Huge intradiploic epidermoid cyst

Olbrzymia torbiel naskórkowa śródkościa

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Abstract

A 60-year-old man presented with an occipital mass under the scalp and complained of headache, nausea, and dizziness. Magnetic resonance imaging showed a well-defined mass in the occipital scalp extending from the scalp through the cranium and several centimetres into the posterior fossa. There were well-defined margins in the deep portion and the mass was totally removed. Histological examination showed that the cystic structure was lined by squamous epithelium containing laminated keratin material. The pathological findings were consistent with the diagnosis of an epidermoid cyst. The patient was discharged free of symptoms.

Key words: cyst, epidermoid, intradiploic, huge, occipital.

Streszczenie

Mężczyzna, lat 60, zgłosił się z powodu guza podskórnego okolicy potylicznej, skarżąc się na bóle głowy, nudności i nieukładowe zawroty głowy. W badaniu za pomocą rezonansu magnetycznego uwidoczniono wyraźnie zarysowany guz podskórny okolicy potylicznej, szerzący się z tkanki podskórnej poprzez czaszkę na głębokość kilku centymetrów do tylnej jamy czaszki. Położona głęboko część guza miała wyraźne granice i guz został usunięty w całości. W badaniu histopatologicznym stwierdzono obecność torbieli wyścielonej naskórkiem wielowarstwowym płaskim zawierającym blaszki keratyny. Wyniki badania histopatologicznego pozwoliły na rozpoznanie torbieli naskórkowej. Chory został wypisany ze szpitala bez objawów.

Słowa kluczowe: torbiel naskórkowa, olbrzymia, potyliczna, śródkoście.

Introduction

Intradiploic epidermoid cysts are rare, benign tumour lesions which arise between the two tables of cranial bones [1]. Intracranial epidermoid cysts account for 1% of all intracranial tumours [2]. They are generally located in the cerebellopontine angle, petrous apex, para-pituitary region, and chiasmal region. Less frequently, intradiploic epidermoid cysts are observed [3]. They may often reach an enormous size without producing neurological symptoms [4]. The goal of surgical treatment should be total resection of the tumour with its capsule [3]. The long-term prognosis is excellent after successful resection [5]. In this paper, we report an unusual case of a huge intradiploic epidermoid cyst.

Case report

A 60-year-old male patient visited our clinic with complaints of headache, nausea, dizziness and an occipital mass under the scalp. The patient had suffered from

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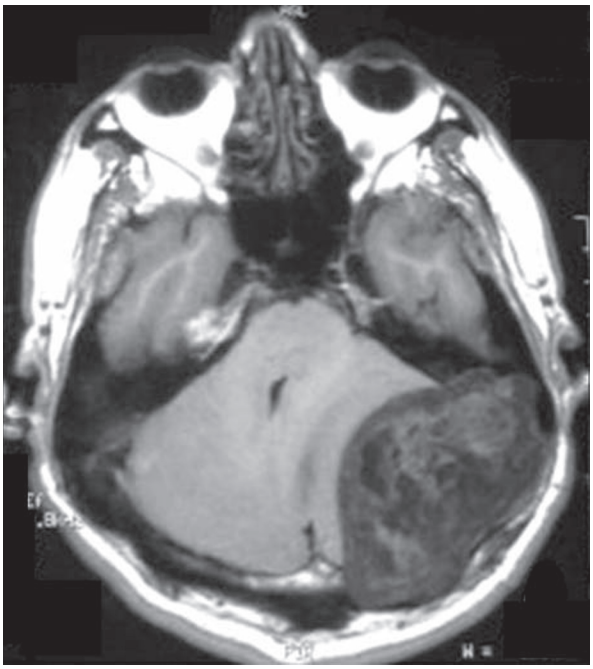

Fig. 1. Preoperative axial T1-weighted magnetic resonance image showing a heterogeneous signal mass with well-defined margins in the left posterior cranial fossa and compressed cerebellum

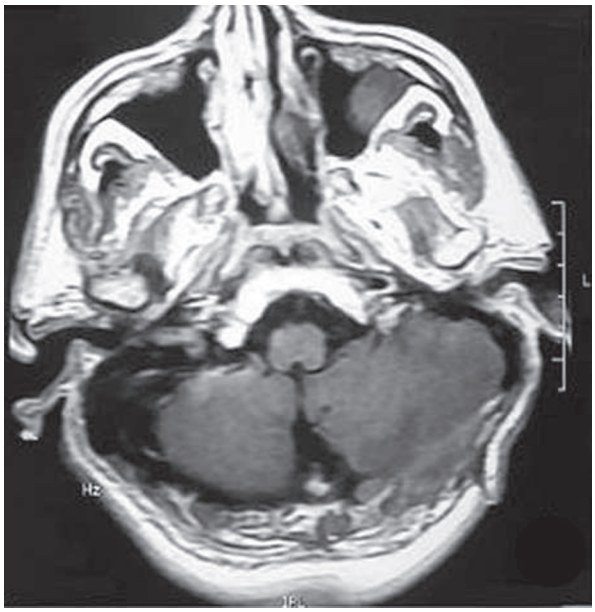

Fig. 2. Postoperative axial T1-weighted magnetic resonance scan reveals total resection of lesion

headache, nausea, and dizziness for the last 2 years and the symptoms had worsened for a month. The patient had palpated the occipital mass for only 2 weeks before he attended our clinic. Physical examination revealed a 4-cm large, soft and painless mass in the left occipital scalp, which had expanded towards the midline over the occipito-cervical incision scar. Past medical history included surgery for a mass lesion of the posterior fossa performed 18 years ago in another medical institution. His complaints before the first surgery included dizziness and severe headache. After the operation, the patient was informed that the lesion was a benign cerebellar tumour. Unfortunately, neither medical records, nor a comprehensive pathology report related to the previous operation were available.

The skull X-ray revealed a 3 × 3 cm occipital craniectomy defect in the cranium. Computed tomography (CT) confirmed the bony defect and showed a hypodense mass lesion extending from the scalp through the cranium and several centimetres into the posterior fossa. Magnetic resonance imaging (MRI) confirmed the presence of a mass lesion (3 × 3 × 4 cm), hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 1). Cranial MRI also showed compression of the left cerebellar hemisphere and fourth ventricle.

The patient underwent immediate operation. During surgery, we observed that the mass had extended beyond the previous craniectomy area and also caused lysis of the bone edges. We observed that the mass originated from the diploe as well as the thinned dura; the left cerebellar hemisphere was compressed. There was no extension to the intradural space. The mass, which was 3 × 3 × 4 cm in size, was totally resected with its capsule (Fig. 2). The pathological evaluations revealed keratinized, multilayered, and thin epithelial cysts. These cysts can be identified as epidermoid cysts without malignant transformation (Fig. 3).

The postoperative course was uneventful and the patient was discharged without neurological deficit. One year later there was no evidence of tumour recurrence.

Discussion

Epidermoid cysts of the skull are rare, benign tumours, accounting for less than 1% of all intracranial tumours [2]. These lesions have been classified as intradural and extradural. Extradural epidermoids account for only 25% of all intracranial epidermoids and are present in the scalp, calvaria, or cranial base [6]. Epidermoid cysts can occur at any age from the first to seventh decade of life [4]. To our knowledge, epidermoid cysts result from a primary failure of neurulation at a later stage in embryogenesis. Similarly, intradiploic

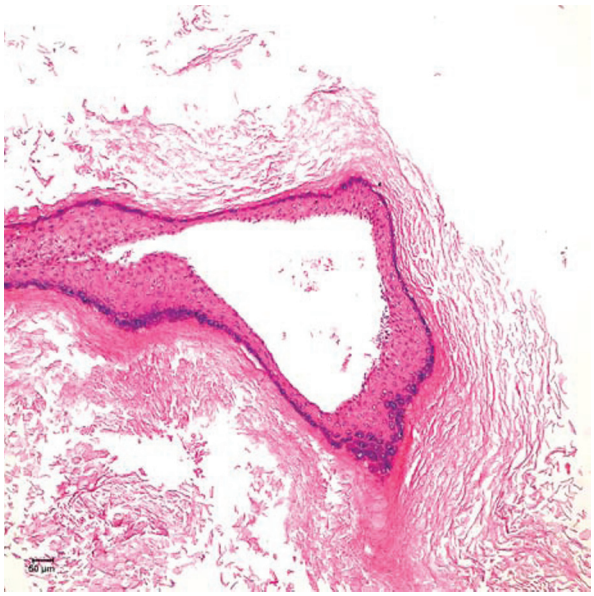

Fig. 3. Photograph showing stratified squamous keratinized epithelium typical of a benign epidermoid tumour (haematoxylin and eosin staining)

cysts are derived from ectodermal remnants that stay within the cranial bones during embryonic development [4]. Because of the lack of data related to the previous surgery in our patient, we cannot establish whether the lesion was derived from a developmental anomaly or acquired.

Iatrogenic epidermoid tumours are rarely seen, and although seeding of epidermal cells has been classically described only after lumbar puncture, the same mechanism could be involved in reported cases occurring after head injury [7] or cranial surgery with use of synthetic material for cranioplasty [8]. In our patient, seeding of epidermal cells may have occurred during the first surgery. Furthermore, it is well known that the capsule of an epidermoid tumour is the living portion of the tumour and any portion remaining after surgical resection is likely to regrow [9]. The recurrence rate of epidermoid tumours ranges between 8.3 and 25.0% [10]. In our case, it is possible that the patient underwent subtotal removal of an epidermoid tumour during his previous surgery. After surgery, local tumour recurrence could occur over a long period.

Epidermoid cysts are slowly growing lesions. Thus, these cysts may cause mild neurological deficits after reaching an enormous size [4]. Signs and symptoms depend on tumour location [1]. They spread in the subarachnoid space and have a tendency to envelop normal structures such as the brainstem, cranial nerves, and blood vessels [3]. The patient described here had been operated on for a mass lesion of the posterior fossa 18 years ago and had neither symptoms nor signs after the surgery. The findings in our patient are compatible with the literature because he did not have pathological symptoms for a long time.

On X-ray, the lesion is well-defined, radiolucent, and centred in the diploic space with sharp sclerotic borders [8]. CT scans reveal a homogeneous area of low attenuation values due to the presence of cholesterol [6]. There is no contrast enhancement, and the cyst interrupts the cranial tables [11]. 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. The signal intensity on T1-weighted images depends on the contents of the cyst [12]. The radiological features of our patient were compatible with an epidermoid cyst.

The differential diagnoses include dermoid cyst, osteolytic metastasis, intradiploic arachnoidal cyst, and, occasionally, hydatid cyst. Dermoid cysts usually occur in relation to suture lines and the midline [13,14]. Compared with epidermoids, dermoid cysts are inhomogeneous on CT scans [11]. Osteolytic metastases have irregular lytic areas destroying both tables of the cranium [14]. Hydatid cysts lack bone erosion and calcifications that are seen in epidermoids [15]. Diffusion-weighted MR imaging can be used to differentiate arachnoid cysts from epidermoid cysts [16]. Epidermoid cysts appear as high signal intensity on diffusion-weighted MR images [17], whereas arachnoid cysts have low signal intensity.

The aim of surgery should be the total resection of the tumour with its capsule. The long-term prognosis is excellent after successful resection [5]. Cranioplasty is recommended after resection to prevent further injuries to the brain after resection [18]. Seizures, infections, cranial nerve deficits, chemical meningitis and also death, when malignant transformation occurs, can be seen as complications [2]. Malignant transformation in these tumours is rare but squamous cell carcinoma could arise. When malignant transformation occurs, treatment options may include chemotherapy or radiotherapy [10].

In conclusion, we have reported the case of a rare huge intradiploic epidermoid cyst. Intradiploic epidermoid cysts should be included in the differential diagnosis of a patient who presents with a slowly progressive scalp mass. The prognosis is good when the cyst is totally resected with its capsule.

Disclosure

Authors report no conflict of interest.

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