

Occipital Intradiploic Epidermoid Cyst with Intracranial Hypertension

Byung-Hyun Park, M.D., Hyun-Sung Lee, M.D., Jong-Soo Lee, M.D.

Department of Neurosurgery, Sun Hospital, Daejeon, Korea

Intradiploic epidermoid cysts of the cranium are rare, benign and slow growing lesion. However, these cysts may cause neurologic deficits due to mass effect. Intradiploic epidermoid cysts are thought to be derived from persistent ectodermal remnants present in the cranial bones during embryogenic development. Here, we report a case of an occipital intradiploic epidermoid cyst with posterior fossa extension. The patient developed intracranial hypertension associated with hydrocephalus due to aqueductal stenosis, and venous congestion secondary to compression of the torcular and the transverse sinus. The imaging studies included a computerized tomography(CT) venogram, which is essential for determining the pathophysiological mechanism of the clinical spectrum and for surgical planning. Near total removal was accomplished and the postoperative course was uneventful. Postoperative imaging studies demonstrated a reversal of the neural structures, but no patency of the sinus, which was presumably indicative of prolonged sinus stenosis. The patient gradually improved and was discharged free of symptoms.

KEY WORDS : Intradiploic epidermoid cyst · Intracranial hypertension · Hydrocephalus · Sinus stenosis.

Introduction

It is generally known that patients with intradural epidermoid cysts present with neurological symptoms. Patients with extradural epidermoid may be asymptomatic, while those with larger extradural intradiploic epidermoid cysts may present with neurologic signs secondary to the mass effect. We postulated that, in the present case, intracranial hypertension is not only caused by hydrocephalus but also partly by venous congestion.

Case Report

A 36-year-old man was treated for dizziness, headache and blurred vision at several outside clinics. One month before admission, he experienced visual dimming, a gradually intensifying morning headache, and an unsteady gait, for which he was referred to our neurosurgical department.

His past medical history was significant for a small lump on the back of his head, present since childhood. He had intermittently suffered from neck pain and headaches over a ten-

year period. A general physical examination revealed no abnormal findings, except for the subtle, non-tender lump on the back of the head.

On neurological examination, he had gait ataxia, a left lateral gaze nystagmus, and blurred vision. Ophthalmic examination revealed edematous optic discs with central venous congestion.

Plain skull radiography demonstrated an irregular, left suboccipital 7×7cm wide bony lytic defect with sclerotic margins (Fig. 1). Computed tomography(CT) sc-

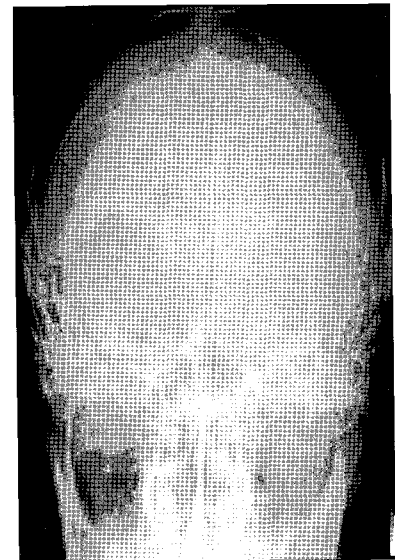


Fig. 1. Towne's view of the skull. The occipital bone showing lytic bony defect with sclerotic margin.

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• Address for reprints : Jong-Soo Lee, M.D., Department of Neurosurgery, Sun Hospital, 10-7 Mok-dong, Jung-gu, Daejeon 301-070, Korea
Tel : +82-42-220-8850, Fax : +82-42-222-7573, E-mail : jslee@sunhospital.com

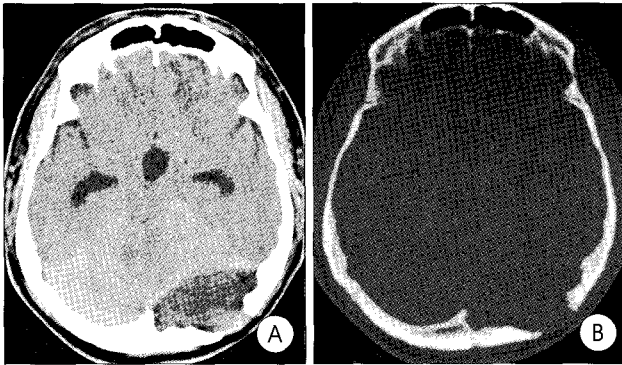


Fig. 2. Axial view of computed tomography showing hypodense lesion extended into the posterior fossa, which compressing the cerebellum and brainstem. Narrowing of the fourth ventricle with enlargement of the temporal horn and the third ventricle is observed (A). Corresponding view with bone window revealing the smooth bony erosion with almost thinning out of the inner table. The outer table is also thinned and partially defective (B).

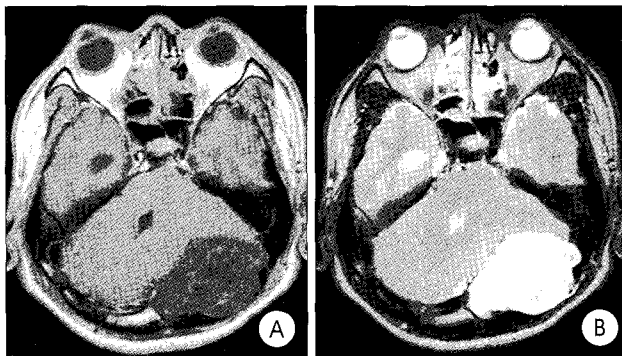


Fig. 3. The mass lesion appears inhomogeneous hypointense on axial T1-weighted image (A) and hyperintense on T2-weighted image (B).



Fig. 4. Venous phase of the conventional angiography with digital subtraction. Non-visualization of the left transverse sinus with resulting venous stasis and many collaterals draining into the left sigmoid sinus are seen on the left internal carotid artery injection.

ing of hypodense material that did not enhance with contrast. There was a smooth, marginal bony erosion with the inner tables almost thinned out and the outer tables sustaining the contour (Fig. 2). Magnetic resonance imaging (MRI) demonstrated a heterogeneous, low-signal intensity on the T1-

weighted images and a markedly increased, heterogeneous signal intensity on the T2-weighted images (Fig. 3). Conventional angiography demonstrated compression of the torcula by an avascular mass and poor opacification of the left transverse sinus (Fig. 4). CT venography with three-dimensional reconstruction revealed compression and displacement of the torcular and a quasi-patent left transverse sinus (Fig. 5).

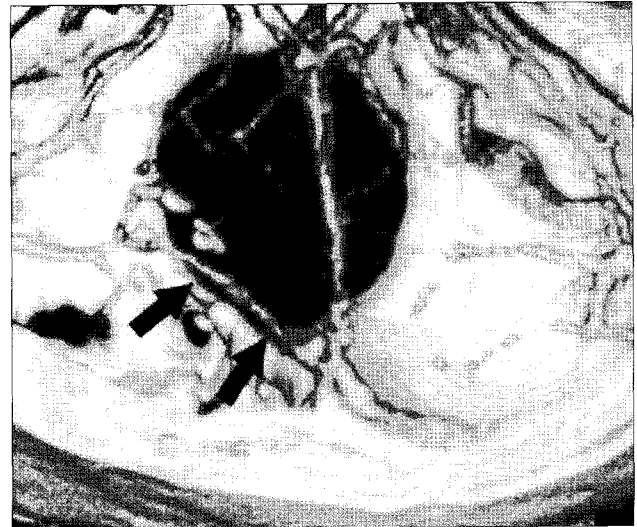


Fig. 5. Superior view of the three-dimensional computed tomographic venogram. The left transverse sinus and part of the torcular is displaced anteriorly and compressed remarkably (black arrows).

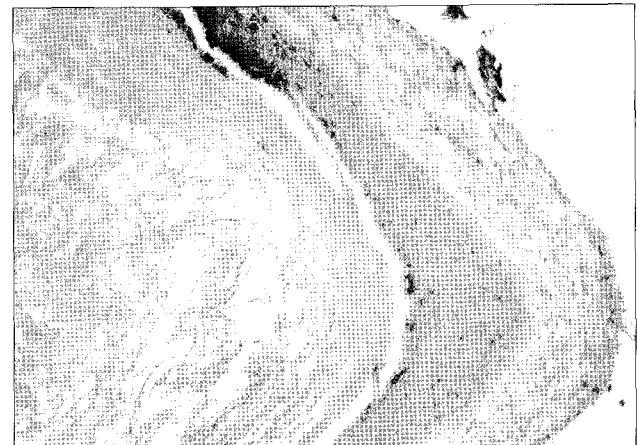


Fig. 6. The wall of the cyst is composed of mature squamous, granular, and horn cells of true epidermis. The cyst is filled with horny material arranged in laminated layers.

Total excision was attempted as possible with a left suboccipital craniotomy. There was a well-encapsulated cystic membrane, and small defects of the thinned outer table were noted. The cyst, which was filled with soft, waxy materials, was near totally evacuated. The inner membrane of the cyst and the underlying dura were cautiously separated with an operative microscope. The dural sinus and the dura mater were inspected and showed no evidence of cerebrospinal fluid leakage. However, we were unable to restore sinus patency, perhaps sec-

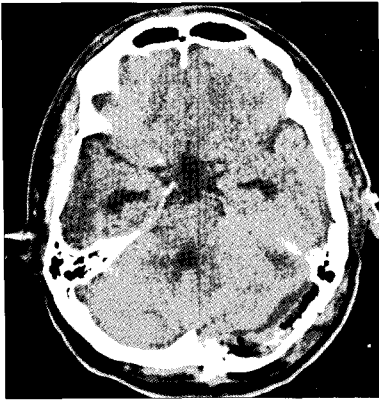


Fig. 7. Postoperative axial computed tomography showing the removal of the mass with restoration of the previously displaced neural structure and with reduction in size of the temporal horns.

secondary to prolonged compression. The procedure was uneventful, and there were no postoperative complications. Histologically, the lesions were composed of a thin capsule of stratified, keratinized squamous epithelium (Fig. 6). The patient improved progressively and was able to resume normal activities. He was discharged

after total removal of the tumor was confirmed by imaging studies (Fig. 7). However, patency of the sinus did not improve radiologically despite restoration to its normal position. The papilledema resolved eight months postoperatively. We do not, however, have any decisive evidence that the intracranial pressure changes were directly related to the venous sinus.

Discussion

Epidermoid cysts are uncommon and comprise about 1% of cranial tumors. Epidermoid cysts are believed to be derived from the ectopic inclusion of epithelial cells during the third to fifth week of embryonic development. About 75% are located intradurally, and 25% are located within the diploic spaces^{1,2,14}. Intradiploic epidermoid cysts are thought to result from the entrapment of the epithelial cells in the mesoderm. Usually, these lesions arise secondary to trauma⁴. This mechanism is more common in the lumbar spine⁹.

Intradural epidermoid cysts mainly affect the cranial nerves and adjacent structures, whereas extradural intradiploic epidermoid cysts cause only local effects¹¹. Enlargement of the intradiploic epidermoid cysts occurs not by active cell division but by desquamation of the normal cells into the cystic cavity. This explains the slow growth and benign nature of the lesion and being evident clinically in adult life¹⁴. Rarely, large lesions can cause neurologic impairment by the mass effect¹².

Histologically, epidermoid cysts are composed of a thin capsule of stratified, keratinized squamous epithelium. The cysts grow through accumulation of desquamated epithelial cells and their breakdown products, keratin and cholesterol^{13,17}. On plain radiography, intradiploic epidermoid cysts demonstrate characteristic, lytic bony erosions with scalloped margins and sharply delineated, sclerotic edges. The outer table is thinned while the inner table is usually destroyed. CT imaging cons-

istently shows epidermoid cysts to be homogeneous, hypodense, and lacking contrast enhancement. Careful evaluation of Hounsfield units may show the lesion to have a higher fat density than cerebrospinal fluid (CSF). Epidermoid cysts on MRI characteristically demonstrate a heterogeneous, low-signal intensity on T1-images and a markedly increased heterogeneous signal intensity on T2-images⁶. The typical appearance is due to the different proportions of fat and aqueous tissue in the cyst⁸.

Although most intradiploic epidermoid cysts are small and asymptomatic, the rationale for surgical intervention is to improve cosmetic problems, relieve neurologic deterioration, and avoid potential malignant degeneration^{2,3,10,20}. Recurrence of the intradiploic epidermoid has been reported¹⁹. Complete removal of the cyst is essential for the long-term prognosis^{1,2,7}. In the present case, the intracranial hypertension, which manifested symptomatically as headaches and the papilledema, was thought to be caused mainly by obstructive hydrocephalus. The postoperative resolution of the hydrocephalus was followed by symptomatic relief.

We also postulated that the sinus and torcular compression caused intracranial hypertension. A previously reported case involved a patient with an intradiploic epidermoid cyst and pure venous congestion without hydrocephalus. In this study, the authors noted a correlation between clinical improvement and pressure change¹¹. Here, we presumed that the sinus stenosis was associated with the intracranial hypertension, based on angiographic findings demonstrating intravenous congestion and dural sinus stenosis^{5,15,16,18}. However, we could not prove this mechanism, because the case was complicated by multiple variables (hydrocephalus and/or venous congestion), and we were unable to obtain evidence of the pressure change related to venous congestion. Although the venous obstruction is postulated as a potential cause of the intracranial hypertension in many clinical situations such as achondroplasia, Crouzon's syndrome and pseudotumor cerebri¹¹, this particular patient was thought to have adapted to the sinus stenosis by developing collateral circulation and sustaining patency of the dominant right transverse sinus. Additionally, the patient experienced symptomatic relief after resolution of the hydrocephalus, without any patency change of the sinus. We concluded that the compression of the torcular and transverse sinus might, but not conclusively, be associated with developing intracranial hypertension.

Conclusion

We report a rare case of an intradiploic epidermoid cyst of the occipital bone with extension into the posterior fossa, resulting in the development of intracranial hypertension. Pathogenetic mechanism of intracranial hypertension is thought

to be development of hydrocephalus and sinus stenosis. Decompression with gross total excision of the cyst resulted in an excellent clinical outcome.

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