CONTINUING EDUCATION PROGRAM: FOCUS...

The vestibulocochlear nerve (VIII)

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KEYWORDS
Crani al nerves; Pathology; Vestibulocochlear nerve (VIII)

Abstract The vestibulocochlear nerve (8th cranial nerve) is a sensory nerve. It is made up of two nerves, the cochlear, which transmits sound and the vestibular which controls balance. It is an intracranial nerve which runs from the sensory receptors in the internal ear to the brain stem nuclei and finally to the auditory areas: the post-central gyrus and superior temporal auditory cortex. The most common lesions responsible for damage to VIII are vestibular Schwannomas. This report reviews the anatomy and various investigations of the nerve.
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The cochlear nerve

Review of anatomy [1]

The cochlear nerve has a peripheral sensory origin and follows a centripetal path. It originates in the cochlear membrane sensory canal, forming the spiral organ (the organ of Corti) and lies on the basilar membrane (Fig. 1). The neuronal fibers of the protoneuron connect to the ciliated cells on the spiral lamina (Fig. 2). The axons are grouped together along the axis of the cochlea (modiolus), forming the cochlear nerve which then enters the internal auditory meatus (IAM). Within the internal auditory meatus, the nerve joins the vestibular nerve to form the vestibulocochlear nerve which crosses the cerebellopontine angle (Figs. 3 and 4) where the cochlear nerve lies postero-laterally to the vestibular nerve. It runs close to the VII nerve, internal auditory artery and the superior lateral cerebellar vein (the vein of Dandy). In the brain stem, the nerve enters the neuraxis in the most lateral part of the bulbopontine groove. The fibers reach two nuclei in the brain stem, the anterior or ventral nucleus and the posterior or dorsal nucleus.

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2211-5684/$ – see front matter © 2013 Published by Elsevier Masson SAS on behalf of the Éditions françaises de radiologie.
http://dx.doi.org/10.1016/j.diii.2013.08.015
Figure 1. Section through the cochlear canal: scala vestibuli anteriorly, scala tympani posteriorly, spiral lamina with the organ of Corti.

Figure 2. Ciliated cells with the cochlear nerve.

Figure 3. High resolution axial T2 MRI: vestibulocochlear nerve in the CPA with division in the internal auditory meatus into the cochlear, anterolateral and posterior vestibular nerves.

Figure 4. T2 weighted axial CT perpendicular to the axis of the nerve: above anteriorly: facial nerve, below anteriorly: the largest, cochlear nerve, above posteriorly: superior vestibular nerve, below posteriorly: inferior vestibular nerve.

Figure 5. Vocal audiogram showing reduced hearing when listening to words.

Clinical features of the cochlear nerve

Damage to the cochlear nerve may be due to a sensory reception organ lesion, causing sensory or endocochlear deafness, or if the damage affects the nerve or central cochlear pathways when the deafness is described as retro-cochlear sensory deafness.

Several tests are often used to investigate deafness:
• vocal audiometry, which involves assessing a person’s understanding of words and different thresholds (Fig. 5);
• tonal audiometry, which is a technique that studies bone (BC) and air (AC) conduction. In people with normal hearing, the BC is the same as the AC and sensory deafness, both curves are reduced (Fig. 6);
• auditory evoked potentials (AEP), which measure electrical conduction in the auditory nerve running the cortex (Fig. 7).

The vestibular nerve

Review of anatomy

The vestibular pathways are a complex, multisensory reflex system belonging to the subconscious balance control system linked to the cerebellum, and the conscious balance control...
The vestibulocochlear nerve (VIII)

Clinical aspects of the vestibular nerve

Damage to the vestibular nerve results in vertigo, a balance disorder, and nystagmus. Four questions should be asked in acute vertigo:

- Is it an emergency?
- Is it true vertigo?
- What is the topography of the lesion?
- What are the consequences of the vertigo and balance problems?

Practitioners should look for any history and cardiovascular risk factors, particularly in the elderly, and for a sudden onset associated with headache and possible concomitant symptoms (motor or sensory problems, swallowing difficulties, etc.) which suggest a possible cerebrovascular accident, which is an absolute emergency as the first hours are essential in order to protect neuronal function.

Clinical examination should look routinely for vertical nystagmus present on direct gaze and for neurological signs, particularly postural deviation (deviation of the index fingers, positive Romberg or Fukuda sign), cerebellar symptoms or involvement of other cranial nerves which may suggest the Wallenberg syndrome or cerebellar infarction.

Clinical examination can be combined with otoscopy and investigation for spontaneous or provoked nystagmus with video-nystagmoscopy.

Any spontaneous nystagmus is pathological. Horizontal nystagmus which reduces with fixed gaze reflects peripheral damage and vertical nystagmus increased on fixed gaze reflects central damage.

The role of imaging in cochlear and vestibular nerve investigations [2]

Which investigations to perform?

The first line investigation for sensory deafness or acute vertigo is MRI, which examines the labyrinth, base of the cranium and brain.

The protocol should include sequences visualizing the various auditory and vestibular anatomical structures and the encephalic neuronal connections. Flair encephalic MRI, diffusion weighted, and high resolution T2 weighted (DRIVE, CISS, FIESTA) on the IAM and anterior and posterior labyrinth, and T1 weighted sequences with and without enhancement, should be performed.

Unenhanced CT is useful to examine the bony structures of the labyrinth or if MRI is contraindicated.

Imaging results in various diseases

Endocochlear deafness [3]

Four types of disease may occur:

- Congenital developmental abnormalities of the labyrinth, either in isolation or associated with multiple malformations causing congenital deafness. This may involve an abnormality of the anterior labyrinth (cochlear abnormality, dilation of the modiolus with Geyser ear), or the posterior labyrinth, the most common being dilatation of the vestibular aqueduct, although labyrinthine
dysmorphic features may be present. Hypoplasia or complete aplasia of the cochlear nerve may also occur;

• acquired disease of the labyrinth, either following injury or infection, with ossifying or autoimmune labyrinthitis (Fig. 8);

• dysplastic bone lesions as a result of damage to the otic capsule (Paget, Lobstein); unenhanced CT is the key investigation to investigate these conditions, which will not be discussed in detail in this article as it is not really its purpose.

• neoplastic disease, an intra-labyrinthine Schwannoma, which we will discuss in the next section.

Retrocochlear deafness [4]

Extra-axial lesions
There is one clearly predominant cause, the Schwannoma (Fig. 9a and b). This is the commonest space-occupying lesion of the IAM.

It is typically a lesion presenting in middle age (after the age of 50 years old). The clinical symptoms are sensory hypoacusis (95% of cases), tinnitus (65% of cases), and vertigo (46% of cases). Sudden onset deafness occasionally occurs. The diagnosis on imaging is based on morphological criteria, appearing as a rounded or ovular lesion with a clearly demarcated regular outline located in the cerebellopontine angle and in the IAM with anterior extension demarcated by the facial nerve. In 84.8% of cases, the tumor develops on the inferior vestibular nerve and therefore is located posteriorly. MRI is also a key part of a diagnosis, the mass being isointense on T1 weighted images, invariably hypointense on high resolution T2 weighted images, and enhanced with gadolinium.

The second most common lesion is a meningioma (Fig. 10a and b) which makes up 10% of cerebellopontine angle tumors and 10% of all intracranial meningiomas [5]. The average age at diagnosis is 45 to 50 years old with a clear female predominance (female to male sex-ratio: 4 to 1). It has various clinical signs which are relatively non-specific and may include vestibular symptoms, facial hypoesthesia or headache. Morphological criteria play a key part in the diagnosis. The tumor is extra-axial, rising from a wide dural base with obtuse attachment angles to the dura mater and associated with bone involvement (usually bony consolidation). Meningioma appearances vary. They are isointense on T1 weighted MRI in the cortex and the T2 weighted image depends on their histological type. Meningothelial and angioblastic meningiomas are hyperintense whereas transitional and fibroblastic meningiomas are iso- or hypointense with the cortex. Meningiomas enhance strongly with contrast. The adjacent dura mater also enhances (the English term for this is the dural tail sign).

Epidermoid cysts are non-neoplastic congenital tumors, responsible for 1% of intracranial tumors. The average age at diagnosis is in the 4th to 5th decade. Clinical features are not characteristic. Symptoms may include mild headaches. Facial sensory abnormalities, due to extension of the tumor along the trigeminal nerve and into the cavum trigeminae are often present. Imaging shows an extra-axial tumor with a clear, irregular polyhedral outline. It is often large when it is discovered. It surrounds rather than displaces the vascular and neuronal structures and is similar in appearance to cerebrospinal fluid (CSF), (hypointense on T1 weighted and hyperintense on T2 weighted sequences, not enhancing with contrast). It is often, however, rather heterogeneous in appearance and slightly “mottled”. It is straightforward, however, using appropriate sequences, to differentiate an epidermoid cyst from a “fluid” structure.

Figure 8. Infectious labyrinthitis. High resolution axial T2 weighted MRI: abnormal signal from the left labyrinth fluid with amputation of the lateral superior semi-circular canal.

Figure 9. a: Schwannoma within the meatus extending to the base of the canal; b: grade 3 CPA Schwannoma extending to the internal auditory meatus. Posterior extension is limited anteriorly by the VII nerve.
such as an arachnoid cyst. Diffusion weighted MRI is the fastest sequence, in which an epidermoid cyst is hyperintense whereas an arachnoid cyst produces the same very hypointense appearances as those of cerebrospinal fluid.

High resolution T2 weighted sequences, which have “binary” contrast (fluid is hyperintense and other structures are hypointense) can also be used to distinguish between an epidermoid cyst which is relatively hypointense compared to the CSF and an arachnoid cyst, which is markedly hyperintense.

Other rarer lesions of the cerebellopontine angle and IAM are seen. Vascular lesions include aneurysms, vascular loops and a dural fistula. These abnormalities are usually hypointense on T2 weighted sequences and, when they are suspected, require additional TOF MR angiography. Lipomas are fatty tumors which develop from the same pluripotent cells which give rise to the meninges (Fig. 11a and b). Lipomas are closely connected to the leptomeninges surrounding the cranial nerves. They often have relatively minor symptoms. They are very straightforward to diagnose on imaging when an unenhanced T1 weighted sequence is performed routinely before using contrast. The diagnosis of lipoma is confirmed on a fat saturation sequence from an IAM mass which is intense on an unenhanced fat saturation sequence. Meningeal diseases [6] include viral neuritis which is diagnosed from enhancement of the arachnoid sheath with normal appearance of the nerves in high resolution T2 weighted images and inflammatory neuroborreliosis or sarcoidosis lesions [7]. Neuroborreliosis is a zoonosis transmitted by tick bites. Diffuse meningeal involvement of the cranial nerves combined with leptomeningeal enhancement in the roots of the cauda equina suggest a diagnosis of neuroborreliosis, which should be confirmed by Lyme’s disease serology (Fig. 12a and b). Rare tumors include meningeal carcinomatosis (the meninges are thickened with multifocal enhancement and undulating outline), tumors of the endolymphatic sac, which are seen particularly in patients suffering from Von Hippel Lindau’s disease. These are hypervascular lesions which enhance and contain cystic and hemorrhagic components. Inter-axial tumors are also found embedded in the CPA (ependymomas and choroidplexus papillomas).

**Intra-axial lesions**
These must be remembered and looked for routinely. Multiple sclerosis (MS) may present initially with vertigo (5 to 7% of cases). MRI is used to investigate for disseminated central nervous system involvement, with supratentorial white matter lesions. MRI should be interpreted along with the other clinical and laboratory findings.
Ischemic bone stem and posterior fossa disease is usually secondary to obstruction of the vertebral artery or posterior inferior cerebellar artery (PICA). It can also be due to vertebral artery dissection, usually following injury. Vertigo is rarely seen in isolation and is often associated with other neurological signs. This is a neurovascular emergency. MRI shows a hyperintense diffusion weighted area with a fall in ADC over the territory supplied by an artery (Fig. 13a and b). Vascular sequences (TOF MR, neck vessel MR angiography) show vertebral artery stenosis or dissection.

Figure 12. Leptomeningeal enhancement of the acousto-facial bundles (a) and trigeminal bundles (b).

Figure 13. Axial T2 weighted (a) and diffusion weighted (b) MRI. Ischemic accident in the right bulbar olive as part of Wallenberg's syndrome with dissection of the right vertebral artery (flow hypointensity not seen on the axial T2 weighted sequence).
Conclusion

MRI is the primary investigation for sensory deafness or central vertigo, either in isolation or associated with other clinical signs. It may be combined with CT to investigate the bony labyrinth and base of the cranium. Sequences should image the different anatomical structures of the vestibulocochlear system (internal ear, IAM and encephalic structures). Clinical features are important (history, endo- or retrocochlear disease, or concomitant neurological signs). A full examination of the head and occipito-cervical junction is needed because of the many causes and relatively non-specific clinical presentation.

TAKE-HOME MESSAGES

- Sudden onset vertigo associated with headaches or neurological symptoms is an emergency and requires MRI.
- Any spontaneous nystagmus is pathological.
- Horizontal nystagmus reduced on fixed gaze is peripheral in origin.
- Vertical nystagmus increased on fixed gaze is central in origin.
- Investigations for sensory deafness should look for disease of the sensory reception organ itself (endocochlear sensory deafness) or damage to the nerve or central cochlear pathways (retrocochlear sensory deafness).
- MRI is the 1st line investigation. Sequences should image the different anatomical structures of the vestibulocochlear system.
- Clinical history, past medical history and clinical signs provide a guide to the diagnosis.

Clinical case

A 24-year-old patient is being followed up for a posterior fossa arachnoid cyst, displacing the cerebellum. A shunt catheter has been inserted. Immediately postoperatively,

the patient developed sudden onset isolated vertigo with left cophosis.

Questions

1. What investigation would you perform?
2. What are you looking for?
3. A CT is normal. Would you add further imaging?
4. Describe the imaging appearances. Does this explain the clinical features? (Figs. 14 and 15).

Answers

1. A CT scan.
2. The image is examined for a hematoma or hemorrhage within the cyst.
3. Yes, a posterior fossa MRI would be performed.
4. The shunt catheter is crossing the wall of the cyst and extends to the IAM, compressing the acoustic-facial bundle, explaining the symptoms due to irritation of the vestibulocochlear nerve.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


