REVIEW ARTICLE

Imaging Diagnostics: Congenital Malformations and Acquired Lesions of the Inner Ear

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Abstract

Introduction: Congenital malformations and acquired lesions of the inner ear are characterised by small structural changes in this region. In recent decades, treatment options have improved considerably. At the same time, there has been a great advancement in diagnostic methods, obtaining high-resolution labyrinth images.

Currently, we use a 64-multislice computed tomography scanner in spiral mode (Brillance 64 Phillips, Eindhoven, the Netherlands), with an overlap of 0.66 mm and an interval of 0.33 mm, 120 kV and 300 mA. The magnetic resonance images were taken with Signa HDxt 1.5 and 3.0 T units (GE Healthcare, Waukesha, WI, USA).

We reviewed the radiological features of the lesions affecting the inner ear. They are classified as congenital (labyrinth malformation and statoacoustic nerve deficiencies) or acquired (otospongiosis, labyrinthitis, Ménière’s disease, inner ear haemorrhage, intralabyrinthine schwannoma and endolymphatic sac tumour).

Conclusion: Magnetic resonance imaging and computed tomography play an essential role in diagnosing patients with inner ear pathology. The technique selected should be chosen depending on the clinical setting. In a generic way, tomography is the method of choice for the study of traumatic pathology or otospongiosis. When tumour or inflammatory pathology is suspected, magnetic resonance is superior. In cases of congenital malformation, both techniques are complementary.

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Introduction

Congenital malformations and acquired lesions of the inner ear (IE) are characterised by small structural changes. Treatment options in this specific region have improved considerably in recent decades. At the same time, there has been great advancement in diagnostic methods, and it is currently possible to obtain high-resolution images of the labyrinth and the statoacoustic nerves.

Computed tomography (CT) and magnetic resonance (MR) imaging of the temporal bone enable excellent anatomical definition of the inner ear and are used in daily practice to study its pathology. Computed tomography is the method of choice for studying the bony structures of the ear, whereas magnetic resonance imaging provides images of both the membranous labyrinth and the cranial nerves.

With the aid of both diagnostic techniques, the radiologist can provide referring ENT specialists with the necessary information to enable a correct treatment approach. We consider that interdisciplinary collaboration is necessary and that all parties should have extensive knowledge of disorders of the inner ear and their diagnosis through imaging.

In this article, we shall describe and illustrate the radiological characteristics of the most common congenital and acquired diseases of the IE, showing the keys to correctly interpreting them on radiological images.

Technique

In general CT images were taken with a thickness of 0.625–1 mm, and reconstructed using a bone algorithm. Currently, we use a 64-multislice CT scanner (Brilliance 64 Phillips, Eindhoven, the Netherlands), with an overlap of 0.66 mm and an interval of 0.33 mm, 120 kV and 300 mA. The magnetic resonance images were taken with Signa HDxt 1.5 and 3.0 T (GE Healthcare, Waukesha, WI, USA).

Radiological Characteristics

The lesions found were classified according to their origin as congenital (labyrinth malformation and statoacoustic nerve deficiencies) or acquired (otospongiosis, labyrinthitis, Ménière’s disease, intralabyrinthine schwannoma and endolymphatic sac tumour).

Congenital Malformations of the Inner Ear

Congenital malformations are defined as variations in the normal anatomical development of the inner ear, which result in functional disorders of the inner ear. The bony labyrinth develops between the fourth and eighth weeks of gestation; from the eighth week, it grows and ossifies. The structural malformations which can be diagnosed using radiological imaging of the inner ear are due to defects forming between the fourth and eighth weeks, whereas subsequent lesions affect the sensory epithelium and cannot be imaged. This is why in long-time series studies only 20% of patients with congenital hearing loss have malformations which are visible radiologically.

Diagnóstico por imagen: malformaciones congénitas y lesiones adquiridas del oído interno

Resumen

Introducción: Las malformaciones congénitas y las lesiones adquiridas del oído interno se caracterizan por pequeños cambios estructurales de esta región. En las últimas décadas, las opciones terapéuticas han mejorado considerablemente, y paralelamente se ha producido un gran avance en los métodos diagnósticos, consiguiendo imágenes de alta resolución del laberinto.

Actualmente se utiliza una tomografía computerizada multicorte de 64 detectores (Brilliance 64 Phillips, Eindhoven, los Países Bajos), un espesor de adquisición de 0,66 y un intervalo de 0,33 mm, 120 kV y 300 mA. Las imágenes de resonancia magnética proceden de los equipos Signa HDxt 1,5 y 3,0 T (GE Healthcare, Waukesha, WI, USA).

Se realiza una revisión de las características radiológicas de las lesiones que afectan al oído interno que son clasificadas según su origen en congénitas (malformaciones del laberinto y deficiencias de los nervios statoacústicos) o adquiridas (otoespongiosis, laberintitis, hemorragia del oído interno, enfermedad de Menière, schwannoma intralabirintico, tumor del saco endolinfático).

Conclusiones: La resonancia magnética y la tomografía computerizada juegan un papel fundamental en el diagnóstico de pacientes con patología del oído interno. La técnica de elección debe ser escogida en función del escenario clínico. De forma genérica, para el estudio de patología traumática u otoespongiosis la tomografía es el método de elección. Ante sospecha de patología tumoral o inflamatoria la resonancia se muestra superior. Para el estudio de patología malformativa ambas técnicas son complementarias.

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Cochlear Aplasia/Hypoplasia

Aplasia is a rare malformation, constituting only 3% of cochlear malformations. Embryonic development is arrested at the end of the third week of gestation and it is usually of unknown etiology. Cochlear hypoplasia constitutes 15% of cochlear malformations and is due to an anomaly in the development of the cochlear duct during the sixth week of gestation.

Patients present with unilateral or bilateral sensorineural hearing loss from birth. Both are diagnosed using imaging tests. Cochlear absence is confirmed in cochlear aplasia. The vestibule and semicircular canals (SCC) are often malformed, globular or dilated and the cochlear nerve and its duct are missing (Fig. 1A). It is important to differentiate aplasia with cystic vestibule from cystic cochleovestibular anomaly (CCVA), since cochlear implants (CI) are contraindicated for aplasia but not for CCVA, the only possible treatment being brainstem implant. It is also important to differentiate this from labyrinthitis ossificans, in which the bulge of the promontory caused by the basal turn of the cochlea can be seen: this characteristic is not present in cochlear aplasia. In cochlear hypoplasia, a small cochlea is observed (1-3 mm) with a single primitive turn. The vestibule and the semicircular canals can be normal or they can be malformed (Fig. 1A).

Malformation of the Vestibule and Semicircular Canals

The development of the semicircular canals starts at the sixth week of gestation and ends at the 22nd week. There are dysplasias which only affect the development of SCCs which are shorter and wider. Other malformations affect the vestibule and the SCCs, but the most common presents with an enlarged globular vestibule and dilated lateral SCC, the last to form (Fig. 1B). Aplasia of the SCCs is less common than dysplasia. It is usually associated with atresia of the oval window and an abnormal trajectory of the facial nerve. In CHARGE syndrome (Coloboma, Heart anomalies, Retarded growth and development, Genital hypoplasia, Ear anomalies) (Fig. 1B) all the SCCs can be absent.

They present clinically with vestibular alteration and can cause different degrees of hearing loss depending on their association with cochlear malformations. In both cases, the diagnostic method of choice is CT.

Deficiency of the Cochlear Nerve

In the majority of cases this is a pathology acquired in the womb or the neonatal period. In cases where a nerve has not existed (malformation at 7 weeks' gestation) the duct will not develop either.

In cases where the nerve has not developed, CT can show atresia or stenosis of variable degree of the cochlear nerve duct, 1.4 mm being considered its normal minimum diameter. As we have already mentioned, most cases are acquired in the womb and the neonatal period; therefore, the diameter of the bony conduit and the inner auditory canal (IAC) are normal, and have low sensitivity for diagnosis. Magnetic resonance imaging is the technique of choice to show deficiencies of the cochlear nerve as it can be visualised directly along its entire length and relative...
measurements can be obtained, in particular of the facial nerve (Fig. 1C).

This pathology should be suspected in cases of profound sensorineural hearing loss, profound deafness in patients with retarded growth and development, CHARGE syndrome, auditory neuropathy spectrum disorder, sensorineural hearing loss acquired in the neonatal period, or in cases of unilateral sensorineural hearing loss.¹¹

Incomplete Partition—Type I (IP-I)/Cystic Cochleovestibular Anomaly
IP-I is a malformation which starts at 5 weeks’ gestation and affects the development of the inner cochlear structure. There are different grades of anomaly. At its mildest, the malformation is confined to the cochlea, with normal vestibule and SCCs, at this grade there is no interscalar septum and the modiolus is absent (Fig. 2A₁).⁵ At its most severe, the cochlea, the vestibule and the horizontal SCC are globularly enlarged and make a single cavity which typically forms a figure-eight contour.⁵,¹² This dysplasia is known as cystic cochleovestibular anomaly (Fig. 2A₂). These findings are confirmed by CT. During inner ear surgery there is a greater risk of meningitis or perilymphatic fistula due to the characteristic dilatation of the inner auditory canal.⁵,⁶

Dilated Vestibular Aqueduct Syndrome
This is the most common pathology detected by imaging in patients with progressive sensorineural hearing loss since childhood.¹³ The malformation is usually bilateral in 90% of cases and is more prevalent in females.

Diagnosis is established by the association of an appropriate clinical symptom context (bilateral sensorineural hearing loss) and characteristic radiological findings. With this malformation there is widening of the vestibular aqueduct and dilatation of the endolymphatic sac can be associated. Computed tomography shows a dilated bony vestibular aqueduct of a calibre exceeding 1.5 mm at the midpoint of its route between the common crura and its external opening.¹⁴ This malformation can be associated with a dilated endolymphatic sac shown on MR, and its projection outside the bone into the posterior fossa (Fig. 2B₁,₂).¹⁵

In 84% of cases, this malformation tends to be accompanied by another inner ear anomaly. The most common cochlear malformation is type II incomplete partition. It is also commonly associated with Prended syndrome which presents with sensorineural hearing loss and goitre.⁸

Incomplete Partition—Type II (IP-II)
Also known as Mondini’s anomaly. This represents 30% of congenital malformations of the IE. There is fusion of the middle and apical turns of the cochlea, with absence of interscalar septum, whereas the basal turn is present.¹⁶ It is a result of arrested development of the ear during the 6th and 7th week of gestation; the cause is genetic or due to infectious or toxic embryopathy.

Patients characteristically present with severe hearing deficit at low frequencies due to apical malformation of the cochlea, whereas high frequencies tend to be preserved. However, other authors such as Sancho Serrano et al., confirm that there is no parallelism between histopathological lesions and hearing deficit.¹⁶

Diagnosis is made using CT (Fig. 2B₂). This type of malformation can be associated with anomalies in the formation of the vestibule and semicircular canals.
Inflammatory Lesions

Inflammatory lesions of the IE may be subdivided into four main categories. Infections, neoplasms, and vascular malformations are the most common. Infections can result from viral, bacterial, or fungal pathogens, and they can present as acute or chronic inflammatory processes. Neoplasms, on the other hand, can be either benign or malignant and may present with a wide range of symptoms. Vascular malformations are another category of inflammatory lesions, and they can be divided into two main types: arteriovenous malformations and venous malformations. These lesions can present with a range of symptoms, including bleeding, pain, and swelling.

Acquired Lesions of the Inner Ear

Acquired lesions of the IE can be subdivided into three types: inflammatory, traumatic, and tumour-related.

Inflammatory Lesions

Inflammatory lesions of the IE can be caused by a variety of factors, including viral, bacterial, and fungal infections. These lesions can present with a wide range of symptoms, including pain, fever, and hearing loss.

Traumatic Lesions

Traumatic lesions of the IE can be caused by a variety of factors, including blunt or penetrating trauma. These lesions can present with a wide range of symptoms, including pain, hearing loss, and dizziness.

Tumour-Related Lesions

Tumour-related lesions of the IE can be caused by a variety of factors, including malignant and benign tumours. These lesions can present with a wide range of symptoms, including hearing loss, dizziness, and tinnitus.

Otospongiosis

This is a condition characterised by the presence of bone absorption foci and is usually associated with hearing loss.

Labyrinthitis

Labyrinthitis is a condition characterised by inflammation of the inner ear. It can be caused by a variety of factors, including viral, bacterial, and fungal infections. These lesions can present with a wide range of symptoms, including hearing loss, dizziness, and tinnitus.
Figure 4  Axial CT of the ears of different patients showing the four radiological grades of otospongiosis. The arrows show the lytic foci of spongy bone. (1a) Isolated lesion of the footplate. (1b) Spongiotic focus in fissula ante fenestram less than 1 mm. (2) Otospongiotic focus above 1 mm not in contact with the middle turn of the cochlear lumen. (3) Spongiotic focus in contact with the cochlear lumen. (4) Confluent otospongiosis around the cochlea.

is characteristic (Fig. 5A). In T2 weighted MR images, there is a decrease in the intensity of the signal from the membranous labyrinth. Magnetic resonance findings in the initial non-bony forms of cochlear obliteration are very relevant with regard to cochlear implants, especially in postmeningitic cases.

Inner Ear Haemorrhage
This is usually secondary to anti-coagulant treatment or trauma (with or without fracture of the temporal bone). Other possible less frequent causes are tumours, labyrinthitis or haematological diseases such as leukaemia or coagulopathies. Clinically, it causes sudden deafness.

Figure 5  (A) Axial CT of right temporal bone showing ossification of the basal turn of the cochlea characteristic of labyrinthitis ossificans (black arrows). (B) T1 weighted coronal plane MR imaging without intravenous contrast showing an increased signal intensity of the membranous labyrinth, corresponding to intralabyrinthine haemorrhage (white arrow) (metahemoglobin).
Figure 6  (A) MR FLAIR image after injection of intravenous contrast showing dilatation of the endolymphatic space particularly prominent in the cochlea. (B) Detail of the left ear image where white arrows indicate dilatation of the endolymphatic space in the cochlea.

Figure 7  (A1) and (A2) T2 and T1 weighted MR images with intravenous contrast, showing a small lesion on the inside of the basal turn of the right cochlea (white arrow) which intensely and homogeneously enhances after injection of contrast. This lesion corresponds to an intracochlear SIL. (B1) and (B2) T1 weighted MR images with and without intravenous contrast, showing a transmodiolar SIL with intense enhancement occupying the cochlea (white arrow) and crossing the modiolus to the bottom of the IAC (arrow tip).
Magnetic resonance imaging is the technique of choice to evaluate the membranous labyrinth. In normal conditions, in T1 weighted sequences without contrast, the membranous labyrinth is visualised as an intermediate signal structure, similar to the brain parenchyma. When there is an intralabyrinthine haemorrhage, there is a characteristic hyperintensity of signal in this sequence (Fig. 7B).\textsuperscript{33,34}

\textbf{Ménière’s Disease}

Also known as endolymphatic hydrops, this is characterised by a triad of vertigo, tinnitus and unilateral sensorineural hearing loss. The physiopathological mechanism which triggers hydrops is an increase in endolymphatic fluid in the IE, because its absorption is reduced due to a malfunction of the endolymphatic sac.\textsuperscript{35} During acute episodes, endolymph expansion in the vestibular system causes ballooning in the perilymphatic space which ruptures the separating membrane. The mixture of the fluids of both compartments bathes the nerve receptors which trigger vertigo. The mechanical disturbance of the organ of Corti causes hearing loss.\textsuperscript{36}

Visualisation of endolymphatic hydrops in Ménière’s disease has always been a major objective for clinicians, and demonstration was only possible in post-mortem histopathological studies; however, it can now be visualised using magnetic resonance imaging.\textsuperscript{37}

The current method for visualising endolymphatic hydrops is based on the endolymphatic space being leak tight and isolated from perilymph, cerebrospinal fluid and blood (Fig. 6).\textsuperscript{38} After injection of intratympanic or intravenous contrast there is only contrast enhancement of perilymph and not endolymph.\textsuperscript{24} The FLAIR sequence is very sensitive to changes in T1 and other subtle alterations in the composition of the fluids, it enables perilymph to be visualised with high signal intensity (bright) surrounding the endolymphatic space which has not taken up contrast (dark).\textsuperscript{39,40}

\textbf{Intralabyrinthine Schwannoma}

Intralabyrinthine schwannomas are benign tumours which arise de novo from the perineural Schwann cell sheath of the intralabyrinthine branches of the vestibulocochlear nerve and initially have no component in the internal auditory canal.\textsuperscript{41} According to their origin and extension, they are classified as: intracochlear (Fig. 7A), intravestibular, vestibulocochlear, transmodiolar in cases where the tumour crosses the modiolus from the cochlea to the bottom of the IAC (Fig. 7B), transmacular when it crosses from the vestibule to the bottom of the IAC, and transotic when it crosses the entire IE from the bottom of the IAC to the middle ear.

It has been published that intracochlear schwannomas originate most commonly in the scala tympani, and later growing towards the scala vestibuli.\textsuperscript{42} They present clinically with tinnitus, vertigo and unilateral progressive sensorineural hearing loss.

In T2 weighted MR sequences they are identified as a ”filling” defect inside the hyperintense perilymph, and intensely enhance after contrast injection.\textsuperscript{43}

\textbf{Endolymphatic Sac Tumour}

The endolymphatic sac is located below the dura of the posterior fossa, at the end of the vestibular aqueduct. Its function is to drain the endolymph from the membranous labyrinth. Endolymphatic sac tumour is a neoplasm which originates in the neuroectodermal epithelium of the membranous labyrinth of the IE. It is a rare tumour which presents with unilateral sensorineural hearing loss. It was initially considered an adenocarcinoma because of its high local destructive capacity and recurrence, however, no histological malignancy has been demonstrated and its capacity to metastasise is almost nil. Anatomopathologically, it is a papillary cystadenoma originating from the pars rugosa (middle third) of the endolymphatic sac. It presents sporadically, but patients with von-Hippel-Lindau disease have a 15% predisposition for the tumour.\textsuperscript{44}

Computed tomography shows localised bone destruction which is characteristically retrolabyrinthine, with central intratumoral bone spicules.\textsuperscript{45} The characteristic MR image is heterogeneous, with hyperintense foci in T1 weighted sequences on haemorrhagic areas, and after injection of contrast, there are areas of decreased uptake due to calcification and haemosiderin deposit (Fig. 8).

\textbf{Conclusions}

Magnetic resonance imaging and computed tomography play an essential role in diagnosing patients with inner ear pathologies.
pathology. These techniques are considered complementary. Computed tomography is important because it precisely delimits the bone component of the otic capsule. With the high capacity of contrast, magnetic resonance enables soft tissues to be evaluated and characterized: using this technique the membranous labyrinth and the cranial nerves can be represented in an excellent way. Furthermore, it is possible to visualize brain anomalies coexisting with malformations or other IE alterations.

We consider it necessary to understand the embryogenesis and the anatomy of the inner ear. Likewise, we must be aware of congenital and acquired disorders which might affect the inner ear. The technique of choice should be chosen in line with the clinical scenario. In general, computed tomography is the method of choice to study traumatic pathology or otospongiosis. When tumoral or inflammatory disease is suspected, magnetic resonance imaging has been shown to be superior. Both techniques are complementary in cases of congenital malformation.

Conflict of Interest

The authors have no conflict of interests to declare.

References