INTRODUCTION

Epidermoid cysts are uncommon tumors, accounting for less than 2% of intracranial tumors [1]. These tumors originate from the remaining ectodermal tissues when the neural tube is closed at the third to fifth gestational weeks [2]. According to their location, epidermoid cysts are classified as extradural and intradural [1]. As one of the extradural types, intradiploic epidermoid cysts occur in any part of the skull and are even rarer tumors [3]. They usually present as a painless local swelling under the scalp. Rarely, they show dural invasion and compress the brain parenchyma [1,4]. Here, we present a rare case of a giant intradiploic epidermoid cyst of the skull presenting with progressive headache.

CASE REPORT

A 57-year-old woman presented with a 1-year history of localized headache in the occipital area. The symptoms were aggravated 2 weeks prior to arrival at our institution. Physical and neurological examination revealed no specific findings. CT and MRI showed an extradural mass measuring 50 × 70 mm in the occipital bone with bony destruction. The patient underwent surgical resection. The tumor was completely removed with its capsule. There was no extension to the intradural space. The pathological report confirmed that the tumor was an epidermoid cyst. Follow-up MRI 24 months after the operation showed no recurrence. The headache was well controlled without any medications. We report a rare case of intradiploic epidermoid cyst with clinical and radiologic features and surgical treatment. It is important to consider this diagnosis for a patient with persistent regional headache with or without a growing scalp mass.

Key Words

Epidermoid cyst; Skull; Occipital bone; Headache; Skull neoplasm.

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Giant Intradiploic Epidermoid Cyst

cyst. The cyst was unilocular and consisted of a grayish brown, sticky material. The wall of the cyst was gray-white and smooth (Fig. 2A, B). The mass was lined by mature squamous epithelium and filled with laminated keratin material (arrow and arrowhead, respectively, Fig. 2C).

Follow-up MRI 24 months after the operation showed no recurrence (Fig. 3). At the last clinical follow-up 24 months after the operation, the headache was well controlled without any medications.

This study was approved by our Institutional Review Board and followed the Declaration of Helsinki (IRB No. 2020-03-033-001). Identifying details (names, dates of birth and other information) were not included in this study. Informed consents were received from the patient.

DISCUSSION

Epidermoid cysts are slow-growing benign congenital lesions. They originate from ectodermal remnants as a result of incomplete cleavage of neural ectoderm from the cutaneous ectoderm in the neural groove during the neural tube closure phase [2,3]. Epidermoid cysts can arise from any part of the craniospinal neural axis, with 90% of epidermoid cysts in the intradural space and 10% of epidermoid cysts in the extradural space [5]. In epidermoid cysts, rupture, hemorrhage, or aseptic meningitis can occur as complications [1,6]. Malignant transformation of epidermoid cysts has rarely been reported [7].

Intradiploic epidermoid tumors usually occur as a result of entrapped ectodermal remnants within the skull bones or, rarely, secondary to trauma [2,8]. In this case report, the patient had no history of head injury prior to the diagnosis of this disease. Therefore, the mass in this case was considered a primary
T1-weighted gadolinium-enhanced MRI usually shows no contrast enhancement of these lesions [22]. Differential diagnoses should include dermoid cysts, hemangiomas, arachnoid cysts, cholesterol granulomas, and/or osseous tumors such as aneurysmal bone cysts, fibrous dysplasia, and eosinophilic granulomas [7,19,24]. Dermoid cysts are more frequently diagnosed in childhood and are usually located along the midline [24]. Hemangiomas have a typical appearance, including a honeycomb or radiating sunburst pattern [24]. Arachnoid cysts, which are very rarely located in the skull, similar to intradiploic cysts, can be distinguished from epidermoid cysts on DWI, where the latter usually present with high signal intensity while the former demonstrate low signal intensity [25-27]. Eosinophilic granulomas usually occur in children and young adults and show contrast enhancement on MRI [24]. In our case, although the mass was difficult to differentiate from other osteolytic skull lesions, our radiological diagnosis was intradiploic epidermoid cyst based on the patient's age and radiological findings, including the paramedian location of osteolytic skull lesion, the lack of dural invasion and contrast enhancement, and diffusion restriction on MRI.

Surgical indications for epidermoid cysts include increased size, progression of neurological deficit, and/or malignant transformation [4]. In intradiploic epidermoid cysts, complete surgical excision followed by appropriate cranioplasty is the treatment of choice. Total removal of these lesions, including the cyst wall, is associated with a good prognosis [3,15,28]. Arko et al. [17] analyzed the outcome of a total of 169 intradiploic epidermoid cysts that were previously described. The authors reported that the recurrence rate was only 5.8%, and of these recurrent cases, malignant transformation to squamous cell carcinoma occurred in 44% (4 of 9 cases) [17]. Most of these cases might be related to incomplete resection of primary in-
tradiploic epidermoid cysts [17]. We completely removed the epidermoid cyst with its capsule from our patient (Fig. 2). The patient was free of recurrence at the 24-month follow-up.

In this article, a rare case of intradiploic epidermoid cyst located in the occipital bone is presented with clinical and radiologic features, and surgical treatment. It is important to consider this diagnosis in a patient who presents with persistent regional headache and/or a slowly growing scalp mass. Correct radiologic evaluation and complete surgical resection are important for favorable long-term outcomes.

Fig. 3. A follow-up MRI at 24 months after surgery shows no residual or recurrent mass. A: T2-weighted MRI. B: T1-weighted MRI. C: Diffusion-weighted imaging. D: T1-weighed gadolinium-enhanced MRI.

Conflicts of Interest
The authors have no potential conflicts of interest.

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