spontaneously. Seromas are typically seen after breast and axillary surgery [6]. Risk factors for seroma formation include obesity, extensive surgery, and the use of electrocautery for skin flap dissection [7–9]. Both hematomas and seromas can lead to wound dehiscence and infection [10,11]. Examination of the wound can provide clues about the character of the fluid collection, and ultrasound or CT scan can help identify these pockets.

We had meticulously dissected the soft tissues during exposure of the high bifurcating carotid endarterectomy in this patient to avoid damage to the parotid or submandibular glands. Nonetheless, it is likely that retraction injury led to indirect damage to these glands, causing salivoma formation. Since the area of drainage involved the right aspect of the neck, vigorous or even passive motion of the head might have contributed to salivoma formation.

If a salivoma is suspected after exclusion of an infectious etiology, we recommend placement of a Penrose drain and avoidance of unnecessary exploratory surgery. Early ENT consultation will assist with management of salivomas.

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.

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

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Behçet's disease presenting as a peripheral vestibulopathy

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ARTICLE INFO

Article history:
Received 9 December 2012
Accepted 10 August 2013

Keywords:
Behçet's
Hearing loss
Vestibular

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 nystagmus. Bithermal caloric testing revealed a 78% right canal paresis. Audiometry revealed profound right sensorineural hearing loss. A diagnosis of vestibular neuronitis was made. His imbalance improved, yet the hearing loss was permanent.

Ten years later, the patient developed mouth ulcers, genital ulcers and red macular coin-shaped lesions over both lower limbs, which were consistent with erythema nodosum. A clinical diagnosis of Behçet's disease was made. Recurrent ulcers were successfully treated with self-initiated oral thalidomide 100 mg twice daily for 3–4 days. A further 10 years later, he experienced a sudden left-sided hearing loss. He had a positive rightward bedside head impulse, left-beating head-shaking nystagmus and a rightward Unterberger test. Ice water caloric stimulation showed low amplitude responses for both ears with a persistent right canal paresis (33%) and video head impulse testing showed marked reduction in the vestibulo-ocular reflex gain for the right ear (0.3–0.6), with a shower of "catch up saccades" for rightward rotation and a small number of saccades for leftward rotation, implying a right, more than left horizontal canal dysfunction (Fig. 1A). Ocular and cervical vestibular evoked myogenic potentials (VEMP) to air and bone conducted sound were absent for right ear stimu-

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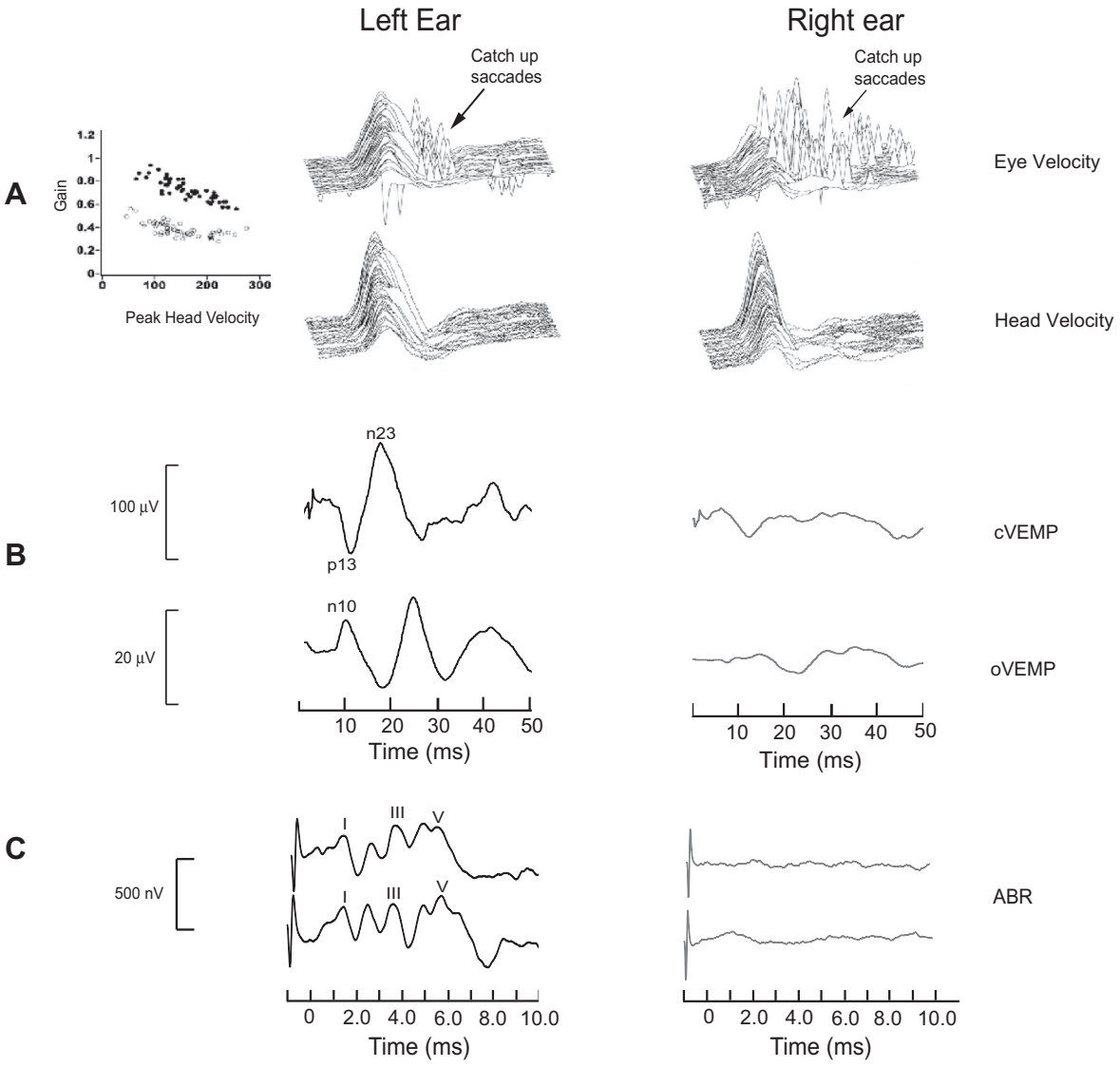

Fig. 1. Results of audiovestibular tests. (A) Video head impulse showing head and eye velocity recorded from a lightweight pair of video goggles while the leftward and rightward horizontal head impulses were performed. The scatterplot indicates vestibulo-ocular reflex gain (eye velocity/head velocity) against the peak velocity for head movement. (B) The traces represent averaged sternocleidomastoid electromyography (for cervical vestibular evoked myogenic potentials [cVEMP]) and averaged infra-orbital surface EMG for ocular vestibular evoked myogenic potentials (oVEMP). cVEMP and oVEMP responses were preserved for left ear stimulation with bone conducted sound but were absent following right ear stimulation. (C) Traces representing an average of 1000 surface recordings to 90 dB HL clicks. Upon left ear stimulation, a normal auditory brainstem response (ABR) with normal waveforms (I–V) and latencies was recorded. Right ear stimulation showed absent waves I–V. No cochlear microphonic was visible.

lation 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 neuronitis. However, hearing loss is not a feature of vestibular neuronitis, which commonly affects the superior division of the

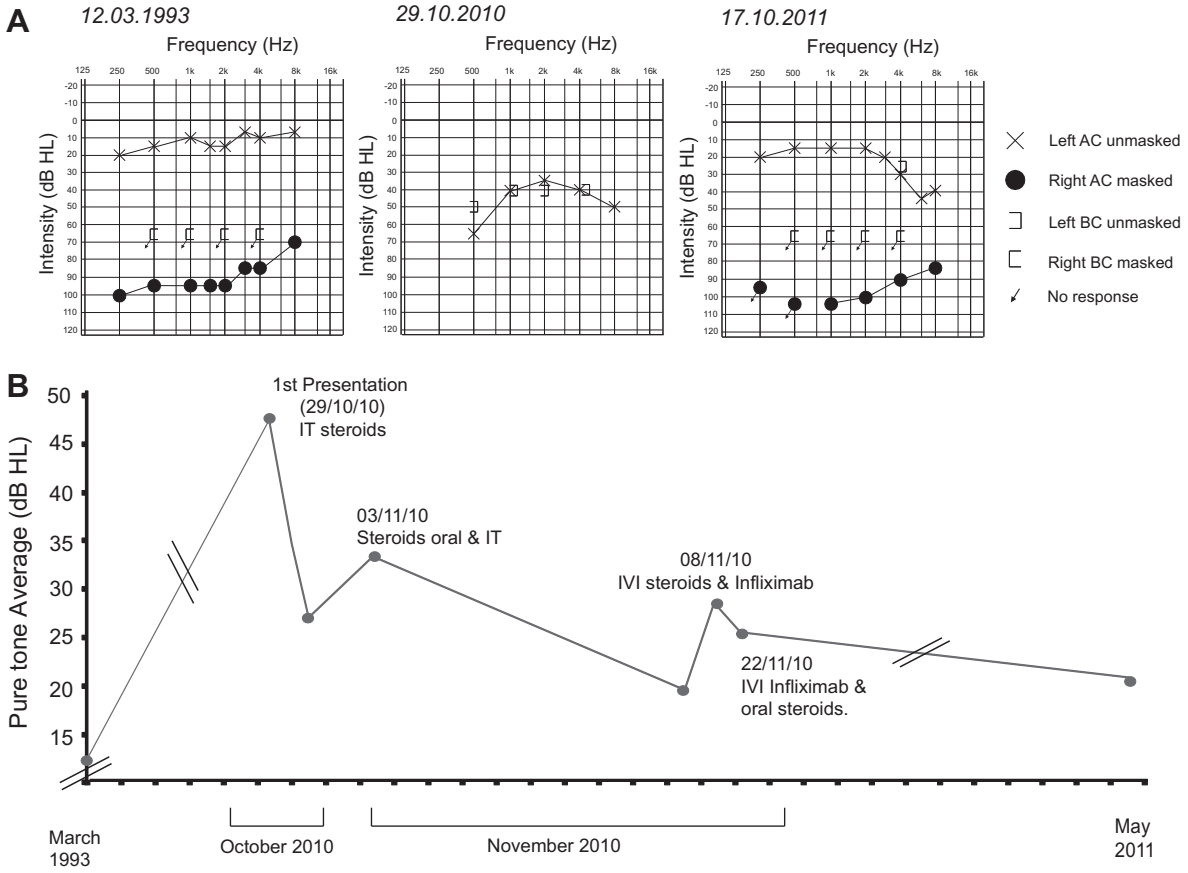

Fig. 2. Pure tone audiograms (A) after the initial episode of audiovestibular symptoms (right) followed by the initial audiogram performed after left sided hearing loss (centre) and final audiogram after treatment (right). (B) Pure tone average (across three frequencies: 500 Hz, 1 kHz, 2 kHz) for the left ear and treatments as a function of time. AC = air conducted, BC = bone conducted, HL = hearing level, IVI = intravenous infusion, IT = intratympanic.

vestibular nerve [2]. 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.

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http://dx.doi.org/10.1016/j.jocn.2013.08.021

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.

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.

2.2. Operation

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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