The Journal of Laryngology & Otology, E10, 1 of 4.

doi:10.1017/S0022215107001491

Online Only Clinical Record

Unilateral hearing loss as primary symptom of craniopharyngioma in a six-year-old girl

R Hofman, H J Rosingh

Abstract

Objective: We report a rare case of otological presentation of craniopharyngioma.

Method: Case report and review of world literature concerning presentations of craniopharyngioma.

Results: A six-year-old girl was referred to our department with unilateral hearing loss. This appeared to be a complete sensorineural hearing loss. Otoacoustic emissions were reproducible on both sides. Magnetic resonance scanning revealed a massive, cystic craniopharyngioma exerting pressure on the patient's ventricular system and brainstem and also invading the internal acoustic canal. The mass was resected via a craniotomy. The patient's hearing loss completely recovered, and she experienced no neurological or endocrinological side effects of the treatment. Craniopharyngioma have a prevalence of 0.13–2:100 000.

Conclusion: Craniopharyngioma is a rare disease. First presentation with otological symptoms is extremely rare. Otoacoustic emissions can differentiate between cochlear and retrocochlear causes of sensorineural hearing loss.

Key words: Hearing Loss; Craniopharyngioma; Otoacoustic Emissions

Introduction

Sensorineural hearing loss in children is mostly congenital and cochlear-related. Rarely, sensorineural hearing loss is caused by retrocochlear pathology. Otoacoustic emissions (OAEs) can ideally differentiate between cochlear and retrocochlear pathology. Magnetic resonance imaging (MRI) can confirm the suspicion of retrocochlear pathology.

Craniopharyngiomas are benign, slow-growing tumours located in the sellar and parasellar regions. Craniopharyngiomas arise from epithelial remnants of the craniopharyngeal duct or Rathke's pouch. Rathke's pouch is a bulging of the embryological pharynx which eventually forms the endocrine part of the pituitary gland.

Craniopharyngioma has a prevalence of 0.13–2: 100 000 and a male/female ratio of 1:1.[1] The peak incidence in children occurs between five and 14 years.

First symptoms are listed in Table I. Headaches, nausea and vomiting occur as a result of hydrocephalus developed as a consequence of increased cerebrospinal fluid pressure. Pressure on the optic nerve or optic chiasma is the cause of visual disturbances. Hormonal imbalance is caused by pituitary or hypothalamic invasion or pressure of the mass on these structures.[2]

The differential diagnosis includes other tumours of the central nervous system; in the present case, this included pilocytic astrocytomas, masses of the pineal gland, tumours of the optical nerve and ependymomas.

Case report

A six-year-old girl with right-sided hearing loss was referred to our department by her general practitioner. Her hearing had been impaired for six months. She had no history of otalgia or otorrhoea. Previously, her hearing on the right had been normal (she had always used her right ear when using the telephone). Apart from recurrent headaches and rhinorrhoea, she had no complaints.

Otoscopy on both sides showed a normal tympanic membrane, and a normal middle ear on the right side. The nasal airway was open and clean. Palpation of the neck revealed no lymphoid nodules.

TABLE I

SYMPTOMS OF CRANIOPHARYNGIOMA

Headaches, nausea & vomiting
Growth retardation
Visual disturbances
Delayed puberty
Hearing impairment

From the Department of Otorhinolaryngology, Isala Klinieken Zwolle, Zwolle, The Netherlands.

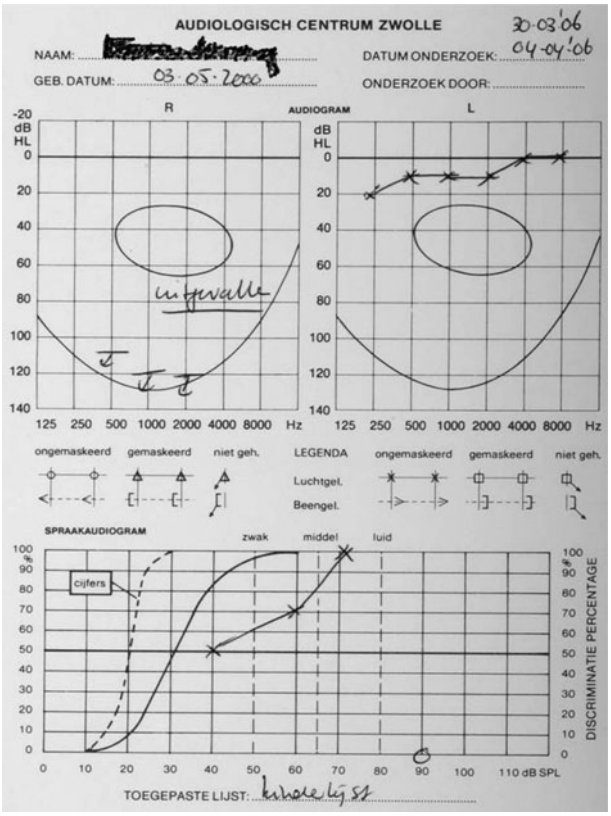

FIG. 1

Pure tone audiometry and speech audiometry on first consultation.

Comprehensive audiometry was performed. Pure tone audiometric examination revealed no response in the right ear and responses of 10–20 dB (NHL) in the left ear. Speech audiometry revealed no response in the right ear and normal responses in the left ear (Figure 1). Tympanometry showed a normal curve on both sides, with normal volume and normal pressure values. The Stenger test indicated no suspicion of aggravation. Brainstem audiometry was performed but was unsuccessful due to the patient's restlessness. Otoacoustic emissions showed normal, reproducible responses on both sides.

A detailed exploration of the patient's history revealed a paediatric consultation earlier that year, due to nausea and vomiting, headaches and abdominal discomfort. The patient's eating and swallowing had been unimpaired. The vomiting had occurred mainly in the morning. The patient's social and intellectual development had been reported as normal. The patient had experienced recent, disturbing events in her life: two of her classmates had died, one of them in a drowning accident, and her aunt and uncle had divorced during the same period. These events had made her anxious and had given her nightmares.

At that time, no abnormalities had been detected on general and neurological paediatric examination. Visual function had been unimpaired. The paediatrician had diagnosed psychosomatic vomiting due to recent emotional events. Psychosocial therapy had been given.

In the current presentation, in consideration of the patient's complete right-sided hearing loss, together with the normal OAEs and the newly discovered aspects of her previous history, we decided to perform emergency brain MRI.

The MRI scan showed a giant cystic mass arising from the cerebellum, exerting pressure on the brainstem and the ventricular system and giving rise to hydrocephalus (Figure 2). The mass was also observed to have invaded the internal acoustic canal on the right side (Figure 3).

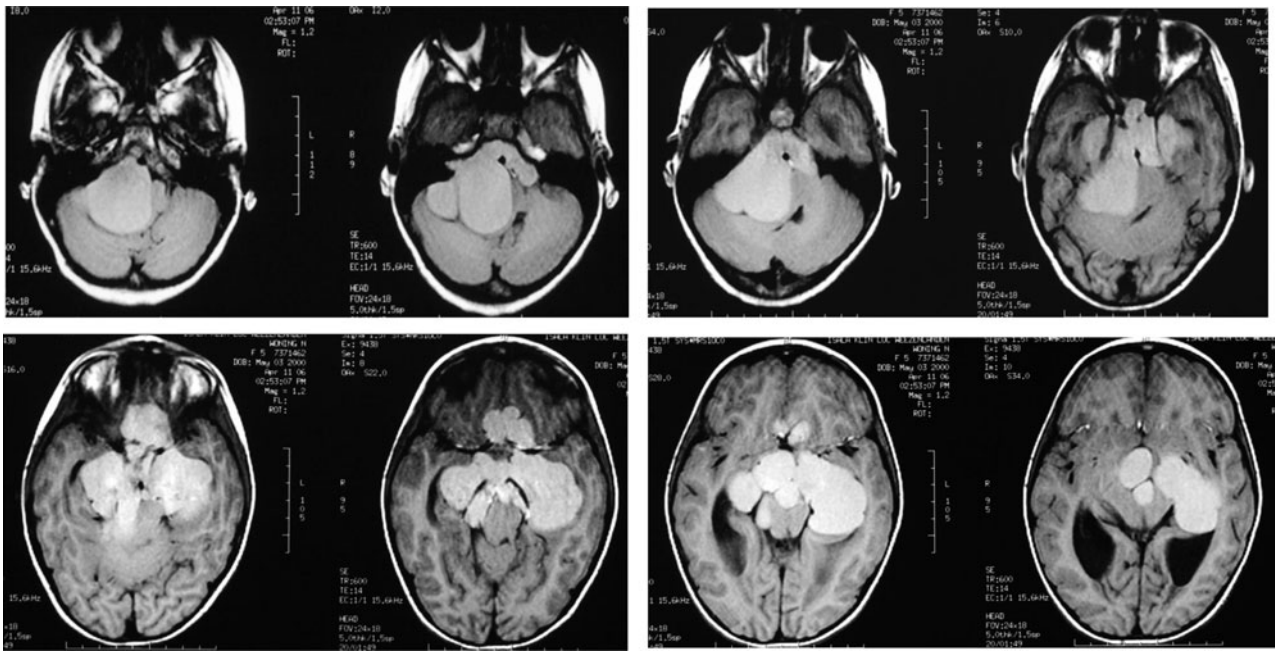

FIG. 2

Emergency axial magnetic resonance imaging scans, showing a cystic mass arising from the cerebellum, exerting pressure on the brainstem and the ventricular system, and causing hydrocephalus (see second row, last scan).

https://doi.org/10.1017/S0022215107001491 Published online by Cambridge University Press

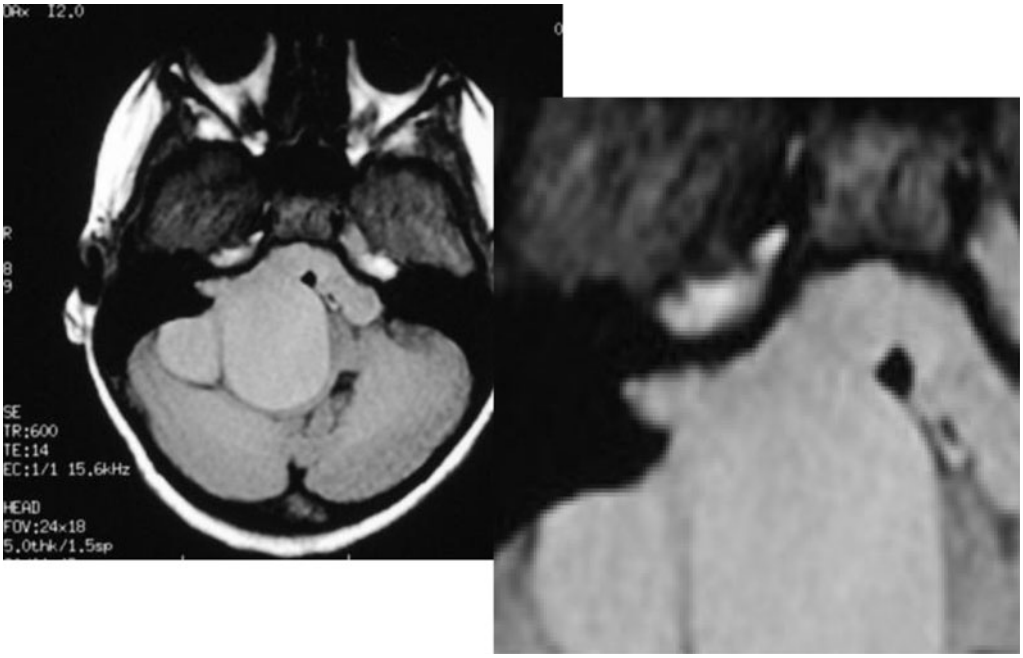

FIG. 3
Emergency axial magnetic resonance imaging scan through the internal acoustic canal; magnified image shows extension of the craniopharyngioma in the internal acoustic canal.

A neurological consultation revealed a positive Babinski reflex on the right, mild right-sided hemiparesis and mild right-sided facial nerve paresis. An ophthalmological consultation revealed dilated pupils on both sides, without visual impairment.

Dexamethasone 2 mg twice daily was immediately commenced, and the patient was immediately referred to the neurosurgeon.

Two days later, the patient underwent craniotomy, during which the whole mass was resected *in toto*. The pituitary shaft was macroscopically unaffected. Histological analysis of the specimen showed craniopharyngioma.

Post-operatively, the patient developed no neurological or endocrinological disturbances. Three days after surgery, MRI showed a small residual mass in the area of the optic chiasma (Figure 4). Hearing on the right side improved to approximately normal values (Figure 5). Obviously, the patient's otoacoustic emissions remained normal.

Discussion

Only 4 per cent of craniopharyngiomas are observed to have posterior fossa extension at initial surgery.[3] To our knowledge, hearing loss is a very rare primary symptom of craniopharyngioma. Buhl *et al.*[4] described the case of a seven-year-old boy who presented with a six-month history of headache, nausea and progressive, unilateral hearing loss. Connolly *et al.* reported three cases of children with craniopharyngioma who had initially presented with either unilateral or bilateral deafness.

A diagnosis of craniopharyngioma is made based on a combination of radiological and histopathological findings. Suprasellar calcifications and a cystic character are typical features on MRI and computed tomography.

Surgical resection by (staged) craniotomy is the primary treatment. Adjuvant radiation is indicated in cases of incomplete resection. One must consider that adjuvant radiotherapy, in addition to prior surgery, can cause neurological, endocrine or audiological morbidity, especially in young children. Overall, the five-year survival rate of craniopharyngiomas is 80 per cent.[5]

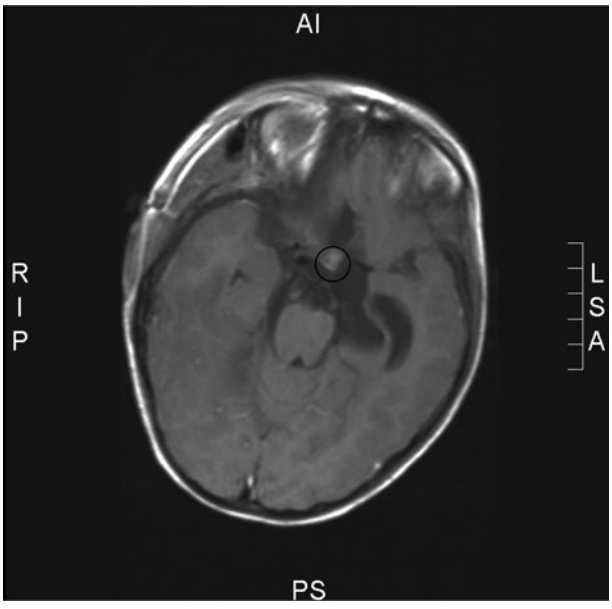

FIG. 4
Post-operative axial magnetic resonance imaging scan, showing a small residual mass in the area of the optic chiasma (circle).

https://doi.org/10.1017/S0022215107001491 Published online by Cambridge University Press

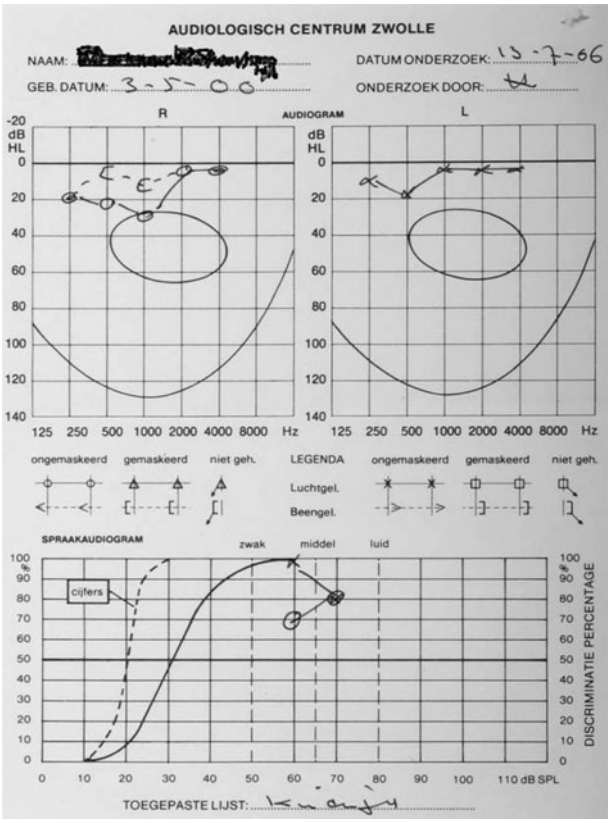

FIG. 5

Post-operative pure tone audiometry and speech audiometry; note that thresholds have fully recovered.

Shuman *et al.*[6] described a craniopharyngioma in another ENT location: the nasal cavity. An eight-year-old boy presented with nasal obstruction and snoring. Adenoidectomy failed to improve the patient's symptoms. Histopathological examination of a nasal mass revealed craniopharyngioma. There was no intracranial extension.

Otoacoustic emissions arise from the outer hair cells of the cochlea. Spontaneous emissions are present in one-third of the normal hearing population. Click-evoked OAEs are present in all normal hearing people. Assessment of OAEs is an easy, cheap and relatively non-invasive diagnostic test used for neonatal hearing screening; it is also used to differentiate between cochlear and retrocochlear pathology.

In the future, we propose to use OAEs in (young) patients with unilateral sensorineural hearing loss, in order to differentiate between cochlear and retrocochlear hearing loss. When OAEs are normal, retrocochlear pathology is suspected and MRI scanning is indicated.

References

1 Bunin GR, Surawicz TS, Witman PA, Preston-Martin S, Davis F, Bruner JM. The descriptive epidemiology of craniopharyngioma. *Neurosurg Focus* 1997;**3**:1
2 Garnett MR, Puget S, Grill J, Sainte-Rose C. Craniopharyngiomas. *Orphanet J Rare Dis* 2007;**2**:18
3 Connolly ES Jr, Winfree CJ, Carmel PW. Giant posterior fossa cystic craniopharyngiomas presenting with hearing loss. Report of three cases and review of the literature. *Surg Neurol* 1997;**47**:291–9
4 Buhl R, Lang EW, Barth H, Mehdorn HM. Giant cystic craniopharyngiomas with extension into the posterior fossa. *Childs Nerv Syst* 2000;**16**:138–42
5 Puget S, Garnett M, Wray A, Grill J, Habrand JL, Bodaert N *et al.* Pediatric craniopharyngiomas: classification and treatment according to the degree of hypothalamic involvement. *J Neurosurg* 2007;**106**(suppl 1):3–12
6 Shuman AG, Heth JA, Marentette LJ, Blaivas M, Muraszko KM. Extracranial nasopharyngeal craniopharyngioma: case report. *Neurosurgery* 2007;**60**(suppl 4):E780–1

Address for correspondence:
Dr Rutger Hofman,
Department of Otorhinolaryngology,
Hanzeplein 1,
PO Box 30001,
9700 RB Groningen, The Netherlands.

Fax: +31 50 3611698
E-mail: r.hofman@kno.umcg.nl

Dr R Hofman takes responsibility for the integrity of the content of the paper.
Competing interests: None declared