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Isolated Aplasia of the Lateral Semicircular Canal without Hearing Loss: A Case Report

I. Azzahiri[1*], S. Kirami[1], M. Benzalim[1], S. Alj[1]

[1]Radiology Department, IBN TOFAIL hospital, Mohammed VI University Hospital, Marrakech, Morocco

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***Corresponding author:** I. Azzahiri
Radiology Department, IBN TOFAIL hospital, Mohammed VI University Hospital, Marrakech, Morocco

Abstract **Case Report**

We present a case of a 25-year-old woman with isolated aplasia of the lateral semicircular canal (LSCC), a rare congenital malformation that results in the absence or underdevelopment of the LSCC. The patient had vertigo, left-sided tinnitus, and persistent chronic headaches but no hearing loss. High-resolution computed tomography (CT) and magnetic resonance imaging (MRI) were used to detect the malformation and associated vasculo-neural conflict. The aplasia of the LSCC was the only abnormality observed. The article explains the embryology of inner ear development and associated anomalies. CT and MRI were found to be effective imaging techniques for detecting ear malformations. The article also explains the importance of proper diagnosis and assessment of hearing loss for patients with this malformation.

Keywords: aplasia, lateral semicircular canal (LSCC), chronic headaches, vasculo-neural.

INTRODUCTION

The inner ear is a complex organ that includes three semicircular canals (lateral, superior, and posterior) that are responsible for detecting changes in head rotation speed and sending sensory information to the brain via the vestibular nerve. Aplasia of the lateral semicircular canal is a congenital malformation of the posterior labyrinth that is characterized by the absence or complete underdevelopment of the lateral semicircular canal [1].

It often occurs in association with other vestibular and cochlear malformations within syndromes. Isolated aplasia of the LSCC is a very rare condition that may be accompanied by hearing loss and vestibular symptoms to varying degrees [2].

We present a case of isolated aplasia of the lateral semicircular canal, without hearing loss, with clinical, computed tomography (CT), and magnetic resonance imaging (MRI) findings related to this condition.

CASE REPORT

A 25-year-old woman presented with a three-year history of persistent chronic headaches, left-sided tinnitus associated with vertigo and cerebellar syndrome, without associated hearing loss. The patient had no personal or family history of ear-related issues and did not report any usage of ototoxic drugs or trauma.

A high-resolution computed tomography (CT) of the temporal bone without contrast injection was performed and revealed isolated aplasia of the left lateral semicircular canal, without any other associated signs. There was no filling of the tympanic cavity or any anomaly of the ossicular chain.

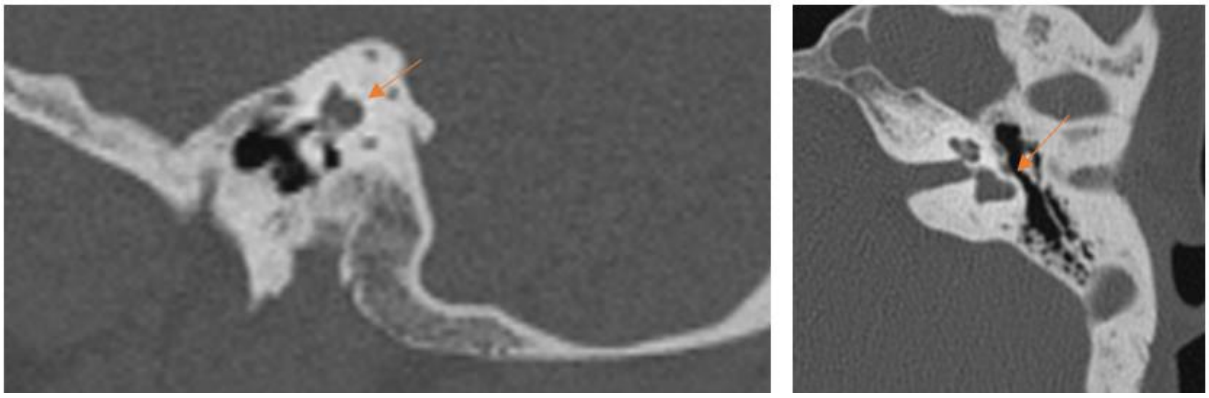

Figure 1: CT scan of the temporal bone without contrast: aplasia of the lateral semicircular canal (arrow)

To better explore the vertigo, magnetic resonance imaging (MRI) of both temporal bones was performed and revealed absence of individualization of the left lateral semicircular canal, while the superior and posterior semicircular canals were normal. No cochlear malformation or pathological contrast enhancement was observed. The left internal auditory canal and cerebellopontine angle were strictly normal, with evidence at the level of the right cerebellopontine angle of a vascular nerve conflict between the right anterior inferior cerebellar artery (AICA) and the homolateral cochleovestibular nerve VIII at the level of its cisternal pathway.

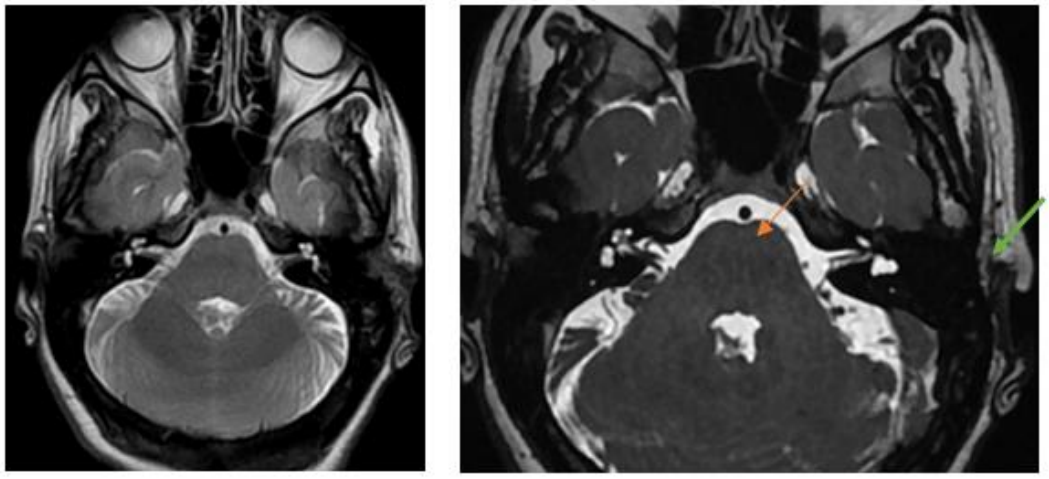

Figure 2: MRI of the cerebellopontine angle showing vascular nerve conflict on the right (orange arrow) and aplasia of the left semicircular canal (green arrow)

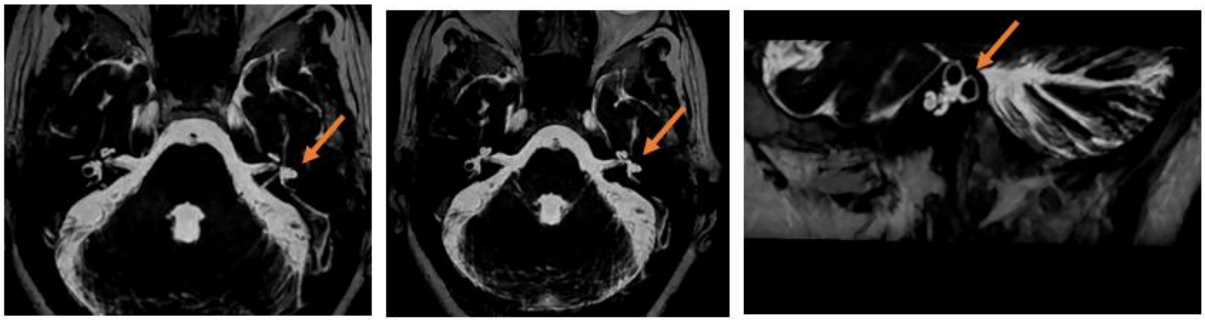

Figure 3: MRI (FIESTA sequence with MIP reconstruction) of the cerebellopontine angle showing aplasia of the left semicircular canal (arrows)

The diagnostic conclusion was isolated aplasia of the left lateral semicircular canal associated with a vascular-nerve conflict between the AICA and the VIII nerve on the right.

It is worth noting that the aplasia of the left lateral semicircular canal may explain the patient's symptoms.

DISCUSSION

Embryologically, the development of the inner ear starts during the 4th week of intrauterine life, with the formation of the otic vesicle from the ectoderm. The superior semicircular canal is the first to form at approximately 6 weeks of gestation, followed by the posterior and lateral canals. It is important to note that the lateral semicircular canal (LSCC) is the last one to develop and is the most commonly affected by aplasia. Aplasia of the LSCC is a rare congenital anomaly that results in the absence of the canal.

This malformation can occur in isolation, without any other associated congenital anomalies or as part of a syndrome [3]. The associated anomalies of these structures are common and variable. They may include stenosis, hypoplasia, and dysplasia of the semicircular canals, vestibular aqueducts, and cochlea. They may include stenosis, hypoplasia, and dysplasia of the semicircular canals, vestibular aqueducts, and cochlea [4].

Semicircular canal (SCC) aplasia is characterized by a complete lack of development of the respective canal and is roughly four times less prevalent than dysplasia. Aplasia of the LSCC is frequently associated with various syndromes, including but not limited to CHARGE syndrome and Noonan's syndrome [5].

Labyrinthine malformations, specifically those including to the posterior labyrinth, are commonly identified through the use of computed tomography (CT) and magnetic resonance imaging (MRI) Among these malformations, the lateral semicircular canal (LSCC) is frequently encountered, accounting for approximately 63% of all posterior labyrinth malformations [3].

Only a limited number of cases of non-syndromic isolated lateral semicircular canal (LSCC) aplasia were identified in the literature.

In our case, LSCC aplasia was associated with vestibular symptoms without detectable hearing loss. This anomaly alone can explain the clinical symptoms in the patient since decreased hearing is not necessarily associated with this malformation. In the context of LSCC dysplasia, the severity of sensori-neural hearing loss varies from normal hearing to severe sensorineural hearing loss, probably related to other combined inner ear anomalies, which requires audiometric and functional investigations in our patient. Our case also presented a contralateral vasculo-neural conflict, which may explain the associated tinnitus.

Computed tomography (CT) and magnetic resonance imaging (MRI) are two non-invasive imaging methods that can be used for the detection of ear malformations. CT provides a precise image of the bony structures of the ear, making it particularly useful for the detection of bony malformations such as the size of the tympanic cavity, the presence and nature of ossicular anomalies, oval and round window atresia, as well as anomalies in the course of the facial nerve canal [6].

On the other hand, MRI is more suitable for evaluating cochlear morphology in cases of insufficient separation or malformations of the modiolus. It also allows for measuring the size of the cochlear division of the vestibulocochlear nerve. MRI is particularly useful for visualizing details of the soft tissues of the inner ear, such as nerves and canals, as well as associated bony structures [6].

The combined use of these two imaging modalities allows for more precise and complete detection of ear malformations [5]. This approach has become increasingly important in recent years due to the increased sensitivity of modern imaging techniques, which have enabled more frequent detection of these malformations.

CT and MRI can also show other anomalies associated with SCC aplasia, such as an abnormal course of the facial nerve canal, ossicular ankylosis, dysplastic modiolus, or stenosis of the internal auditory canal [2]. In our case, all structures of the inner ear appeared normal except for the isolated aplasia.

The main differential diagnosis of SCC aplasia is fibrous or calcified obliteration of the SCC in cases of labyrinthine ossification, which is typically associated with a history of progressive hearing loss after an episode of meningitis [5]. The cochlea is also obliterated with involvement of the SCC in cases of labyrinthine ossification.

CONCLUSION

Semicircular canal (SCC) aplasia is a malformation of the posterior labyrinth characterized by a complete lack of development of the respective canal. It is typically accompanied by other vestibular and cochlear malformations within the context of syndromes.

Non-syndromic isolated lateral semicircular canal (LSCC) aplasia remains very rare. CT scan and MR imaging represent non-invasive modalities that can be used for the detection of ear malformations.

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