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Radiology in Focus

Case report: greatly enlarged jugular fossa with progressive sensorineural hearing loss

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Abstract
A case of a five-year-old child with a greatly enlarged jugular fossa and high jugular bulb with associated progressive sensorineural hearing loss is presented. While various forms of this anatomical variant have been described by many authors, this is an extreme example, and progressive symptoms are most unusual.

Key words: Hearing loss, sensorineural; Child; Jugular vein

Case report

A five-year-old female child presented to the Royal National Throat, Nose and Ear Hospital with a history of progressive hearing loss and occasional tinnitus involving the right ear. She had been born by caesarean section at 35 weeks gestation after an uneventful antenatal course. Her neonatal period and infancy were normal, with normal developmental milestones. There was a vague family history of mild unilateral hearing loss involving the maternal father and brother, but no objective details could be obtained. Her mother first suspected that her child had reduced hearing, when she was three years old, and it was at this stage, that she initially presented. She was assessed as normal on impedance audiometry. She returned for follow-up five months later owing to continued worries by her mother about the hearing loss. Clinical examination and audiometry were normal, both at this stage, and at a subsequent two-month follow-up. Three months later however, audiometry showed low frequency sensorineural hearing loss on the right side (0.25 kHz down to 60 dB and 500 kHz down to 50 dB). Hearing on the left side remained normal. Since this time, progressive right-sided low and mid-range frequency hearing loss of a further 25 dB has occurred.

Computerized tomography (CT) scanning with high resolution images of the temporal bones revealed a massively enlarged right jugular fossa (Figure 1). The bony margins of the vestibule and posterior semicircular canal could not be clearly defined, and raised the suspicion of bony erosion. The right internal auditory meatus appeared normal, as did the cochlea. The left jugular fossa and inner ear structures appeared normal. On account of the continued progressive symptoms and possible bony erosion on the right side, magnetic resonance imaging (MRI) with magnetic resonance angiography (MRA) was performed. On axial and coronal

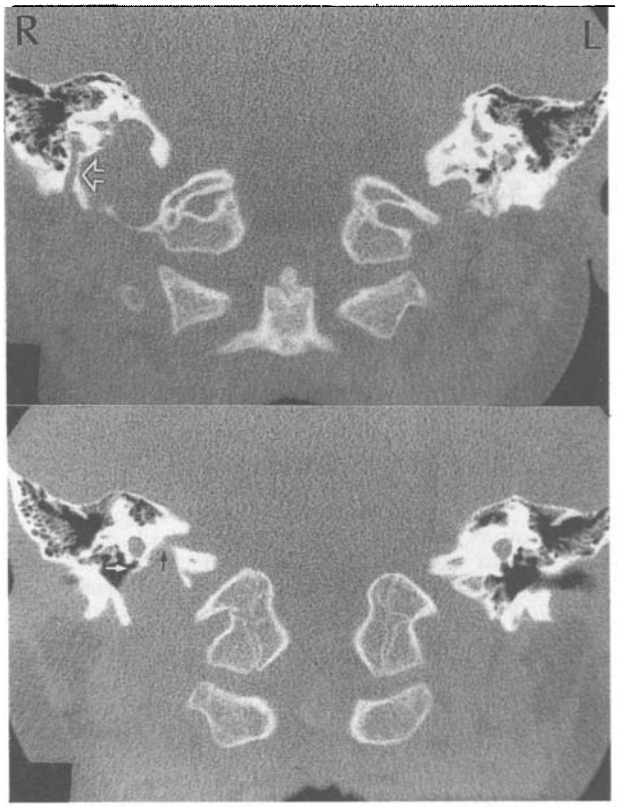

Fig. 1.

Coronal CT images through the petrous bones demonstrating the greatly enlarged right high jugular bulb and diverticulum. Note the close relationship of the right jugular bulb with the descending portion of the facial nerve canal (open white arrow). The right high jugular bulb and diverticulum (black arrow) is seen extending above the level of the round window (white arrow.) Note the apparent erosion into the right vestibule.

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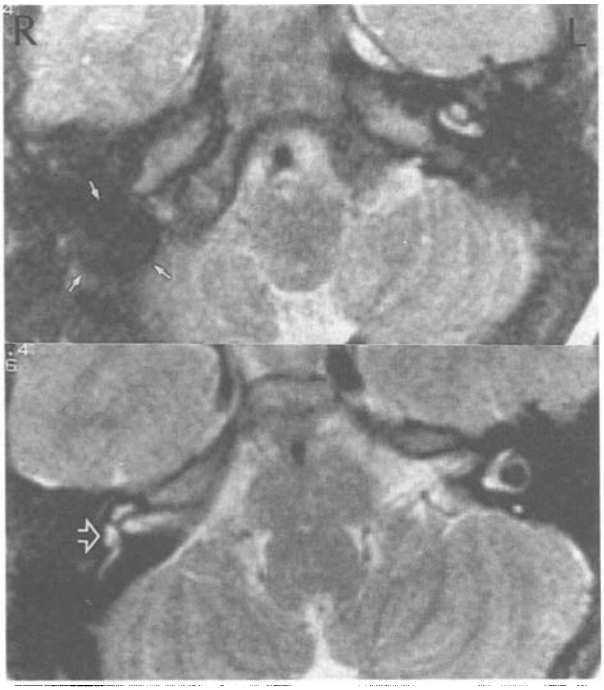

FIG. 2.

Axial fast spin echo (FSE) T_2-weighted images through the petrous bones demonstrating signal void in the region of the right jugular fossa (small white arrows). Note the normal right vestibule (open white arrow) and posterior semicircular canal.

fast spin echo (FSE) T_2-weighted images (Figure 2), the right vestibule and posterior semicircular canal were well visualized and appeared intact, and in particular, were not encroached upon by the enlarged jugular fossa. The right cochlea and internal auditory meatus (IAM) appeared normal, as did the left-sided inner ear structures. MRA clearly demonstrated a massively enlarged sigmoid sinus and high jugular bulb (Figure 3), with an associated

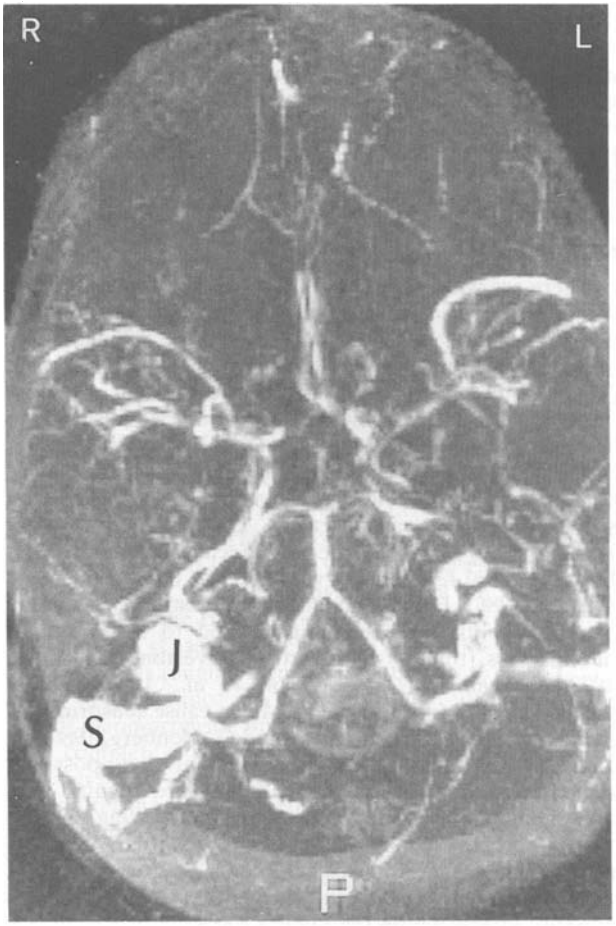

FIG. 3.

MRA demonstrating the greatly enlarged right-sided sigmoid sinus (S) and jugular bulb (J). Note the significantly decreased venous return down the left side.

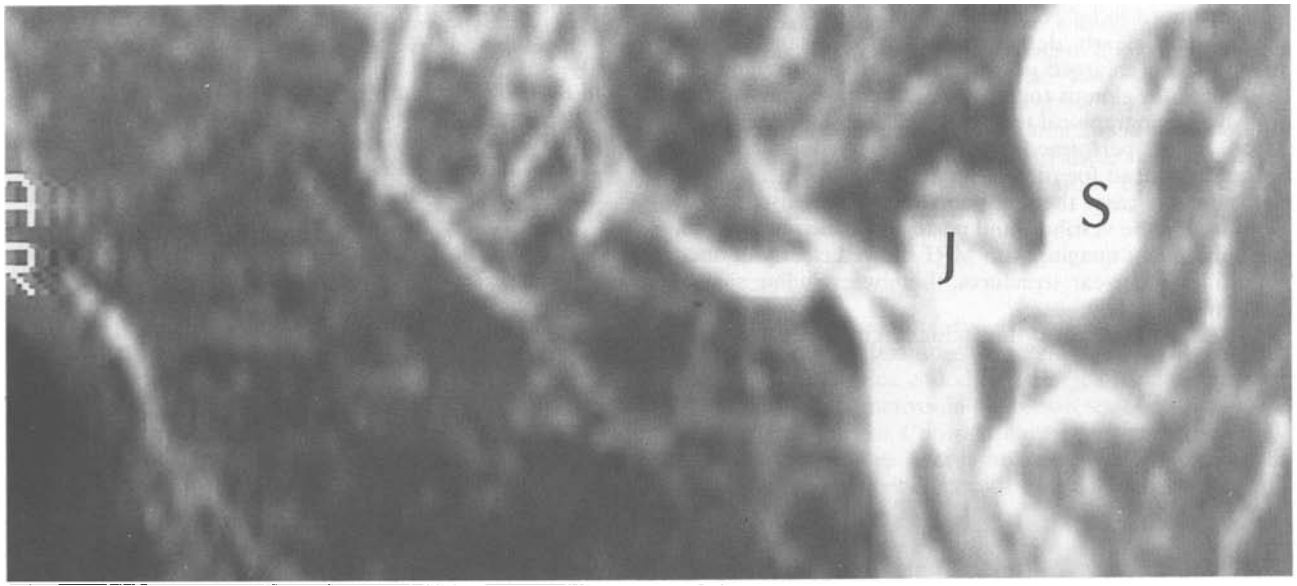

FIG. 4.

MRA oblique saggital view of the right-sided sigmoid sinus (S) and jugular bulb (J). Note the turbulent flow within the jugular bulb diverticulum.

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diverticulum. Turbulent flow was noted within the jugular bulb diverticulum (Figure 4). Decreased venous return was noted down the left side. There was no evidence of a glomus tumour nor arteriovenous inflammation. It is worth mentioning that the child lay still for the entire examination, approximately nine minutes, without requiring sedation.

Discussion

There are wide variations in the size, shape and position of jugular fossae and associated jugular bulbs in the general population. Extensive work on the radioanatomy of temporal bones has revealed that high jugular fossae (situated above the lower border of the round window), occur in approximately 24 per cent of individuals and are commonly associated with a diverticulum of the jugular bulb (Wadin, 1986). These high jugular fossae occur more commonly on the right side and may encroach on surrounding inner ear structures. The cochlear aqueduct is the structure most frequently affected by a high fossa, but no clinical relationship with this has been established (Wadin, 1986). Encroachment on the vestibular aqueduct has been both reported, and related to sensorineural hearing loss, tinnitus and vertigo (Overton and Ritter, 1973; Graham, 1977; Bergeron *et al.*, 1991). Direct intrusion of the fossa on the IAM can cause sensorineural hearing loss and tinnitus (Stern and Goldenberg, 1980; Lo and Solti-Bohman, 1984). Erosion of the posterior semicircular canal by a medially directed jugular bulb diverticulum with associated vertigo and hearing loss has been reported (Phelps and Lloyd, 1990). The exact mechanism of causation of symptoms is often not clear, as in this case, but may in part be due to turbulent flow and pressure effects within the jugular fossa.

Summary

The following important points arise from this case:

(1) The findings are an extreme example of a normal variant. Progressive symptoms are unusual in such cases, and underlying vascular tumours, in particular, glomus tumours, need to be excluded.

(2) MRA clearly demonstrates the vascular anatomy related to an enlarged jugular fossa and can exclude the presence of a glomus tumour, thereby preventing further invasive angiographical techniques. It may be possible for MRA to be performed in cooperative young children without the need for routine sedation.

(3) In this case, the CT scans raised the suspicion of erosion into the vestibule and posterior semicircular canal, but subsequent imaging with MRI showed clearly defined fluid-filled inner ear structures, thereby excluding significant encroachment.

(4) Management in this patient continues to be a dilemma. Her hearing continues to diminish on the right side, but surgical intervention is high risk. The management plan in this case is continued close follow-up in the Department of Audiological Medicine.

Children with unilateral hearing loss are known to be at risk for reduced attention, poor localization skills and slight deficits in their cognitive development. If there are no difficulties in any of these areas and if this child's hearing for speech is good, then there will be no need to aid the abnormal ear. If however she develops problems with her learning skills and school progress, then a BE101 type aid with a vented mould will be considered. The aim would be to boost her own hearing without boosting the background noise.

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