

Epidermoid cyst of the brain stem. Case report

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Abstract

Brain stem epidermoid cysts are extremely rare lesions. Only nine cases have been reported. Management of the epidermoid cyst is decompression of cyst contents and removal of cyst capsule. But in some cases resection of the cyst may result in a poor outcome because of cyst wall adhesion into the brain stem.

Keywords: Brain stem, epidermoid cyst, magnetic resonance imaging.

1 Introduction

Epidermoid cysts were first described by the CUVÉLIER in 1829. BAILEY gave a detailed histological description in 1920 [9].

Epidermoid cysts account for 0.2% to 1.8% of all intracranial tumors [1, 2, 5, 6, 10]. They are most commonly located in the cerebellopontine angle and in the parasellar region [6, 9, 11, 16, 18]. Epidermoid cysts are filled with soft white flaky material containing a high concentration of cholesterol crystals.

Until 1992, only nine cases of epidermoid cysts involving the brain stem have been reported [3, 7–10, 12, 15]. Here a further case is reported and discussed.

2 Case report

This 2-year-old boy presented with a two month history of headache and bad temper. In neurological examination the patient had a right central facial palsy and right hemiparesis. Sagittal T2 weighted magnetic

resonance images showed a cyst located intraaxially within the pons (Figure 1). Axial T2 weighted images showed an extraaxial exophytic component projecting into left prepontine cisterna (Figure 2). There was no contrast enhancement or peritumoral edema on the T1 and T2 weighted images.

The patient underwent a left suboccipital craniectomy while in prone position. The extraaxial portion of the cyst and fifth cranial nerve was seen in the lateral pontine cisterna by extracting the left cerebellar hemisphere medially. The cyst was seen to be extended into the pons.

We removed the epithelial debris with suction. But did not make any attempt to remove the intraaxial portion of the cyst capsule which adhered to the pons. The pathological diagnosis was an epidermoid cyst (Figure 3).

The patient was discharged ten days after operation with no neurological deficits. Follow-up MRI images revealed residual tumor within the pons (Figure 4). The patient was still asymptomatic five months after surgery.

3 Discussion

Epidermoid cysts probably develop from inclusion of ectodermal elements at the time of closure of the neural groove between the 3rd and 5th weeks of embryonic life [1, 9]. Epidermoid cysts can occur at any age; approximately 50% of patients are between 20–40 years old. They occur with equal frequency in both sexes [18]. They may occur in multiple locati-

ons in the nervous system. Approximately half are intradural or intracranial. One fourth to one half of epidermoid cysts arise in the cerebellopontine angle and the posterior fossa cisterns [6, 18]. The suprasellar region is also commonly involved [6]. They are also found in the sylvian fissure and in the lateral or fourth ventricle (10% to 20%) [9, 18]. Epidermoid cysts of the brain stem are very rare with only nine cases previously reported [3, 7–10, 12, 15].

Epidermoid cysts appear as hypodense or isodense central areas surrounded by a capsule that cannot be enhanced on computed tomographic scans [4, 6, 14, 17].

On MRI, the signal intensity of an epidermoid cyst depends on the amount of lipid, cholesterol, and keratin within the cyst [1]. On T1 images, it usually exhibits an intermediate signal intensity between the signal intensities of the brain and the cerebrospinal fluid [13]. On T2 weighted images, signal intensities of epidermoid cysts are usually greater than that of either brain or CSF [13, 17]. Epidermoid cysts do not typically exhibit peritumoral edema [18].

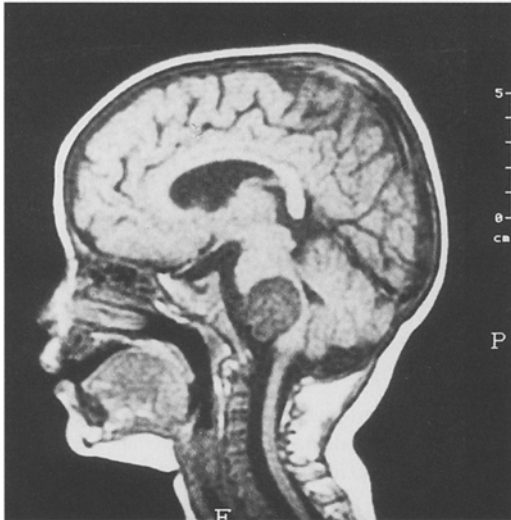


Figure 1. Sagittal T2 weighted magnetic resonance image showing a cyst located intraaxially within the pons.

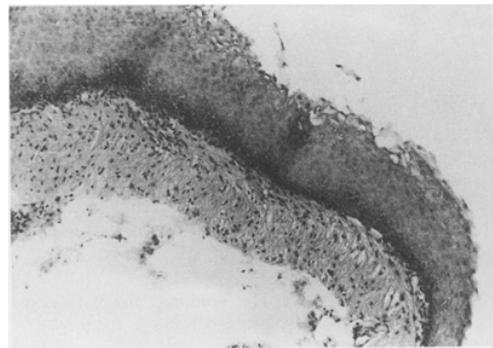


Figure 3. Pathology of the cyst wall revealed as an epidermoid cyst. Multilayered squamous cells directly invade upon nervous tissue (H.E. ×20).

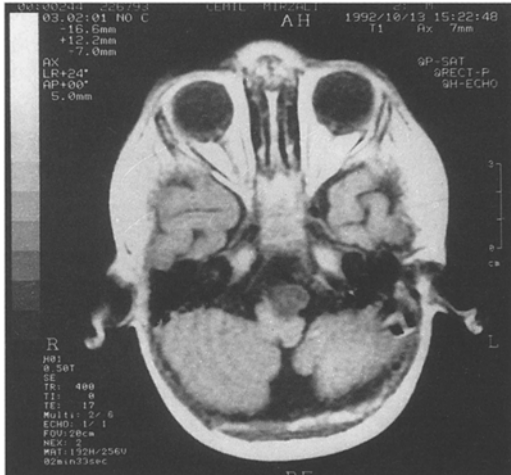


Figure 2. Exophytic component of the cyst projecting into the left prepontine cistern.



Figure 4. Follow-up MRI image showing residual tumor within brain stem.

Symptoms of patients with epidermoid cysts may result from involvement of important neural structures by the tumor or the exertion of gentle distortion and compression on the subarachnoid space. Clinical presentation of the epidermoid cysts is variable and may include: cranial nerve deficits, mental symptoms, cerebellar deficits, and pyramidal tract abnormalities [17]. Chemical meningitis may develop due to leakage of epidermoid cyst contents. The patient may have repeated attacks of meningitis, probably owing to spilling of cholesterol or fatty acids into the subarachnoid space.

There are various difficulties during the operation of epidermoid cysts. Although the walls of some cysts can be removed easily, the walls of others may adhere to arteries and important neural structures and so prevent total excision [17]. The extent of

tumor resection determines to a great extent morbidity and mortality [1, 9].

In our case the epidermoid cyst was located within the pons. The exophytic component of the cyst projected extraaxially into the left prepontine cistern and the cyst wall was adherent to the brain stem. During surgery decompression and removal of non-adherent small portions of the cyst capsule was performed.

If even a single epidermoid tumor cell remains after resection, cysts might recur. But a more aggressive resection might result in a poorer outcome.

We support previous observations a non-total resection of adherent brainstem epidermoid cysts, even though they may require re-operation, can reduce morbidity and mortality rates [9].

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