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Papilledema secondary to vestibular schwannoma: An atypical case without intracranial hypertension

Œdème papillaire secondaire à un schwannome vestibulaire : un cas atypique sans hypertension intracrânienne

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ABSTRACT

In most cases, vestibular schwannomas with papilledema are associated with intracranial hypertension secondary to hydrocephalus (obstructive or communicating). We describe the atypical case of a 39-years-old man who presented with bilateral papilledema revealing a vestibular schwannoma, but without hydrocephalus and with normal intracranial pressure. Ophthalmologic signs were completely resolved after tumor removal. The pathophysiological mechanism generally described to explain bilateral papilledema in such cases is tumor-induced hyperproteinorachia. However, in the absence of hydrocephalus or intracranial hypertension, this case raises the question of the mechanisms involved in the visual impairment related to vestibular schwannoma.

RÉSUMÉ

Les schwannomes vestibulaires avec œdème papillaire sont en général associés à une hypertension intracrânienne secondaire à une hydrocéphalie (communicante ou obstructive). Nous décrivons ici le cas atypique d'un homme de 39 ans ayant présenté un œdème papillaire révélateur d'un schwannome vestibulaire, mais sans hydrocéphalie et avec une pression intracrânienne normale. L'exérèse tumorale a permis une régression complète des signes ophtalmologiques. En l'absence d'obstruction ventriculaire, le mécanisme physiopathologique généralement décrit pour expliquer l'atteinte visuelle est l'hydrocéphalie secondaire à une hyperprotéinorachie. Cependant, en l'absence d'hydrocéphalie ou d'hypertension intracrânienne, ce cas soulève la question des mécanismes impliqués dans l'atteinte visuelle secondaire à la présence d'un schwannome vestibulaire.

1. Introduction

Vestibular schwannomas (VS), or acoustic neuromas, are the most frequent cerebellopontine angle tumors [1]. They are usually revealed by auditory and labyrinthine symptoms. Ophthalmologic signs such as papilledema are not commonly associated with VS [2], but are consistently secondary to increased intracranial pressure

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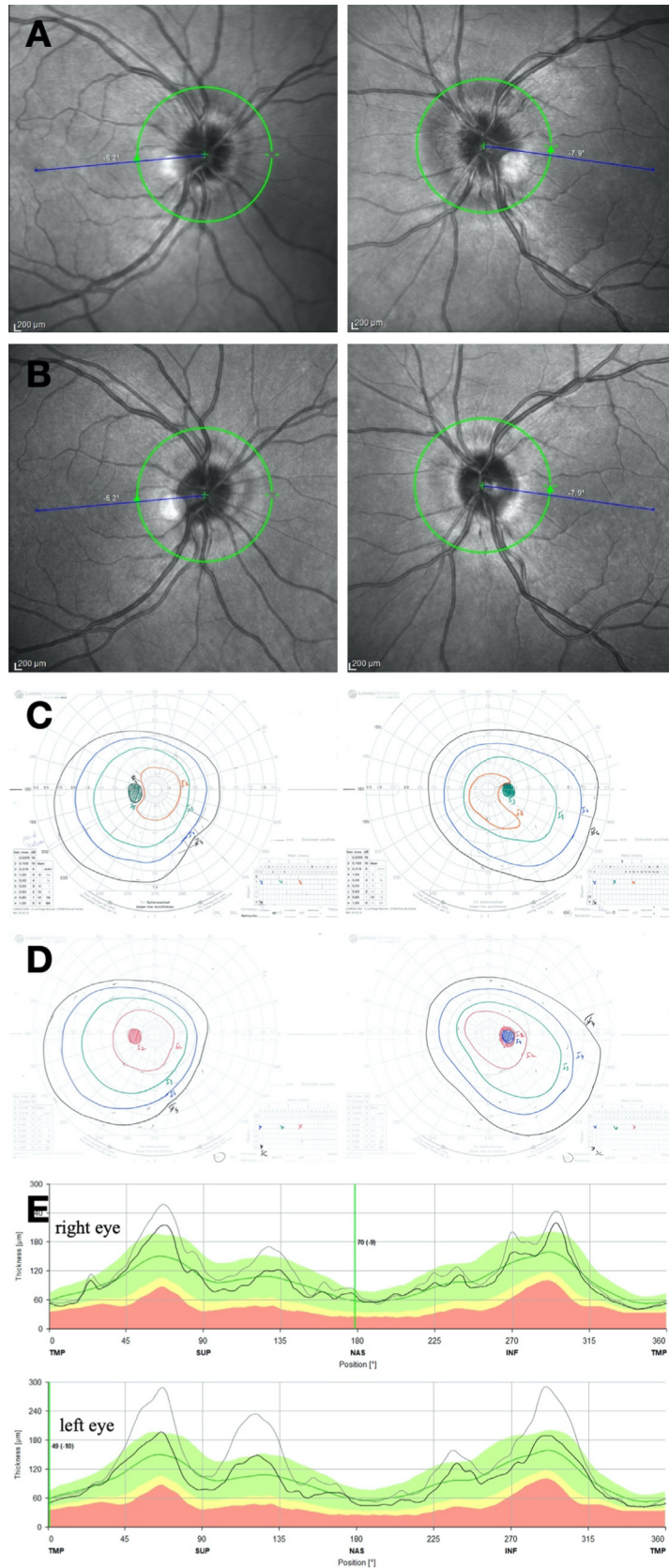


Fig. 1. Pre- and postoperative ophthalmologic findings. A. Bilateral papilledema on preoperative funduscopy. B. Bilateral papilledema resolution 3 months after surgery. C. Enlarged bilateral blind spots on preoperative visual field. D. Postoperative visual field improvement. E. Optical coherence tomography (OCT) showing a decrease of optic fibers oedema (thickness) for both eyes after surgery; the grey line represents preoperative thickness, the black line represents 3-months postoperative thickness, and the green line represents the average thickness of optic fibers.

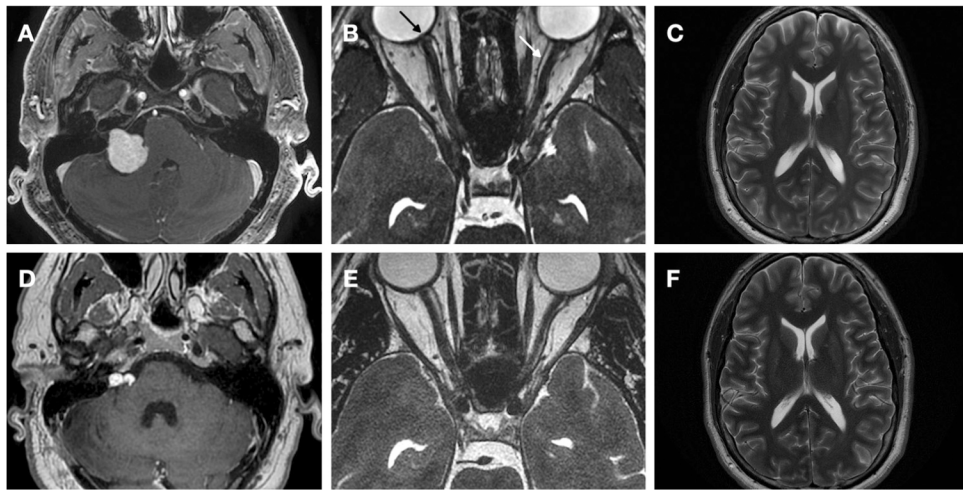


Fig. 2. Pre- and postoperative magnetic resonance imaging (MRI). A. Axial 3D T1-weighted fat-suppressed gadolinium-enhanced (FSPGR) MRI showing right cerebellopontine angle tumor extending into the right canal auditory, with brainstem compression. B. Axial 3D fast imaging employing steady-state acquisition (FIESTA) MRI showing a posterior globe surface flattened (black arrow) and a slight dilatation of optic nerves sheaths (white arrow). C. Pre-operative Axial T2-weighted MRI showing the absence of hydrocephalus (frontal ventricles). D. Axial 3D T1-weighted CUBE gadolinium-enhanced MRI showing subtotal tumor resection. E. Axial 3D-FIESTA MRI showing a decrease of optic nerves sheaths dilatation. F. Postoperative Axial T2-weighted MRI showing no change in the size of ventricles.

(ICP > 20 cm H₂O) [3]. Hydrocephalus, whether communicating or obstructive, is the leading cause of intracranial hypertension in VS patients [4]. However, few cases of VS with papilledema without hydrocephalus have been described, but these cases were mostly accompanied with raised ICP. We present here an atypical case of patient with symptomatic bilateral papilledema revealing a VS, without intracranial hypertension or hydrocephalus.

2. Case presentation

A 39-years-old male with no medical or surgical history, presented with headaches and visual impairment progressing for 2 weeks. He also complained of right hearing loss and static disorders that have been evolving for 9 years.

Neurologic examination showed right-sided perceptive deafness and tinnitus, unstable gait, positive Romberg's sign and no right facial palsy or hypoesthesia.

Ophthalmologic examination revealed bilateral papilledema on funduscopy, confirmed by optical coherence tomography (OCT). Bilateral visual acuity was preserved (10/10, P2). Visual field showed enlarged bilateral blind spots (Fig. 1A, C, E).

Preoperative MRI showed a 26 mm × 21 mm (volume = 6.5 cc) right cerebellopontine angle tumor suggestive of a Koos grade 4 vestibular schwannoma [5], with slight dilatation of optic nerve sheaths and a flattened posterior globe surface. No hydrocephalus or cerebral venous sinus thrombosis was identified (Fig. 2A–C).

The lumbar puncture showed hyperproteinorachia (1.33 g/L, $n < 0.45$) and an opening pressure of 18 cm H₂O.

Tumor surgery allowed a subtotal removal using a retrosigmoid approach with facial nerve monitoring (Fig. 2D).

Postoperative course showed a progressive visual improvement, persistent right deafness, regression of right tinnitus, improvement of cerebellar syndrome and a partial right facial palsy (House and Brackmann grade 2) [6] resolving in 3 months. Histological examination confirmed the vestibular schwannoma.

Papilledema decreased at 1 month and was completely resolved at 3 months follow-up. Visual field was improved at 3 months follow-up (Fig. 1B, D, E).

The size of the ventricles remained stable and the dilatation of the optic nerve sheaths decreased (Fig. 2E, F).

3. Discussion

We reported a patient with a VS associated with bilateral papilledema without raised ICP, which is unusual regarding to other published cases and pathophysiological aspects.

Few cases of VS with papilledema have already been described with similar clinical and radiological features, but the authors did not report intracranial pressure measurements as they did not perform lumbar puncture, not allowing them to confirm their hypothesis [7,8]. To our knowledge, only one previous case without raised ICP has been reported [9], but authors described however an ICP measured above 20 cm H₂O (21 cm H₂O). Also, Grainger et al. [10] reported another case of VS with papilledema without ICP (14 cm H₂O), but this could be explained by their measurement method, i.e. under general anesthesia. However, it is important to remind that ICP value may fluctuate throughout the day and could depend on the measurement method [11]. Moreover, intracranial hypertension is classically defined as > 27 cm H₂O and normal ICP < 20 cm H₂O, with the so-called “grey zone” between 20 and 27 cm H₂O [3].

In the absence of obstruction, the most widespread pathophysiological hypothesis to explain papilledema in VS is intracranial hypertension (also known as pseudotumor cerebri). The non-obstructive hydrocephalus could be the result of Pacchioni granulations obstruction due to VS's excessive proteins production [12–14]. But in our case, the absence of elevated ICP may suggest an additional mechanism in relation to papilledema. Like spinal nerve sheaths, optic nerves may physiologically participate to the CSF resorption [15,16]. In this view, we can suggest that hyperproteinorachia may block the CSF resorption capacities of optic nerve sheaths, thus leading to a CSF trapping into the sheath and then to a local “hypertension” of the optic nerve manifested by papilledema. Interestingly, Jefferis et al. [17] showed papilledema regression after optic nerve sheath fenestration in VS with disc oedema, which is also an argument for optic nerve involvement in CSF resorption.

It should be noted that vestibular schwannomas secrete many factors such as TNFalpha or interleukines [18]. One can also mention a direct damage to the optic nerve through the tumor secretion, as described in TNFalpha-related ototoxicity [19]. However, such a pathologic mechanism concerning the optic nerves has never been described, and would require further explorations at the cellular or molecular level.

Finally, in our case we cannot exclude an ICP measurement error (i.e. one-time versus continuously), and thus an unawareness of a very early or slight intracranial hypertension but without raised ICP at the time of lumbar puncture. Indeed, Funnell et al. [20] studied a population of idiopathic intracranial hypertension, with or without papilledema, and defined a 24-hour intracranial pressure threshold at 13.6 cm H₂O (average) as a good predictor for papilledema.

4. Conclusion

The involvement of the VS in visual impairment recorded in our case seems clearly established since its resection allowed a full visual recovery. However, the pathophysiological mechanism is not deciphered, and appears to be independent of intracranial pressure. Investigations at cellular level are mandatory to better elucidate the link between VS and papilledema.

Human and animal rights

The authors declare that the work described has been carried out in accordance with the Declaration of Helsinki of the World Medical Association revised in 2013 for experiments involving humans as well as in accordance with the EU Directive 2010/63/EU for animal experiments.

Informed consent and patient details

The authors declare that they obtained a written informed consent from the patients and/or volunteers included in the article and that this report does not contain any personal information that could lead to their identification.

Disclosure of interest

The authors declare that they have no competing interest.

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Author contributions

All authors attest that they meet the current International Committee of Medical Journal Editors (ICMJE) criteria for Authorship.

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