Otitic hydrocephalus: case report and literature review

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Abstract

Otitic hydrocephalus is characterized by elevated intracranial pressure without focal neurologic abnormalities. The pathogenesis of otitic hydrocephalus remains unclear, but it usually occurs secondary to lateral sinus thrombosis. With the advent of new antibiotics, there has been a spectacular decrease in the complications of otitis media. Otogenic intracranial hypertension, always an uncommon condition, is seen only very rarely nowadays. Here we present a case of otitic hydrocephalus and discuss about its pathophysiology, clinical findings, and treatment.

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1. Introduction

Otitic hydrocephalus is one of the rare intracranial complications of middle ear infection, consisting of elevated intracranial pressure without focal neurologic abnormalities other than those due to the elevated pressure [1]. Over a 20-year period, Gower and McGuirt [2] noted only 5 patients with otitic hydrocephalus from 100 cases of intracranial complications of infectious ear disease.

Taylor was the first to describe patients with increased intracranial pressure with a normal composition of cerebrospinal fluid (CSF) in the absence of a tumor [3]. However, Quincke in 1897 was the first who recognized this condition and published a thorough report; in this study, Quincke termed it as serous meningitis. Throughout the history, many other terms have been used, including pseudotumor cerebri, toxic hydrocephalus, meningeal hypertension, and intracranial hypertension of unknown cause [3], but otitic hydrocephalus is the term suggested by Symonds [4], and although it is an unfortunate misnomer [5], it now seems to be the accepted term in the surgical literature when the increased intracranial pressure, in the absence of focal neurologic signs and normal CSF findings, is associated with middle ear or mastoid disease [6].

The leading symptoms of otitic hydrocephalus (headache, drowsiness, blurred vision, nausea and vomiting, and sometimes diplopia) are common to all causes of raised intracranial pressure [6]. The onset may be many weeks after an acute otitis media or many years after the start of chronic middle ear disease. Clinical examination reveals papilledema and usually lateral rectus palsy due to cranial nerve VI stretching on 1 or both sides. Optic atrophy can eventually develop [4].

Elevated CSF pressures with normal CSF biochemistry comprise the classic findings of otitic hydrocephalus. Because lumbar puncture in the presence of raised intracranial pressure associated with papilledema is dangerous because of the risk of cerebellar coining, it should be done with caution [4].

The pathogenesis of otitic hydrocephalus still remains doubtful. Although thrombosis of the lateral venous sinus is almost always a constant component of this disease, debate remains as to whether concomitant superior sagittal sinus thrombosis is essential to produce the clinical picture [6].

2. Case reports

The patient was a 28-year-old man with a history of blood-stained, foul-smelling, and bilateral otorrhea for many years who presented with a progressively worsening headache especially behind the right eye, low-grade fever, diplopia, and blurring of vision for 7 days for which he came to the hospital. On examination, bilateral cholesteatoma with granulations and purulent, thick, and foul-
smelling otorrhea was seen. Because of headache and right abducens nerve palsy in the presence of cholesteatoma, with the diagnosis of Gradenigo’s syndrome, a computerized tomography (CT) scan was performed. Computerized tomography examination did not give enough information on the temporal bone cellular system because of low resolution and thick sections, but erosion of dural and sinus plate especially in the left was evident. A right canal wall down tympanomastoidectomy was performed. Granulations and a polyp were removed from the middle ear and mastoid antrum. The lateral sinus plate and the dural plate were eroded, but there was no lateral sinus thrombosis or suppurative focus in the petrous apex.

His symptoms did not get better after the operation; fever and headache worsened, and an inflamed tender area was evident in the left side of the neck. Funduscopy showed the presence of established papilledema with hemorrhages and exudates on both sides. A contrast-enhanced CT scan of the temporal bone and brain showed the presence of “empty delta” sign in the region of left sigmoid sinus (Fig. 1), but the ventricular system was normal. Contrast-enhanced CT scan of the neck revealed a rounded hypodense area with rim enhancement medial to the left sternocleidomastoid muscle, consistent with the internal jugular vein thrombosis (Fig. 2). Pre- and postgadolinium magnetic resonance imaging (MRI) revealed marked enhancement in the region of the left sigmoid and lateral venous sinuses, consistent with the presence of thrombus and probable perisinus inflammation. This thrombus extended up to, but did not include, the confluens sinuum. Signal void in the region of the right lateral venous sinus confirmed its patency (Fig. 3). The lumbar puncture (LP) revealed an opening pressure greater than 250 mm H₂O, but microscopic findings were normal.

With the diagnosis of otitic hydrocephalus, topical antibiotic drops, intravenous antibiotics, high-dose steroid, acetazolamide, and mannitol were started. A left-side canal wall down tympanomastoidectomy was performed 7 days after the first operation, and all granulations were removed from the middle ear and mastoid antrum. There was a lateral sinus thrombosis with a perisinus abscess from which pus was drained and erosion of the dural plate. The sinus was opened and the infected clot was removed, but the internal jugular vein remained untouched.

The patient made a rapid and complete recovery and was discharged from the hospital within a few days. After
2 months, the abducens palsy recovered completely, and his visual acuity and funduscopic examination results were normal.

3. Discussion

Antibiotic use may have changed the incidence and morbidity of intracranial complications of otitis media. Because these complications have declined markedly since the advent of antibiotics, many contemporary otolaryngologists have been unexposed to these complications. Furthermore, patients’ symptoms are now often masked, so patients appear remarkably well despite the presence of potentially fatal complications [7].

Otitic hydrocephalus is a rare complication of otitis media and stems from either acute or chronic otitis media. Even though the intracranial complications of otitis media are far less common compared with the preantibiotic era, they do, however, still occur and may still be fatal [8].

In patients with otologic disease, complaints of headache, diplopia, or photophobia should warrant an evaluation. The presence of lateral sinus thrombosis mandates further investigation for additional intracranial complications [9].

The precise mechanism underlying the development of otitic hydrocephalus is unknown. Symonds (1937) suggested that a nonobstructing mural thrombosis extending from the lateral sinus into the sagittal sinus was required to produce the features seen in otitic hydrocephalus [6].

Because superior sagittal sinus thrombosis should be associated with more neurologic deficits than are found in otitic hydrocephalus, it is postulated that a mural nonobstructing thrombus extending to the superior sagittal sinus was required to produce the features seen in otitic hydrocephalus [6].

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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 [11]. 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 [12]. However, ligation of the internal jugular vein in the neck does not cause hydrocephalus; thus, that mechanism must be the suspect.

One explanation may be the wide anatomical variation in the venous drainage of the intracranial contents, and this may explain why not all episodes of lateral sinus thrombosis lead to otitic hydrocephalus. Clemis and Jerva [11] described the venous patterns, showing that there is a right predominance in most subjects. Furthermore, the superior sagittal sinus is more often in continuity with the right lateral sinus in the ratio of 6:4 or 7:3 [13]. If a dominant venous sinus becomes obstructed, in the presence of inadequate cross communication at the trochlear, venous drainage may be sufficiently impaired to cause raised intracranial pressure. Probably because of this anatomical variation, otitic hydrocephalus is expected to be seen more commonly in right-sided ear disease [13], although our case occurred after left-sided sinus thrombosis.

Other possibilities include increased intracranial pressure secondary to brain edema as brain biopsies reveal interstitial edema [4], or a vasomotor reflex phenomenon originating from the thrombosed epithelium of the vein [14], but none of these mechanisms have been proven.

Radiologic findings help us to identify possible physiologic mechanisms of otitic hydrocephalus. Magnetic resonance imaging is the imaging modality of choice because it allows for superior evaluation of the venous sinuses [4]. In patients with a history of otitis media and symptoms such as vomiting, headache, and visual impairment, MRI scan should be carefully studied to facilitate the early diagnosis of a potentially life-threatening complication [15]. Magnetic resonance venography (MRV) can be used for the determination of intrasinus thrombosis, which could not be diagnosed by classic venography [5].

The goals of the therapy are eradication of ear disease and lowering of the elevated intracranial pressure. If the ear disease is acute, it may be resolved spontaneously. Persisting middle ear infection has to be treated on its own merits. Surgical procedures should include cleaning the disease completely from middle ear and mastoid and draining the perisinus abscess if present. Currently, complete mastoidectomy with evacuation of all middle ear and mastoid disease, drainage of the perisinus abscess, and clot removal, along with high-dose appropriate antimicrobial medications administered intravenously, is considered to be adequate to control suppurative thrombophlebitis. Ligation of the internal jugular vein is reserved for those cases in which sepsis continues despite an adequate surgical procedure and appropriate intravenous antibiotic therapy [4]. Treatment of raised intracranial pressure includes the use of steroids, diuretics such as acetozolamide, and hyperosmolar dehydrating agents (mannitol). Repeated lumbar puncture has been advocated, but this is not free from risk in the presence of raised intracranial pressure. Furthermore, in some articles, repeated LPs are referred to have only historic importance [5,7]. Because otitic hydrocephalus occurs regularly in conjunction with lateral sinus thrombosis, anticoagulation therapy may be considered as a therapeutic option. Although some authors recommend anticoagulants in such cases [15,16], most writers agree that there is no regular place for anticoagulation except in those very rare instances where spreading thrombosis has reached to other intracranial sinuses [9].

4. Conclusion

In patients with acute or chronic ear infections, complaints of headache, blurred vision, diplopia, or photophobia
may be a heralding sign of an intracranial complication. Otitic hydrocephalus is an uncommon complication of otitis media, with potentially significant morbidities, necessitating a high index of suspicion.

References