Behçet’s disease presenting as a peripheral vestibulopathy

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Abstract

Prolonged acute spontaneous vertigo can be secondary to acute vestibular neuritis or posterior circulation ischaemia. We present a 66-year-old man who first developed an acute vestibular syndrome with profound unilateral hearing loss 34 years ago. First treated as vestibular neuritis, he subsequently developed manifestations of Behçet’s disease, including mouth ulcers, genital ulcers and erythema nodosum over a period of 10 years. Subsequently, sudden sensorineural hearing loss affecting his only hearing ear responded to immunomodulation, confirming an autoimmune cause for the audiovestibular symptoms. This report serves as a reminder that vestibular neuritis seldom causes hearing loss; ischaemic, infective and autoimmune causes should be sought when an acute vestibular syndrome is accompanied by hearing impairment.

1. Case report

A 66-year-old man first presented in 1979 with acute spontaneous vertigo lasting 12 hours, right-sided hearing loss, tinnitus and imbalance. He had third degree left-beating spontaneous vertigo lasting 12 hours, right-sided hearing loss, tinnitus and imbalance.

Keywords:
Behçet’s
Hearing loss
Vestibular

Acknowledgements

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

http://dx.doi.org/10.1016/j.jocn.2013.10.011

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but preserved for left ear stimulation, indicating dysfunction of the right otolith organs (Fig. 1B). Auditory brainstem responses showed absence of waves I–V upon right ear stimulation (Fig. 1C). These abnormalities represented a predominantly right-sided audiovestibular loss.

Audiometry revealed a new 30–40 dB decline in low frequency hearing for the left ear, with a pure tone average (PTA) of 45 dB HL (normal range –10 to 15 dB HL) (Fig. 2A) [1]. All audiovestibular symptoms were now attributed to exacerbations of Behçet’s disease. He was therefore treated with intra-tympanic dexamethasone after which his hearing improved to a PTA of 32.5 dB HL. He undertook home-audiometry performed on a portable Widex SP3 device (Widex, Copenhagen, Denmark). PTA fluctuations between 18.75 and 46.25 dB HL were recorded (Fig. 2B); one episode of hearing loss was also associated with acute spontaneous vertigo lasting 4 hours. During the ensuing 4 week period, sudden drops in hearing threshold were treated with pulsed intravenous (IV) methyl prednisolone (two IV infusions of 1 g over 2 days) and two further intra-tympanic steroid injections followed by oral prednisone, commencing at 50 mg daily. To enable withdrawal of steroids, he was administered IV infliximab (400 mg over 1 hour, two doses at 6 weeks apart). Although he developed tremulousness, malaise, and fevers directly following the second IV infusion, no further episodes of hearing loss or vertigo occurred. A follow-up series of home-audiometry recorded PTA of 20–25 dB HL.

2. Discussion

This patient first presented with an acute vestibular syndrome of a single episode of severe, long-lasting, spontaneous vertigo, which is most commonly attributed to vestibular neuritis. However, hearing loss is not a feature of vestibular neuritis, which commonly affects the superior division of the
Neuronitis affecting the inferior nerve or both divisions can cause hearing loss, but is exceedingly uncommon, accounting for <2% of patients [2]. Acute vertigo with unilateral hearing impairment can occur in herpes zoster oticus, Meniere’s disease, labyrinthine haemorrhage, labyrinthine infarction, anterior inferior cerebellar artery infarction, and less commonly, in autoimmune inner ear diseases. The absence of vesicles and profound right-sided hearing loss across all frequencies made the first two diagnoses unlikely. The subsequent development of mouth ulcers, genital ulcers and typical skin lesions identified Behçet’s disease as the underlying disorder [3]. The later recurrence of vertigo and hearing impairment, which responded to immunosuppression, confirmed an autoimmune cause for his audiovestibular symptoms.

First described in 1937 by Dr Halusi Behçet in Istanbul, Behçet’s disease was prevalent along the Silk Road, but became widespread with immigration [3]. It is a chronic relapsing inflammatory disorder of unknown aetiology, affecting small vessels of nearly all organs [3]. The underlying histopathologic findings include leukocytoclastic vasculitis, fibrinoid necrosis of postcapillary venules, or perivascular neutrophilic accumulations [4]. Originally described as a classical triad of oral ulcers, genital ulcers and ocular disease including anterior uveitis, it is now acknowledged to be multisystemic. Central nervous system involvement occurs in 10–25% of patients and can span the entire neuraxis [5]. Audiovestibular symptoms include vertigo, sudden sensorineural hearing loss, tinnitus and orthostatic imbalance [6,7]. Vertigo lasting several days, occurring alone or with hearing impairment, has been described. The prevalence of otological involvement ranges between 12–80% in separate studies [6,7]. Audiometry in these patients showed mild sensorineural hearing loss without specific attributes that enable its recognition. Based upon diverse auditory brainstem response abnormalities ranging from normal waveforms in the presence of sensorineural hearing loss (implying cochlear pathology) to delayed I–III or I–V interpeak latencies (indicating brainstem involvement), the auditory pathways are thought to be involved at multiple sites [8]. Caloric weakness [7] implying abnormal horizontal canal function and prolonged VEMP peak latencies (suggesting central vestibulopathy) have been reported [9].

This study draws attention to vertigo and hearing loss as initial manifestations of Behçet’s disease. It reinforces the necessity to carefully investigate acute vestibular syndrome when it is accompanied by severe sensorineural hearing loss. Vestibular neuronitis is seldom responsible for vertigo accompanied by hearing impairment and should only be considered after alternate diagnoses are excluded.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

References

Intracranial hypotension in the setting of concurrent perineural cyst rupture and subarachnoid hemorrhage

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ARTICLE INFO

Article history:
Received 10 October 2013
Accepted 18 October 2013

Keywords:
Aneurysm
Coma
Epidural blood patch
Intracranial hypotension
Perineural cyst
Subarachnoid hemorrhage
Tarlov cyst

ABSTRACT

Although most patients with intracranial hypotension typically present with headaches, the rest of the clinical spectrum is characteristically non-specific and often quite variable. In a patient with concurrent pathologies that can produce a similar clinical picture, a high index of suspicion must be maintained to achieve the correct diagnosis. The authors report a patient with intracranial hypotension in the setting of concurrent perineural cyst rupture and subarachnoid hemorrhage. A 63-year-old woman with a family history of ruptured intracranial aneurysms presented after a sudden thunderclap headache and was found to have diffuse subarachnoid hemorrhage. Imaging revealed anterior communicating and superior hypophyseal artery aneurysms. Following the uneventful clipping of both aneurysms, the patient experienced a delayed return to her neurological baseline. After it was noted that the patient had an improved neurological examination when she was placed supine, further investigation confirmed intracranial hypotension from perineural cyst rupture. The patient improved and returned to her neurological baseline after undergoing a high-volume blood patch and remained neurologically intact at postoperative follow-up. Although intracranial hypotension is known to be commonly associated with cerebrospinal fluid leak, its causal and temporal relationship with subarachnoid hemorrhage has yet to be elucidated.

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1. Introduction

The confounding nature of the symptoms of intracranial hypotension, along with the infrequency with which it occurs, may make the condition a diagnostic quandary for neurosurgeons. Although most patients typically present with headaches, the rest of the clinical spectrum is characteristically non-specific and often quite variable. This can lead to a significant delay in diagnosis, resulting in increased morbidity and possible mortality [1]. In a patient with concurrent pathologies that can produce a similar clinical picture, a high index of suspicion must be maintained to achieve the correct diagnosis. We present a 63-year-old woman who developed intracranial hypotension after subarachnoid hemorrhage (SAH) in whom diagnosis of a large perineural cyst in the upper thoracic spine was delayed. In this report, we aim to explore the relationship between the two entities.

2. Case presentation

2.1. History and presentation

This 63-year-old woman with a family history of ruptured intracranial aneurysms presented to our institution after the sudden onset of a severe “thunderclap” headache. On arrival, the patient complained of headache and neck stiffness with intermittent nausea and vomiting. She was somnolent but easily rousable and otherwise neurologically intact (Glasgow Coma Scale score of 14). A CT scan of the brain obtained upon admission demonstrated diffuse SAH with blood in the Sylvian fissures bilaterally as well as in the basal cisterns (Fig. 1). CT angiography of the brain confirmed the presence of both an anterior communicating artery aneurysm and a right superior hypophyseal artery saccular aneurysm (Fig. 2). The patient’s medical history included mucocutaneous systemic lupus erythematosus (mSLE) that was controlled with hydroxychloroquine, but she had no history of hypertension or tobacco use. All initial laboratory values were within normal parameters.

On admission to the intensive care unit, the patient was promptly placed on our institutional SAH protocol, including strict blood pressure control, hydration, vasospasm monitoring, and seizure prophylaxis. Preparation for surgery was delayed because of difficulty acquiring blood products for possible transfusion because mSLE caused significant agglutination during the blood type and cross matching. Three days after her admission for SAH, the patient underwent clipping of both the right superior hypophyseal and the anterior communicating artery aneurysms with placement of an external ventricular drain.

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