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Letter to Editor

Intradiploic epidermoid cyst of the temporal and occipital bone with intracranial extension

Epidermoid tumours are rare, benign, slow growing, congenital tumours composed of epithelial cell remnants and cholesterol [1]. These tumours represent approximately 0.2–1.8% of all intracranial neoplasms. They are thought to form from surface ectoderm, which becomes enclosed within the neural tube between the 3rd and 5th week of embryogenesis. Tumour enlargement is associated with the processes of desquamation and degradation of cell membrane, and leads to development of clinical symptoms. Epidermoid cysts most often present in patients over forty years of age with headaches and signs of raised intracranial pressure with further symptoms determined by the location of the cyst. Cushing described intradiploic epidermoids in 1922 [2]. Most common localization is intradural (90%); especially in the cerebellopontine angle and parasellar region; however intradiploic intraosseous epidermoids occur usually accompanied by milder clinical symptoms [3,4]. Computed tomography (CT) imaging generally reveals a nonenhancing, lobulated, hypodense mass with well defined borders. On magnetic resonance imaging (MRI) epidermoid cists are hypointense on T1-weighted, and hyperintense on T2-weighted, diffusion-weighted imaging (DWI) and fluid-attenuated inversion recovery (FLAIR) sequences [5]. In differential diagnosis the dermoid cyst, haemangioma, eosinophilic granuloma, aneurysmal bone cyst and fibrous dysplasia should be taken into account. When radiologically confirmed, epidermoid tumours should be surgically completely removed because of possible complications e.g. recurrence or granulomatous meningitis. Malignant transformation of epidermoids occurs rarely although has been reported [6]. We report a rare case of intradiploic epidermoid cyst of the temporal and occipital bone with a posterior fossa component. Approval of the institution Ethical Committee was not necessary for the case. Informed consent of the patient was obtained. A 56-year-old male presented with a history of dizziness, balance disturbances and memory disorders, which lasted for a few weeks. Neurological examination showed no abnormalities. Head CT was performed on Toshiba Aquilion Scanner and showed expansive, intradiploic lesion of left temporal and occipital bone, posteriorly to temporal pyramid and mastoid process. The outer and inner table of the diploic space was involved and destroyed with the tumour intracranial extension localized in the adjacent left cerebellar hemisphere. The tumour was predominantly hypodense on CT scan with a small hyperdense region in the posterior fossa portion what was supposed to be an area corresponding with blood. MRI was performed subsequently on Simens Avanto 1.5 T and showed an expansive, intradiploic mass that penetrated to the posterior cranial fossa having well defined borders and causing a slight mass effect with no surrounding oedema. No considerable contrast enhancement was observed (Figure 1). The patient was qualified for surgical removal of the lesion. A left-sided, vertically oriented, retromastoid skin incision was performed. The tumour related, complete bone destruction was revealed with a glossy, lobulated mass penetrating the dura and herniating into the left cerebellar hemisphere. There was macroscopic evidence of old blood products however it was not confirmed in further histologic examination. The cyst was dissected completely and subsequent dura- and cranioplasty was performed. In postoperative course, the patient temporarily presented mild symptoms of aseptic meningitis, which was successfully treated with steroids and antiedematous therapy. Neurological condition was intact. Histological examination revealed keratin contents and fragments of the squamous epithelium. The patient was discharged home and remains under the care of the clinic. Complete relief of the preoperative symptoms was observed. Review of the literature revealed that intradiploic epidermoids were first described in 1838 and have since been reported sporadically in small groups of patients [7]. Moreover, an intraparenchymal extension of intradiploic epidermoids occurs extremely rarely and only a few cases have been reported so far. In the patient we present, a considerable posterior fossa part of the lesion was found. In clinical course, intradiploic epidermoid cyst usually presents as a painless, slowly growing lump palpable on the scalp. In the case we report, there was neither scalp swelling nor abnormalities on palpation. Diagnostics was based on the clinical symptoms, which were cerebellar disorders and memory disturbances, however in other patients, severe neurological deficits or epileptic seizures can develop due to the tumour size and location. A lesion can involve the inner and (more

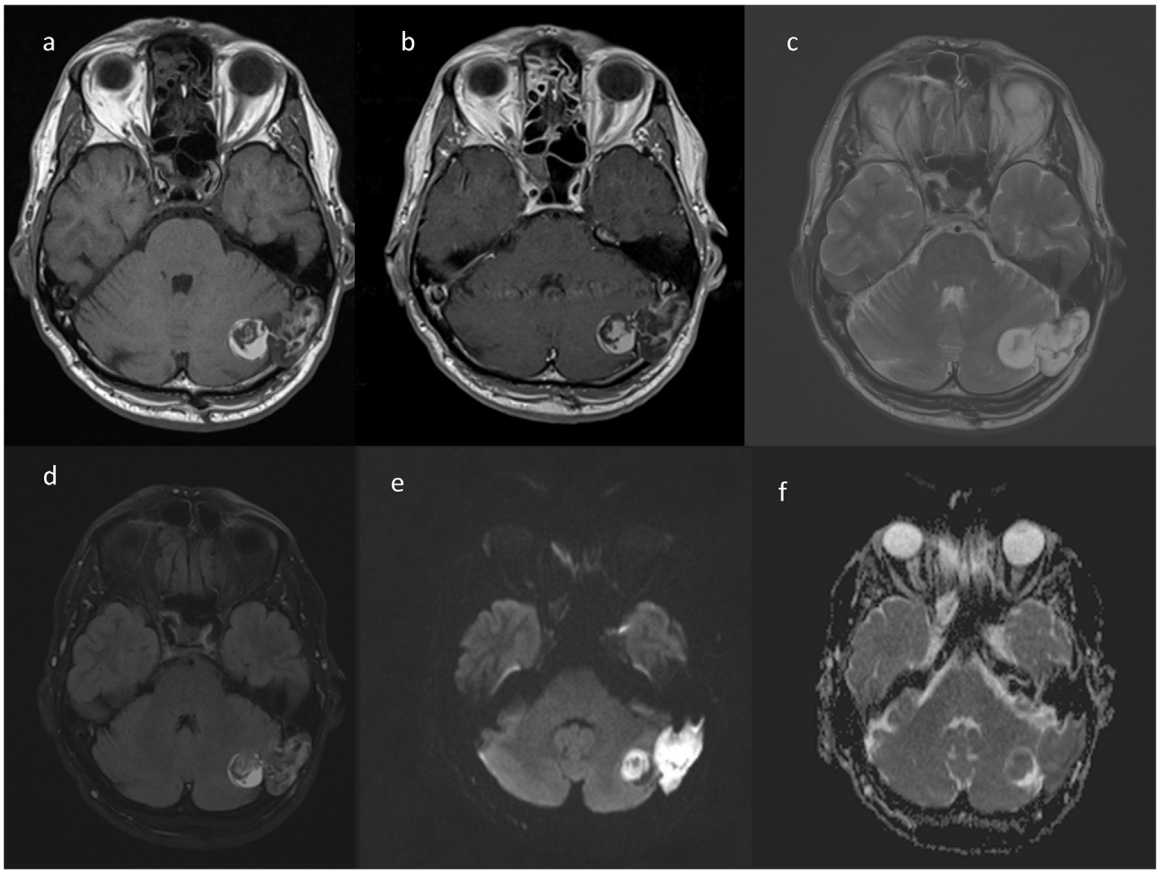

Fig. 1 – (a) Axial T1-weighted image shows mixed-intensity retropyramidal posterior fossa mass with intracerebellar component being more hyperintense. (b) Axial T1-weighted post-gadolinium image demonstrated no enhancement. (c) Axial T2-weighted image shows mostly hyperintense CSF-similar lesion with some irregular hypointense areas. (d) Axial FLAIR image shows mixed intensity lesion with marked hyperintense intracerebellar portion. (e) and (f) DWI b1000 image and ADC map show restricted diffusion in the lesion.

frequently) the outer table of the skull bone. In the patient, the both tables were destroyed with explicit destruction of the inner table and combined with posterior fossa extension of the tumour. Epidermoid cysts are hypodense on CT due to the fat content, however, when a volume of cell debris is growing, the tumour density may become similar to water/CSF. A higher density of epidermoids can be associated also with increased protein content or calcifications. In the described patient, the tumour was mostly hypodense on CT but a small hyperdense region was also noticeable being supposed of possible bleeding into the lesion; what was confirmed intraoperatively. No CT contrast enhancement was observed in the tumour what is typical for epidermoids. On MRI, the signal intensity was not homogeneous, especially on T1-weighted image, where areas of increased signal were displayed due to the possible composition of lipids, cholesterol, keratin or intracystic haemorrhage. FLAIR sequence showed mixed intensity lesion with a hyperintense region in the intracerebellar portion, what was only partially helpful since epidermoids usually demonstrate a high FLAIR signal due to cholesterol and epithelial remnants. A DWI sequence demonstrated the characteristic for epidermoids, hyperintense lesion signal due to diffusion restriction. Apparent diffusion coefficient (ADC) map showed a low signal of the tumour. In some patients spontaneous or iatrogenic cyst rupture can cause aseptic meningitis [8]. In this patient, episode of mild aseptic meningitis was observed postoperatively but the patient fully recovered. Some authors reported cases of intradiploic epidermoid cyst presenting as a spontaneous intracerebral haematoma [9]. Remnants of bleeding into the cyst were also found during the surgery.

Conclusions

(1) Epidermoid cysts are benign, congenital, predominantly intracranial tumours, which grow slowly. Intradiploic epidermoid cyst is a rare condition especially when accompanied by parenchymal involvement.
(2) Radiological diagnostics of epidermoids sometimes is a difficult challenge, however in most cases MRI study is sufficient for accurate diagnosis.
(3) Epidermoids should be completely and carefully excised to avoid recurrency and aseptic inflammation.

Conflicts of interest

None declared.

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Piotr Dąbrowski[a,b,*]

[a]*Department of Diagnostic Imaging, Western Hospital in Grodzisk Mazowiecki, ul. Daleka 11, 00-825 Grodzisk Mazowiecki, Poland*

[b]*Department of Neurosurgery, Western Hospital in Grodzisk Mazowiecki, ul. Daleka 11, 00-825 Grodzisk Mazowiecki, Poland*

Rafał Kidziński

Emilia Frankowska

Department of Diagnostic Imaging, Western Hospital in Grodzisk Mazowiecki, ul. Daleka 11, 00-825 Grodzisk Mazowiecki, Poland

Rafał Górski

Maciej Bujko

Department of Neurosurgery, Western Hospital in Grodzisk Mazowiecki, ul. Daleka 11, 00-825 Grodzisk Mazowiecki, Poland

*Corresponding author at: Department of Diagnostic Imaging, Western Hospital in Grodzisk Mazowiecki, ul. Daleka 11, 00-825 Grodzisk Mazowiecki, Poland

E-mail address: piotrdabrowski2000@gmail.com (P. Dąbrowski)

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