Intradiploic Leptomeningeal Cyst of the Posterior Fossa —A Case Report—

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Summary

A case of an intradiploic leptomeningeal cyst of the posterior fossa in a 7-year-old boy is reported. Plain skull films demonstrated characteristic findings. The cause of the lesion was attributed to a head injury at 2 years of age. The pathogenesis and pathomechanism are discussed in contrast with supratentorial leptomeningeal cysts.

Key words: Leptomeningeal cyst, posterior fossa, growing skull fracture

Introduction

It is well known that head injuries with a linear skull fracture in infancy sometimes cause a growing skull fracture with or without a leptomeningeal cyst. This condition, however, is mainly located in the supratentorial portion of the cranium.

Leptomeningeal cysts in the posterior fossa are rare, and to our knowledge, there have been only four cases reported in the literature. The leptomeningeal cyst of the posterior fossa develops as an intradiploic cyst and never destroys the outer table of the skull. In this paper, a case of an intradiploic leptomeningeal cyst of the posterior fossa is presented.

Case Report

A 7-year-old boy was admitted to our hospital on March 26, 1979, complaining of a mass formation in the occiput. His birth and development had been uneventful. At 2 years of age he had fallen from a height two meters on to a concrete floor, and had sustained a cerebral concussion and a linear skull fracture in the occipital bone. He had been hospitalized for 2 weeks. After the accident he had been in good health until January, 1979, when he noticed an occipital mass.

The physical condition on admission was quite normal except for a hard palpable mass in the occiput. Neurological findings were unremarkable.

Plain films of the skull demonstrated a cystic lesion in the occipital bone, 6 x 6 x 2 cm. in size, hemmed with a osteosclerotic margin which extended from the external occipital protuberance nearly to the foramen magnum (Fig. 1). The inner wall of the bone cyst drew almost the same curve as the inner table of the skull on the lateral projection, but the thin outer wall was markedly protruded outward. The inner and outer walls of the cyst continued to the inner and outer tables at the upper margin of the cyst (Fig. 1A). Towne's projection showed an oval osteosclerotic margin encircled by a large round calcified margin which indicated the boundary of the cystic mass lesion (Fig. 1B). These findings indicated that there must be a large intraosseous cyst such as a cystic bone tumor, an epidermoid cyst or an intradiploic leptomeningeal cyst.

In an operation on March 27, 1979, a midline straight scalp incision was made to expose the suboccipital mass. The thin external table of the occipital bone was present all around the bulging. Immediately after a small perforation was made in the outer wall of the cyst, a moderate amount of clear colorless CSF gushed out abruptly through the opening. The thickness of the outer wall of the cyst was less than 1 mm. Removal of the outer wall unroofed the cystic cavity. The cyst was 22 mm. deep at the center. Fine bony spicules were connected between the inner and outer walls. There was a hole, 2.5 x 3 cm. in size, approximately at
the center of the cavity, through which normal cerebellar tonsils and the medulla oblongata could be seen (Fig. 2). The hole corresponded to the smaller oval-shaped osteosclerotic margin encircled by a large round margin that had been observed radiographically. The margin of the hole was irregular with many calcified and non-calcified spicules just like stalactites. The arachnoid membrane and degenerated dura mater were identified along the margin of the hole. The calcified inner wall was removed carefully and the adhesion of the dura to the wall was separated to expose the undegenerated dura. Both posterior inferior cerebellar arteries were identified at the normal sites. A galeal flap of 3.5 x 4 cm. was applied to the dural defect for watertight dural plasty. Cranio-plasty with methylmethacrylate resin was performed in the upper third of the bony defect. The postoperative course was uneventful and at present 1 year after the operation, he attends elementary school in good health.

Discussion

The characteristic findings in this case were formation of the intradiploic cyst filled with cerebrospinal fluid in the occipital bone of the posterior fossa and free communication of CSF between the cystic cavity and the cisterna magna via a defect through the arachnoid, dura and inner table. The outer table of the skull and the outer wall of the cyst were very thin but not destroyed (Fig. 3).

Although there was apparently no arachnoid membrane encapsulating the cyst, the diagnosis of leptomeningeal cyst might be accepted according to the definition of the Head Injury
Glossary,\textsuperscript{2} in which a leptomeningeal cyst is defined as a persistent cystic accumulation of cerebrospinal fluid with progressive loss of bone and dura occurring at the site of a previous fracture.

Concerning the pathomechanism of this condition, we agree with Taveras and Ransohoff,\textsuperscript{7} who offered a possible explanation of supratentorial leptomeningeal cysts. In these supratentorial lesions the whole thickness of the skull, both the inner and outer tables, is eroded, and a pulsating or nonpulsating cystic mass not covered by bone is palpable under the scalp.

In our case a linear skull fracture of the occipital bone with a dural tear occurred in a head injury at 2 years of age. The arachnoid membrane herniated into the diploic tissue simultaneously through the dural tear and the fractured inner table. The herniated arachnoid sac might extend into the diploic tissue with pulsating pressure, eroding the inner table and diploic tissue. Therefore, the cyst might become larger in the diploe by the water hammer effect.

In case of leptomeningeal cysts in the posterior fossa, the outer table is usually not destroyed and the cyst develops into the intradiploic tissue. Dunkser and McCreary\textsuperscript{3} postulated that the much thicker occipital bone might permit the arachnoid to herniate only through the inner table, but not through the entire thickness of the skull. Hillman et al.\textsuperscript{5} pointed out that the thick musculature behind the inferior portion of the occipital bone might cushion the force of the trauma so that only the inner table might be fractured and the arachnoid sac might be trapped between the inner and outer table, resulting in the formation of the intradiploic cyst. Another feature of the posterior fossa is the presence of a large subarachnoid space, the cisterna magna, between the skull and brain. This might reduce the expansion of the pulsating pressure effect in the bony tissue in contrast to the supratentorial lesion which is compressed almost directly by the pulsating brain.

Although the outer table is not destroyed, we believe that this condition also could be called a growing or enlarging skull fracture, because the pathomechanism of this condition might be essentially the same as that of the supratentorial leptomeningeal cyst and only be modified by the anatomical characteristics of the posterior fossa.

Four cases of leptomeningeal cyst in the posterior fossa have been reported in the literature (Table 1).\textsuperscript{1,3,5} Cerebellar symptoms and signs were noted in two cases due to inward compression by the bulging cyst wall. Our case is similar to the second case of Hillman in that no cerebellar sign was present.

Differential diagnosis of an intradiploic cyst should include an epidermoid cyst,
aneurysmal bone cyst and a cystic form of fibrous dysplasia, as well as a leptomeningeal cyst. The diagnosis of an intradiploic leptomeningeal cyst might not be so difficult if the preceding fracture of the posterior fossa in infancy is obvious. Useful roentgenographic findings are a partial defect of the inner table in plain sagittal tomography and inflow of radioactive substance into the cyst in cisternography. The CT scan is certainly useful although no reports with CT scans have been seen yet. Dural plasty is essential for treatment of this condition as is so for treatment of supratentorial growing skull fracture.

References