal of the tumor together with its capsule, wich is the only living and growing part of the tumor.

CONCLUSION

Intradiploic epidermoid cysts of the skull are rare, slow growing, mostly benign tumors. However complications are not uncommon. CT and MRI plays an important role in the diagnostic and surgical approach. The prognosis is generally good after complete excision of the tumor and its capsule, with the goal of preventing recurrences, inflammation and the possibility of malignant transformation.

DECLARATIONS

Authors' contributions

HB contributed to the conception, drafting, and reporting of the case. AM and AA acquired the clinical data. HB and AA contributed to the revision of the manuscript. All authors have read and approved the final manuscript.

Availability of data and materials

All data generated and analysed in this study are included in this article.

Consent for publication

The patient provided written informed consent for publication of patient clinical details and clinical images.

Competing interests

The authors declare that they have no competing interests.

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