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The pathophysiology of otitic hydrocephalus

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Abstract

The pathophysiology of otitic hydrocephalus remains controversial. It has been argued that involvement of the superior sagittal sinus, by, at least, a mural thrombus is a necessary component of this disease.

We present a case of otitic hydrocephalus where on magnetic resonance imaging (MRI) normal luminal and mural flow within the superior sagittal sinus is demonstrated. The presence of thrombus in the lateral venous sinus alone appears sufficient in this case to impede venous drainage of the intracranial contents into the neck and produce a rise in the cerebral venous pressure and a subsequent increase in the CSF pressure. The presence of a superior sagittal sinus mural thrombus is not required.

Key words: Hydrocephalus, otitic; Cerebrospinal fluid; Sinus thrombosis

Introduction

Quincke in 1896 first described a condition where the intracranial pressure was raised in the absence of focal neurological signs and the CSF was of normal composition. Since the initial description it has been described by several names (Foley, 1955) and varied aetiologies have been associated with its occurrence (Fishman, 1984). Names in common usage today include benign intracranial hypertension, pseudotumour cerebri and otitic hydrocephalus. The latter was coined by Symonds (1931), and although it is an unfortunate misnomer (Doyle *et al.*, 1994), it now seems to be the accepted term in the surgical literature when the condition is associated with middle ear or mastoid disease.

The clinical features of otitic hydrocephalus are those common to all causes of raised intracranial pressure (namely nausea, headache, papilloedema, and possible VI cranial nerve palsy) but in this situation the CSF composition is normal, there is no evidence of ventricular dilatation and focal neurological signs are absent.

Controversy exists as to the exact pathophysiology of otitic hydrocephalus and, although thrombosis of the lateral venous sinus is almost universally accepted as a constant component of this disease, debate remains as to whether concomitant superior sagittal sinus thrombosis is essential to produce the clinical picture.

Case history

A seven-year-old male was admitted with a 10-day history of right otalgia and otorrhoea with associated lethargy. He was pyrexial (40 °C) and tender over his right mastoid process. A lumbar puncture showed normal CSF under normal pressure. A right cortical mastoidectomy was performed which revealed pus in both the middle ear and mastoid segments; however, the dural and sinus plates were intact. He made a rapid recovery and was discharged two days after operation.

The patient was re-admitted 10 days after his initial presentation with nausea, lethargy, headache, left VI nerve palsy and papilloedema. The right tympanic membrane was unremarkable.

MRI of the brain was performed together with 2D phase contrast MR venography (Figures 1 and 2). Normal blood flow was present in the left lateral venous sinus and superior sagittal sinus but was absent in the region of the right transverse and sigmoid sinus. Coronal images of the brain were obtained pre- and post-gadolinium and demonstrated marked enhancement in the region of the right lateral venous sinus, consistent with the presence of thrombus and probable perivenous inflammation. This thrombus extended up to, but did not include the torcula. Signal void in the region of the left lateral venous sinus

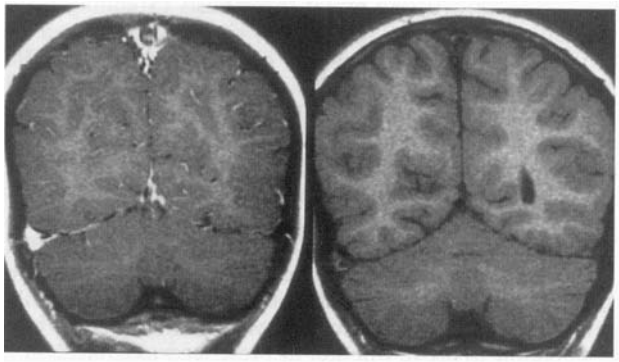

FIG. 1

T1 coronal images, pre- and post-gadolinium. Marked enhancement in the region of right lateral venous sinus following contrast is consistent with the presence of thrombus in this region. Normal blood flow in the superior sagittal sinus with peripheral signal void due to slow flowing blood adjacent to the sinus wall, and higher intensity signal centrally due to faster flowing blood in the middle of the sinus (a). The pre-contrast image is included for reference (b).

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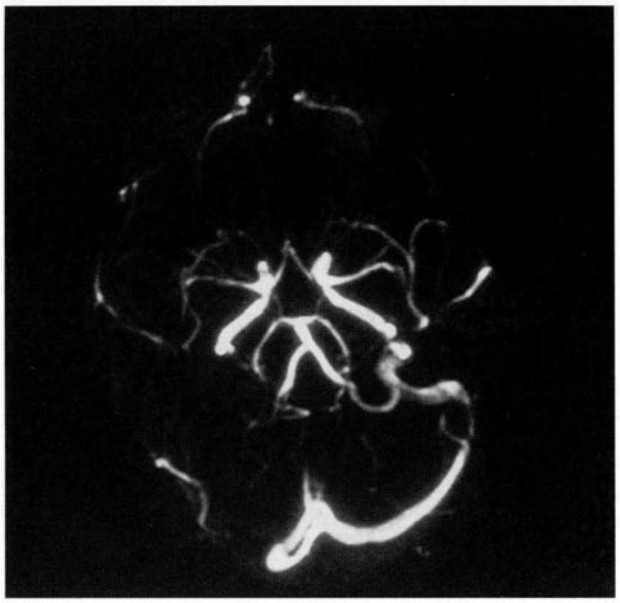

FIG. 2

Axial 2D phase contrast magnetic resonance venogram, demonstrating absence of blood flow in the region of the right transverse and sigmoid sinus.

confirmed its patency and a normal pattern of enhancement in the superior sagittal sinus also indicated normal blood flow.

The child was treated with high dose intravenous antibiotics and following a therapeutic lumbar puncture, during which a CSF pressure of 450 mm of water was recorded and 30 ml of normal CSF removed, the child made a rapid and complete recovery.

Discussion

The exact mechanism of otitic hydrocephalus still remains a source of debate. Symonds (1937) suggested that a non-obstructing mural thrombosis extending from the lateral sinus into the sagittal sinus was required to produce the features seen in otitic hydrocephalus. Its presence would directly interfere with the transfer of CSF to the venous sinus (Lund, 1978; O'Connor and Moffat, 1978). It is essential that only partial obstruction occurs, as sudden, complete obstruction of this sinus by thrombus would produce a very different clinical picture. In sharp contrast to lateral sinus thrombosis, sagittal sinus thrombosis is associated with significant mortality and morbidity.

Evidence of such a thrombus partially obstructing the sagittal sinus in a patient with otitic hydrocephalus has only recently been demonstrated (Doyle *et al.*, 1994) but it is not possible to prove whether its presence was coincidental or the reason for the development of otitic hydrocephalus.

An alternative mechanism proposes that the presence of thrombus in the lateral sinus leads to impeded venous drainage into the neck, especially if the thrombus occurs in a dominant lateral sinus. Extension to the superior sagittal sinus is not required (Clemis and Jerva, 1976; Powers *et al.*, 1986). An increase in the intracranial pressure may then be produced either by direct transmission of the raised venous pressure to the CSF or by impeding the function of the arachnoid villi. An increased pressure in the superior sagittal sinus leads to reduced CSF absorption by the arachnoid villi and consequently an increase in CSF pressure (Fishman, 1984; Powers *et al.*, 1986).

There is a wide anatomical variation in the venous drainage of the intracranial contents (Woodhall, 1936) and this may explain why not all episodes of lateral sinus thrombosis lead to otitic hydrocephalus. If a dominant venous sinus becomes obstructed, in the presence of inadequate cross communication at the torcula, venous drainage may be sufficiently impaired to cause raised intracranial pressure. Even if this cross communication is adequate, a thrombus extending to involve the torcula may impair its potential as a collateral. It follows that the extent of the thrombus within the lateral sinus may also be a significant factor in determining if otitic hydrocephalus develops.

In our case thrombus was demonstrated in the right lateral venous sinus (which is usually the dominant sinus) and extended up to but did not include the torcula (Figure 1). The post-gadolinium coronal brain MR images demonstrated a normal pattern of enhancement in the superior sagittal sinus with peripheral signal void due to slow flowing blood adjacent to the sinus wall, and higher intensity signal centrally due to faster flowing blood in the middle of the sinus (Figure 2). Mural thrombus was not present in this sinus.

Conventional X-ray angiography fails to opacify the superior sagittal sinus completely due to mixing of contrast free blood from the hemisphere contralateral to the side of contrast injection entering the sinus. Definite exclusion of the mural thrombus in this region is therefore difficult.

Until now proponents of Symonds' theory have argued that the imaging methods were not good enough to detect a partially obstructed superior sagittal sinus. The advent of the MRI has clearly changed this. The presence of a mural thrombus in the superior sagittal sinus is *not* a requirement of this condition. The degree of thrombosis together with sufficient asymmetry in the venous drainage system is enough to determine whether a lateral venous thrombosis leads to otitic hydrocephalus. If mural thrombosis in the superior sagittal sinus is found on MRI, its presence may either be taken as a coincidence or it may well be contributing directly to impairment of arachnoid villi function. Alternatively, extension of thrombus into this adjacent sinus may be the point at which compromise of the venous drainage becomes significant enough to cause otitic hydrocephalus.

Conclusions

Extension of thrombus from the lateral sinus into the superior sagittal sinus is unnecessary to produce otitic hydrocephalus. It appears that in the presence of an appropriate venous sinus drainage pattern lateral sinus thrombosis can lead to a rise in the cerebral venous pressure and a subsequent increase in the CSF pressure. We would concede, however, that in cases where a mural thrombus is identified in the superior sagittal sinus, both mechanisms could play a part in the development of otitic hydrocephalus.

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