Int. Adv. Otol. 2009; 5:(2) 277-280

CASE REPORT

Intracranial Hypertension: A Rare Complication Following Glomus Jugulare Surgery

Rosa Perez-Mora, Luis Lassaletta, Javier Gavilan

Department of Otolaryngology. La Paz University Hospital, Madrid/Spain

Benign intracranial hypertension or pseudotumor cerebri has been traditionally defined as increased intracranial pressure with normal cerebrospinal fluid composition in absence of a space-occupying lesion. Intracranial hypertension is a rare complication following otolaryngological surgery. A case of intracranial hypertension following infratemporal surgery of a glomus jugulare tumor is presented to discuss the terminology and physiopathology of this uncommon condition.

Submitted : 04 July 2008 *Revised : 14 April 2009* *Accepted : 10 May 2009*

Glomus jugulare (GJ) are a slow-growing benign tumor of the temporal bone with invasive features [1]. Primary treatment is total surgical removal through the infratemporal fossa approach [2]. Main complications of GJ surgery are bleeding, cerebrospinal fluid (CSF) leak, aspiration, cranial nerves paralysis, cochleovestibular fistulae and deafness [2,3]. The terms benign intracranial hypertension (BIH) and pseudotumor cerebri syndrome are indistinctly used to describe patients with increased intracranial pressure in the absence of a space-occupying lesion. The composition of CSF and other neuroradiologic tests are normal in these patients [1, 4]. Increased intracranial pressure has been rarely described following cranial base surgery [5, 6], being extremely unusual following GJ surgery [3].

A case of intracranial hypertension (ICH) associated with elevated intracranial venous pressure due to sigmoid sinus occlusion following infratemporal surgery for a GJ tumor is presented.

Case Report

A 19-year-old man was referred with a 3-month history of right hearing loss and pulsatile tinnitus. Medical history was unremarkable. Otoscopy showed a reddish mass in the posterorinferior segment of the right tympanic membrane. Tonal audiometry showed conductive hearing loss of 45 dB. Radiological findings on computed tomography (CT) scans, magnetic resonance imaging (MRI) and angiography were compatible with glomus jugulare class C1 tumor (Fisch classification) (Figure 1-3). An infratemporal approach with total tumor resection was performed. The immediate postoperative period was uneventful, except for a House-Brackmann (HB) [7] grade IV facial paresis, and the patient was discharged seven days following surgery.

Three weeks after surgery, the patient developed a light headache and blurred vision. On examination, he was normotensive and afebrile. Visual acuity was reduced, and symmetric bilateral papilledema was identified. Right sided-facial palsy was the only neurological deficit. Unenhanced CT, MRI and angiography of the brain showed postoperative changes, with hesitant delta sign in the right transverse sinus (Figure 4). Lumbar puncture revealed increased CSF opening pressure of 41 cm H2O (normal: lower than 20 cm), with normal CSF constituents. Electrolytes, calcium levels, and full blood count were normal. A diagnosis of ICH was made, and acetazolamide therapy was initiated, with a prompt improvement of the symptomatology.

Corresponding address:
Rosa Mª Pérez Mora
Servicio de Otorrinolaringología Hospital Universitario La Paz
Paseo de la Castellana, 261 28046 - Madrid.
Phone: 34912071651; Fax: 34917277050; E-mail address: rmpmora@yahoo.es

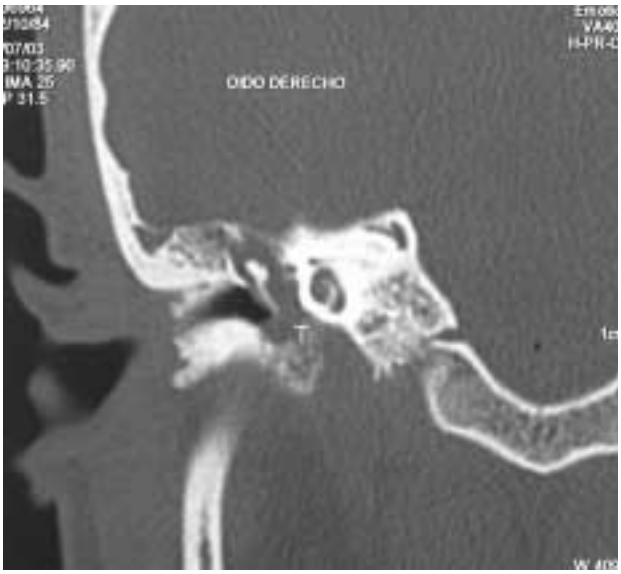

Figure 1. Coronal CT scan showing a soft-tissue mass into middle ear, with minimal bony destruction and extension into hypotympanum and the jugular bulb (T).

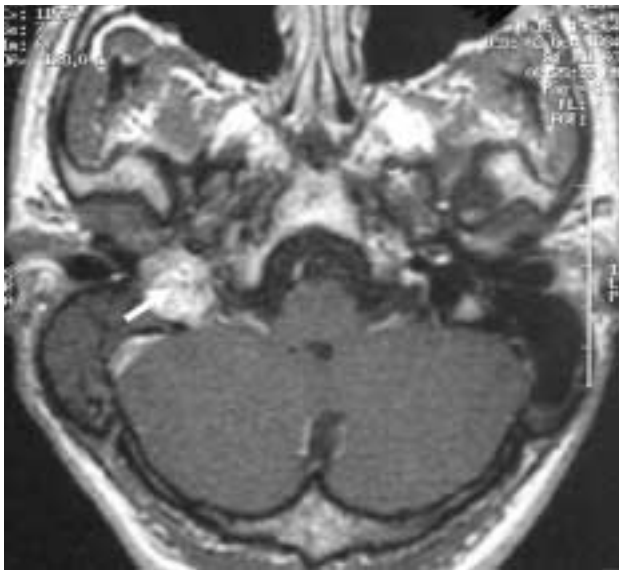

Figure 2. Axial gadolinium-enhanced MR demonstrating right glomus jugulare with middle ear extension (arrow).

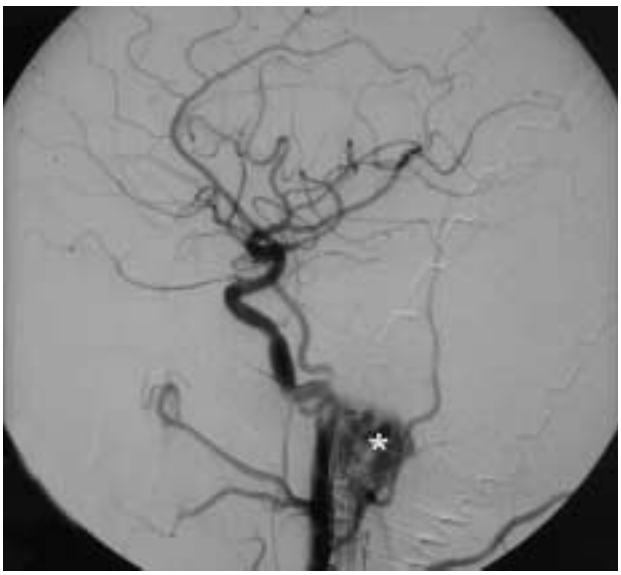

Figure 3. Angiogram showing a vascular lesion (asterisk) 2 cm in diameter in the jugular foramen. The main vascular supply arose from the right occipital artery.

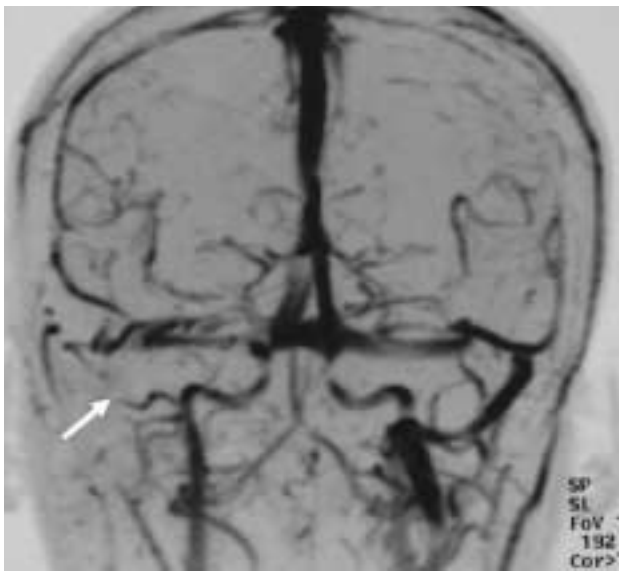

Figure 4. Coronal MR venogram demonstrating occlusion of the right transverse sinus (arrow).

Twenty months after surgery, the patient is still asymptomatic without any treatment. Facial function is HB grade I (normal), and MRI shows no tumor recurrence.

Discussion

The terms pseudotumor cerebri or BIH refer to a syndrome characterized by increased intracranial pressure with normal CSF composition without evidence of another underlying brain pathology [4-6]. In recent years, association of any medication or disorder with ICH has been defined as a secondary pseudotumor syndrome (STS) or a secondary cause of ICH [1, 5]. This condition has been described in cases of arteriovenous and carotid-cavernous fistulae, catheter-induced subclavian vein thrombosis, venous sinus

compression by meningiomas, dural sinus thrombosis and iatrogenic disruption of venous drainage [6, 8, 9]. The occurrence of ICH following otolaryngological surgery is very rare. It has been reported after radical neck dissection, and, very uncommonly, following jugular vein ligation during GJ surgery [3].

The pathogenesis of ICH is still uncertain, although a CSF homeostasis disorder is suggested [4]. Many theories have been proposed such as excess CSF production, increase in cerebral blood volume or brain water content and decreased CSF absorption [9, 10]. The most widely accepted theory postulates impaired CSF absorption, which may result from either an abolition of the pressure gradient between the CSF and the superior sagittal sinus, produced by intracranial venous hypertension, or from an increased resistance to drainage at the level of the arachnoid granulations [1, 11]. This intracranial venous hypertension may be due to dural sinus occlusion (in our case a sigmoid sinus occlusion caused by surgery). Since sigmoid or transverse sinuses occlusion is more common than symptomatic ICH, other factors may be involved [9]. Unilateral obstruction of a transverse sinus is not always well tolerated, especially in the dominant side [11]. Since the superior sagittal sinus drains directly into the right transverse sinus in 59-70% of cases [12-15], obstruction of the right lateral sinus is more likely to result in secondary inadequate cerebral venous drainage.

Occasionally, patients are asymptomatic and come to medical attention when papilledema is detected on routine eye examination [11]. However, headache is the most common symptom of ICH, and described by almost all patients [4, 5]. It is typically daily, generalized, worse in the morning, and aggravated by eye movements and Valsalva maneuvers. Headache may also be throbbing, with nausea, vomiting and photophobia. Neck, shoulders and arm pain are often involved. Although not specific to ICH, transient visual symptoms (obscurations, blurring and scotomata) and diplopia are commonly noted and sometimes the presenting manifestations [4]. Pulsatile tinnitus is also common. Other symptoms such as numbness, incoordination, decreased sense of smell, weakness or dizziness have been also described.

The hallmark of ICH is papilledema, which may be asymmetric [5]. However, the absence of papilledema does not rule out the diagnosis.

ICH can only be diagnosis by exclusion [3, 7] based on a complete history, complete neurological and ophthalmological examinations and radiological studies followed by a lumbar puncture [8]. CT scan shows normal or small ventricular size, enlarged optic nerve sheaths, empty sella syndrome and enlargement of the subarachnoid space. MRI is the preferred imaging study in patients with suspected intracranial hypertension [4, 5, 11]. Nonspecific MRI findings include a partially empty sella, flattening of the posterior sclera, dilation and tortuosity of the optic nerve sheath and sometimes gadolinium enhancement of the optic disc [5]. MR angiography provides a direct visualization of the dural sinuses and can be used to assess sinus flow abnormalities.

CSF examination is critical for the diagnosis of ICH [3], especially when imaging findings do not provide compelling evidence of intracranial hypertension. An opening pressure of greater than 25 cm H2O is accepted as diagnostic criteria.

Most cases if ICH are successfully managed with medical treatment. Carbonic anhydrase inhibitors (acetazolamide) are the main treatment of papilledema as they decrease the rate of CSF production. Corticosteroids are generally reserved for urgent and short-term therapy in patients with visual loss resulting from fulminant papilledema. Surgery is indicated for worsening of vision that cannot be controlled medically [4, 5]. The two major techniques are optic nerve sheath fenestration and more frequently, lumboperitoneal shunting. In case of dural sinus trombosis, the direct endovascular tromboliytic therapy is a safe and effective treatment.

References

1. Angeli SI, Sato Y, Gantz BJ. Glomus jugulare tumours masquerading as benign intracranial hypertension. Arch Otolaryngol Head Neck Surg 1994; 120:1277-1280.

2. Sanna M, Jain Y, De Donato G, Lauda L, Taibah A. Management of jugulare paragangliomas: the Gruppo Otologico experience. Otol Neurotol 2004; 25:797-804

3. Brookes GB, Graham MD. Benign intracranial hypertension complicating glomus jugulare tumor surgery. Am J Otol 1984; 5:350-354.

4. Friedman DI. Pseudotumor cerebri. Neurol Clin N Am 2004;22:99-131.

5. Binder DK, Horton JC, Lawton MT, McDermott MW. Idiopathic intracranial hypertension. Neurosurgery 2004;54:538-551.

6. Lam BJ, Schatz NJ, Glaser JS, Bowen BC. Pseudotumor cerebri from cranial venous obstruction. Ophthalmology 1992; 99:706-712.

7. House JW, Brackman DE. Facial nerve grading system. Otolaryngol Head Neck Surg 1985; 93:146-7.

8. Beck DW, Kassell NF, Drake CG. Glomus jugulare tumor presenting with increased intracranial pressure: case report. J Neurosurg 1979; 50:823-825.

9. Karahalios DG, Rekate HL Khayata MH, Apostolides PJ. Elevated intracranial venous pressure as a universal mechanism in pseudotumor cerebri of varying etiologies. Neurology 1996; 46:198-202.

10. Johnston I, Hawke S, Halmagyi M, Teo C. The pseudotumor syndrome: Disorders of cerebrospinal fluid circulation causing intracranial hypertension without ventriculomegaly. Arch Neurol 1991; 48: 740-747

11. Keiper GL, Sherman JD, Tomsick TA, Tew JM. Dural sinus thrombosis and pseudotumor cerebri: unexpected complications of suboccipital craniotomy and translabyrinthine craniectomy. J Neurosurg 1999; 91:192-197.

12. Ayanzen RH, Bird CR, Keller PJ, McCully FJ, Theobald MR, Heiserman JC. Cerebral MR venography: normal anatomy and potential diagnostic pitfalls. AJNR Am J Neuroradiol 2000; 21:74-78.

13. Ernemann U, Löwenheim H, Freudenstein D, Koerbel A, Heininger A, Tatagiba M. Hemodynamic evaluation during ballon test occlusion of the sigmoid sinus: clinical and technical considerations. AJNR Am J Neuroradiol 2005; 26:179-182.

14. Hwang SK, Gwak HS, Pack SH, Kim DG, Jung HW. The experience of ligation of transverse or sigmoid sinus in surgery of large petroclival meningiomas. J Korean Med Sci 2002; 17:544-548.

15. Miabi Z, Midia R, Rohrer SE, Hoeffner EG, Vandorpe R, Berk CM, Midia M. Delineation of lateral tentorial sins with contrast-enhanced MR imaging and its surgical implications. AJNR Am J Neuroradiol 2004; 25:1181-1188.