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Traumatic porencephalic cyst and cholesteatoma of the ear

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Abstract
Porencephalic cyst expanding into the ear is a very rare complication of temporal bone fracture. We report a case of a 20-year-old male who developed a traumatic porencephalic cyst of the temporal lobe expanding into the ear through a tegmen fracture in association with a cholesteatoma. The clinical presentation was otitic meningitis.

This occurrence was not encountered in any of the cases reviewed in the literature. The diagnosis, pathogenesis, treatment and prognosis of this condition are reviewed.

Key words: Cysts; Brain; Cholesteatoma; Ear Trauma

Introduction

The definition of porencephaly most commonly used is that of LeCount and Semerak,[1] who described it as a 'defect communicating with ventricles or separated from them by a thin layer of brain tissue, and covered on the outside by the arachnoid'. The cavity is filled with cerebrospinal fluid.

Porencephaly was initially described by Heschl[2] as a congenital anomaly caused by anoxia, disturbances of nutrition in the mother, or defects of foetal development.[1] Currently, with the advent of tomography and other advanced radiological techniques, acquired cases of porencephaly are now considered more prevalent than congenital ones.

Although this condition is rare, a porencephalic cyst most often appears after cranial trauma in children, and the condition has been termed 'growing fracture of the skull'.[3–5]

A computed tomography (CT) scan is often the only neuroradiological procedure needed to make the diagnosis. On CT, the area of the porencephalic cyst is well-defined and its contents have the density of cerebrospinal fluid. This density does not change after the use of contrast enhancement material, indicating that the cyst is an avascular space-occupying lesion. The absence of a vascular capsule surrounding the porencephalic cyst distinguishes porencephaly from primary and secondary malignancies or abscesses, but, porencephaly must not be confused with low-density neoplasms.[6]

Porencephalic cysts are generally not reversible and do not resolve spontaneously. Relieving the intercavity pressure may not significantly reduce the size of the cyst, but will help prevent the cyst from expanding into surrounding areas, which could cause headaches and other neurological symptoms.[7] In such symptomatic cases, the pressure may be relieved by cystoperitoneal or ventriculoperitoneal shunts, or by opening the cyst to the subarachnoid space.[8]

The prognosis of porencephaly is independent of the cause and depends on the location and extent of the lesion. If sufficiently large, the cyst can cause compression or pressure on adjacent structures.

Case report

A 20-year-old Caucasian male presented with fever, malaise and headaches. Neurological examination revealed meningeal signs and absence of neurological deficits. There was a history of two severe cranio-encephalic traumata (CET), at the ages of three and 14, both resulting from car accidents, the latter one leading to 25 days period of coma and CSF leak with spontaneous resolution.

A CT scan was performed, and he was admitted to neurosurgery with the diagnosis of meningitis secondary to right middle-ear infection.

On the same CT scan, besides the infectious process of the middle ear, a porencephalic cyst was seen, communicating with the moderately enlarged right lateral ventricule, and passing through an extensive tegmen tympani and tegmen antri defect. The right middle and external ear

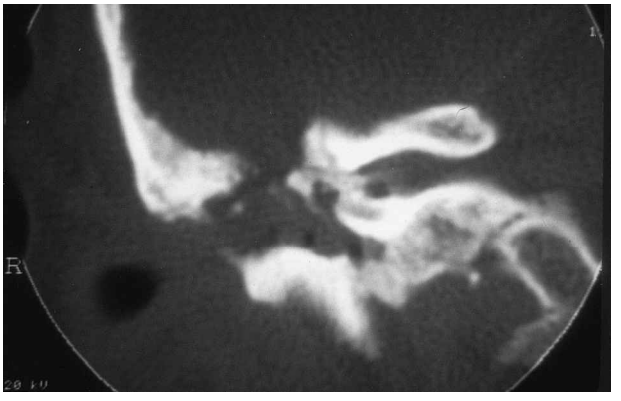

Fig. 1
CT scan showing tegmen defect and middle and external ear filled with soft tissue.

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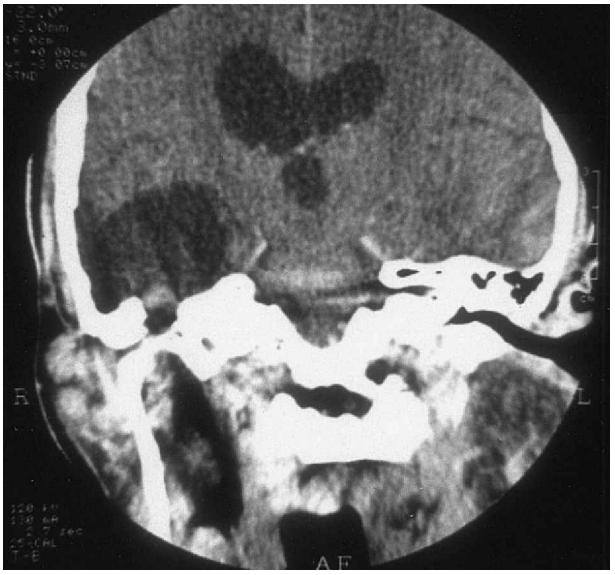

FIG. 2

CT scan showing 'granulomatous' lesion adjacent to the porencephalic cyst.

were totally filled with soft tissue (Figure 1). A small granulomatous lesion, was present, under the right temporal lobe, near the antero-superior vault of the petrous bone, and adjacent to the wall of the cyst (Figure 2).

Treatment with antibiotics was initiated with good resolution of the infectious process.

An ENT consultation was requested. Purulent right otorrhoea was observed, as well as a marked stenosis of the external ear canal, obscuring the tympanic membrane. An audiometric study was performed, disclosing a conductive hearing loss, with air-bone gap of 20 dB.

After the resolution of the acute infectious process, a magnetic resonance imaging (MRI) was requested for further study. Although it provided some better images (Figure 3), MRI did not definitely clarify some of the doubts of CT, namely the possible nature of the granulomatous lesion previously observed (Figure 4).

A radical mastoidectomy was performed about two months after the diagnosis of meningitis. During surgery, an extense cholesteatoma located in the epitympanic region, and extending to the middle fossa was observed.

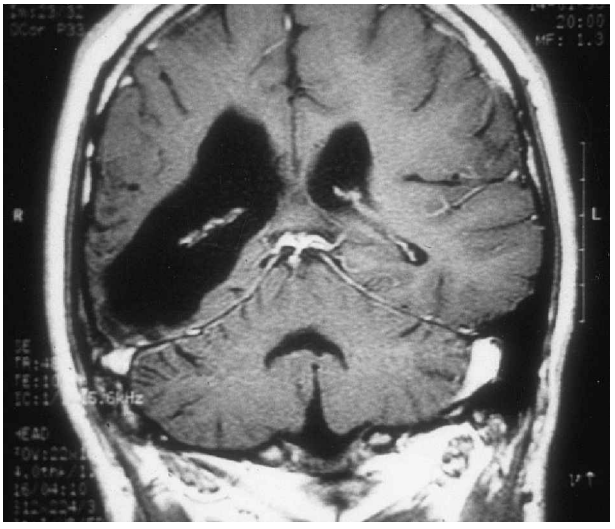

FIG. 3

MRI image – right porencephalic cyst.

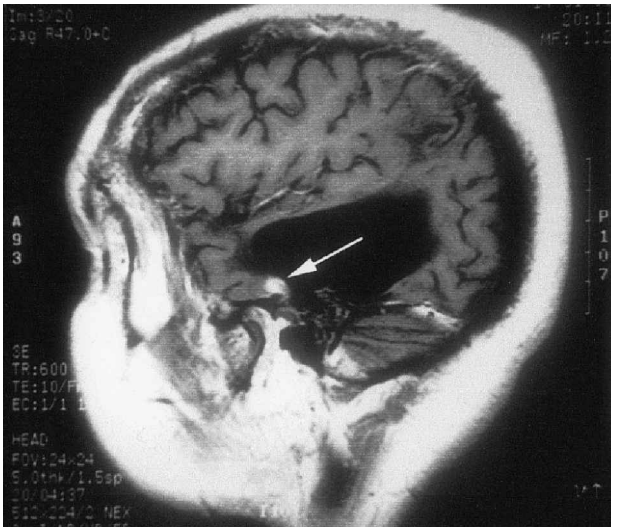

FIG. 4

MRI image showing 'granulomatous' lesion (white arrow) adjacent to the porencephalic cyst.

It was adjacent to the cyst formation, which passed into the middle ear through a large tegmen defect (Figure 5). The ossicular chain was antero-inferiorly dislocated. In the act of cholesteatoma removal, an accidental tear of the hernial sac occurred with consequent CSF leakage. The sac was repositioned in the middle fossa and the osseous defect repaired with fascia temporalis. The mastoid was obliterated with temporalis muscle, with transient resolution of the CSF leak. Nevertheless, in the first week after surgery CSF leak recurred and a second operation was programmed, this time using a middle fossa approach, by the neurosurgeons.

Twenty months after surgery the patient is well, with no CSF leak and no signs of residual or recurrent cholesteatoma.

Discussion

Porencephaly is now defined as either a congenital or acquired local defect of the cerebral hemisphere communicating with the ventricular system. Congenitally, it can be caused by anoxia, disturbances of nutrition in the mother, or defects of foetal development. Acquired porencephaly results from infarction, haemorrhage, a focal inflammatory lesion, repeated episodes of localized arterial spasm,

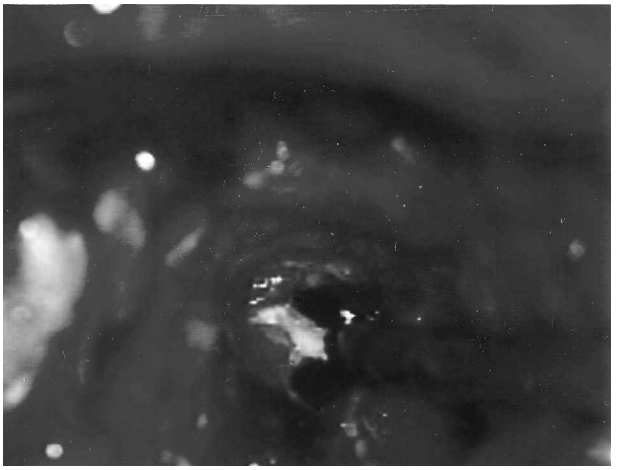

FIG. 5

Surgery: cholesteatoma and cyst.

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trauma or post-surgical changes that result in destruction of cerebral tissue.[9] Jaffe[10] particularly emphasized the importance of trauma in porencephaly and many other authors have described several cases in which trauma was determined to be the cause.[11]

The associated hypoperfusion results in destruction of grey and white matter that may then induce ischaemic encephalomalacia and focal necrosis. These processes are followed by cystic degeneration, which is the precursor of the porencephalic cyst. Enlargement of the cyst is thought to occur from mechanical forces, such as pulsations of the brain and the flow of cerebrospinal fluid into the cyst, where a membrane or other obstruction at the point of communication with either the ventricle or the subarachnoid space allows cerebrospinal fluid to flow into the cyst, but blocks flow out as in a ball-valve mechanism. These forces may result in increased pressure inside the cavity.[12]

Another proposed mechanism for the formation of a porencephalic cyst is elevated intracranial pressure, which may cause defects in the ventricular wall. Cerebrospinal fluid then enters these defects and splits the cerebral parenchyma, ultimately forming a porencephalic cyst.[13]

The case presented here certainly illustrates the importance of trauma, even later in life, in the development of porencephalic cysts. The following factors are characteristic in the development of the traumatic porencephalic cyst of the ear: temporal bone fracture through the tegmen tympani or tegmen antri, or both, dural tear, manifested by a cerebrospinal fluid leak with brain being found in the ear during the period of fresh trauma and severe trauma to the brain; the pia and arachnoid have expanded into the fracture line, slowly enlarging the bony defect and protruding into the attic and tympanic cavity; the brain above the defect dissolves, leading to a connection between the temporal horn of the lateral ventricle and the arachnoid cyst. The cyst may expand into the external ear canal.[14]

The diagnosis is made by CT scan or MRI, demonstrating a cyst communicating with the middle-ear space and the lateral ventricle. Histological studies are not necessary for the diagnosis of porencephaly.[11]

The other striking feature of this case was the existence of a cholesteatoma in the middle ear. Cholesteatoma may be congenital or acquired. The pathogenesis of acquired cholesteatoma is probably multifactorial but eustachian tube malfunction and infection play an important role.[15] Cholesteatoma in children shows a more extensive and rapid growth in the middle ear and mastoid than its counterpart in adults.[16]

Some features in this particular case may raise questions concerning the true aetiology and pathogenesis of this cholesteatoma, although a process of epitympanic blockage by the porencephalic cyst has been the mechanism which probably leads to its development. The existence of a previous true congenital cholesteatoma cannot be totally excluded, nor can the possibility of two completely independent processes. Nevertheless, it is reasonable to assume a relationship between the two co-existent diseases.

Treatment of porencephalic cysts of the ear has been by mastoidectomy, covering the defect with fascia, and obliteration of the mastoid cavity with the sternocleidomastoid muscle,[11] or with a combined mastoid and middle fossa approach.[17] In our case, we tried first to do a mastoidectomy with obliteration. For the purpose of a definitive resolution there was, nevertheless, the need of neurosurgical help, with a middle fossa approach. This illustrates clearly, that an ENT-neurosurgical collaboration is important, in order to get the best results.

Conclusion

As far as we know, a simultaneous traumatic porencephalic cyst and cholesteatoma of the same ear has not been reported before.

The otologist should be aware of the possibility of porencephaly when he treats a patient who has otitic meningitis or cerebrospinal otorrhoea, particularly when the patient has a history of basal skull fractures and massive cerebral injury.

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