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Clinical case

VIIIth nerve cavernous hemangioma mimicking a stage 1 acoustic schwannoma[☆]



Hémangiome caverneux du nerf cochléo-vestibulaire imitant un petit neurinome de l'acoustique

G. Bonfort^{a,d,*}, F. Veillon^c, C. Debry^a, P. Kehrli^b, S. Chibbaro^b

^a Department of ENT, Hautepierre University Hospital, Strasbourg, France

^b Department of neurosurgery, Hautepierre University Hospital, Strasbourg, France

^c Department of medical imaging, Hautepierre University Hospital, Strasbourg, France

^d ENT department, Legouest, Instructional Military Hospital, 27, avenue de Plantières, BP 90001, 57077 Metz cedex 3, France

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ABSTRACT

Objective. – To report a case of VIIIth nerve cavernous hemangioma, a very rare differential diagnosis among the various pathologies presenting as small enhancing entities into the internal auditory canal. It is one of the most challenging when imaging is not able to differentiate it from an intrameatal vestibular schwannoma.

Methods and results. – We report a cavernous hemangioma extruding from the internal auditory canal, diagnosed after a left translabyrinthine resection in a 45-year-old man complaining of profound sensorineural hearing loss, with no facial paresis or dizziness. The preoperative differential diagnosis of a vestibular schwannoma was impossible, due to the absence of calcifications that usually characterize temporal bone hemangiomas. Clinical presentation, radiological features and treatment considerations are discussed along with up-to-date review of pertinent literature.

Conclusions. – When considering an apparent small intra-auditory canal schwannoma, otoneurologists should be aware of the rare possibility of a cavernous hemangioma. Early diagnosis and surgical treatment may improve the functional outcome, possibly preserving neural integrity.

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RÉSUMÉ

Objectifs. – Décrire un cas d'hémangiome caverneux du nerf cochléo-vestibulaire, diagnostic différentiel très rare parmi les pathologies variées siégeant dans le conduit auditif interne. Cette entité est particulièrement difficile à discerner lorsque l'imagerie ne permet pas sa différenciation d'un schwannome vestibulaire de stade 1 selon Koos.

Méthodes et résultats. – Nous rapportons un cas d'hémangiome caverneux du conduit auditif interne, découvert chez un homme de 45 ans opéré par voie translabyrinthique pour ce que l'on suspectait être un schwannome vestibulaire intra-méatal, à gauche. Il se plaignait de surdité neurosensorielle profonde, sans troubles vertigineux ni parésie faciale. La différenciation préopératoire était radiologiquement impossible, du fait de l'absence des trabéculations osseuses qui caractérisent habituellement les hémangiomes de l'os temporal. Nous discutons les caractéristiques cliniques et radiologiques de la pathologie, ainsi que les perspectives thérapeutiques.

Conclusions. – Face à un apparent petit schwannome intra-canalaire, les oto-neurologistes doivent être avertis qu'il puisse s'agir d'un hémangiome du VIII. Lorsque le diagnostic est précoce, le traitement chirurgical préservant le nerf améliore possiblement la fonction.

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Mots clés :

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Schwannome de l'acoustique

Hémangiome de l'os temporal

Nerf facial

Abbreviations: CH, Cavernous Hemangioma; CPA, Cerebellopontine Angle; CSF, Cerebrospinal Fluid; CT, Computed Tomography; IAC, Internal Auditory Canal; MRI, Magnetic Resonance Imaging; VS, Vestibular Schwannoma.

[☆] The work was carried out at Hautepierre University Hospital, Strasbourg, France.

* Corresponding author.

E-mail address: gratienbonfort@yahoo.fr (G. Bonfort).

1. Introduction

Vascular lesions of the CPA (cerebellopontine angle) and IAC (internal auditory canal) are quite rare lesions including cavernoma, hemangioma, hemangioblastoma, hemangioendothelioma, vascular ectasia and arterio-venous malformations. Differential diagnoses on the location also include lesions such as schwannomas, meningiomas, hamartomas, lipomas, cholesteatomas, sarcoidosis, rhabdomyomas, lymphoma and metastasis.

Only a few cases of cavernous hemangioma (CH) of the IAC have been reported in international literature and surprisingly only half of them have been histologically confirmed [1–3]. Due to the variety of pathologies in the IAC, the preoperative diagnosis is very challenging.

2. Case Report

A 45-year-old man was known and followed in our combined neurosurgery/ENT clinic for 3 years with a history of progressive, left-sided hearing loss and episodic tinnitus. MRI demonstrated a mass extending into an enlarged left IAC, up to the fundus but without scalloping, showing a progressive volume increase from 4 to 10 mm length within 24 months, thought to be a vestibular

schwannoma (VS) (Fig. 1). The patient did not complain of vertigo and there was no evidence of facial nerve dysfunction. Videonystagmography showed symmetrical vestibular responses. Due to the tumoral growth and profound deafness, the lesion was excised through a left translabyrinthine approach with the patient in park bench position using facial nerve monitoring. The lesion was totally extra-arachnoidal and adherent to the cochlear and inferior vestibular nerves (Fig. 2). Macroscopically it was a 10-mm rounded pink and reddish soft mass being confirmed on histopathology to be a CH, without any bone trabeculation (Fig. 3). In the patient's medical as well as in his family history, there was no notion of cavernomatosis. The patient's postoperative course was uneventful and he was discharged home at day 4. At 6 months follow-up, the patient reported a generalized well-being and MRI showing neither residual nor pathology recurrence.

3. Discussion

3.1. Background

CH is a very rare benign vascular tumor, and it is even rarer when involving the temporal bone where two main forms might be found: cranial nerve-related and primary intra-osseous forms [4,5].

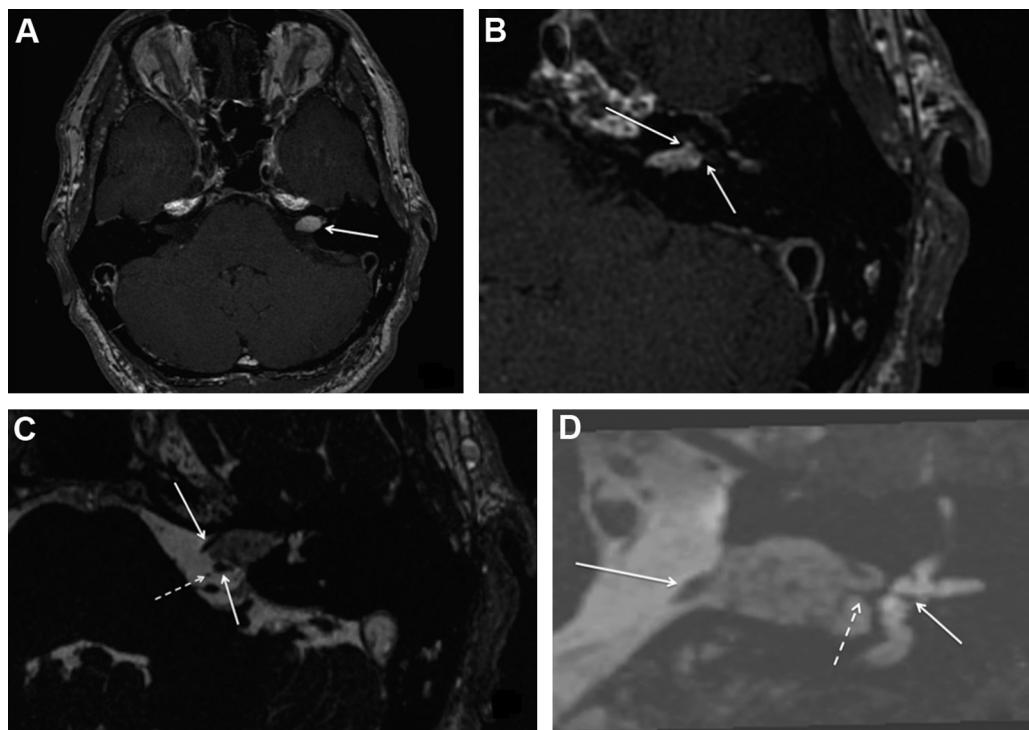


Fig. 1. Cerebral weighted MRI (Unfortunately CT was not performed preoperatively); A: axial post contrast T1 showing an enlarged left IAC (diameter 5.6 mm vs 4.0 mm for the healthy side), a well-delineated 10-mm tumor (white arrow) with strong and homogeneous gadolinium enhancement, mimicking a stage 1 vestibular schwannoma; B: focus on the left IAC axial post contrast T1 shows the fundus filled with the enhancing lesion, which invades the cochlear recess of vestibule (left arrow) and adopts the inferior vestibular nerve direction towards the saccule of the inner ear (right arrow); C: left pontocerebellar angle in axial T2-FIESTA depicting a well-circumscribed hyperintense tumor into the IAC. In contact with the lesion at the porus sections of the cranial nerves may be observed, the VIIth anteriorly (superior arrow) to the lesion and the VIIIth (inferior arrow) posteriorly, with the antero-inferior cerebellar artery (AICA, dotted arrow) (Labyrinthine artery unperceivable); D: focus on the left IAC T2-FIESTA coronal reconstruction shows the moderate-hyperintense tumor enlarging the IAC. VIIth nerve (left arrow), transverse crista falciformis (dotted arrow) and the hyperintense inner ear labyrinth, with its hypointense utricular macula (right arrow), are shown.

IRM cérébrale en coupes axiales (Nous ne disposons malheureusement d'aucun scanner préopératoire faute de réalisation) ; A : séquence T1 avec injection de gadolinium, illustrant au sein du conduit auditif interne gauche, une lésion tumorale (flèche blanche) bien délimitée de 10 mm, qui élargit le CAI (5,6 mm de diamètre contre 4,0 mm côté sain) et qui prend le contraste de manière intense et homogène. Ceci correspondant à l'aspect radiologique d'un schwannome vestibulaire de stade 1 ; B : focalisation sur le fundus du conduit auditif interne gauche en séquence T1 injectée. Il est entièrement envahi par la lésion qui emplit le récessus cochléaire du vestibule (flèche de gauche) et qui poursuit la direction du nerf vestibulaire inférieur vers le saccule (flèche de droite) ; C : séquence T2-FIESTA centrée sur l'angle ponto-cérébelleux gauche. À hauteur du porus et au contact de cette tumeur ronde et hyperintense, courrent les nerfs du paquet acoustico-facial, avec en avant de la lésion le VII (flèche supérieure) et en arrière le VIII (flèche inférieure) ainsi que l'artère cérébelleuse antéro-inférieure (AICA, flèche pointillée) (Artère auditive interne non perceptible) ; D : en séquence T2-FIESTA, reconstruction coronale focalisée sur le CAI gauche, illustrant la dilatation du CAI exercée par la tumeur qui est modérément hyperintense. On y visualise le VII (flèche de gauche), la crête falciforme transverse (flèche pointillée) et le labyrinth de l'oreille interne en hypersignal, incluant la macule utriculaire en hyposignal (flèche de droite).

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