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Case report

Isolated lateral semicircular canal aplasia: Functional consequences

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ABSTRACT

Introduction: Lateral semicircular canal aplasia is a malformation of the inner ear, usually associated with vestibular and cochlear malformations in the context of congenital malformation syndromes. We report a rare case of a young patient with isolated lateral semicircular canal aplasia and no associated vestibular symptoms.
Clinical case summary: A 20-year-old man with no personal or family history presented with persistent unilateral tinnitus for three years with no associated vestibular symptoms. Moderate unilateral right sensorineural hearing loss was detected. Magnetic resonance imaging demonstrated isolated aplasia of the right lateral semicircular canal. Videonystagmography revealed right hyporeflexia. Vestibular evoked myogenic potentials were absent after stimulation on the right side and normal on the left side.
Discussion: Although the morphological abnormalities appeared to be isolated on imaging, the patient presented functional signs of global cochlear, semicircular canal and otolithic lesions, probably related to a developmental disorder of the membranous labyrinth. Functional investigations must be performed in the presence of isolated semicircular canal aplasia, even when it is an incidental finding, to exclude more extensive labyrinthine lesions.

1. Introduction

Malformations of the inner ear can affect the cochlea, vestibule, or both structures simultaneously [1]. Lateral semicircular canal (LSCC) aplasia is a malformation of the posterior labyrinth, usually associated with other cochlear and vestibular malformations in the context of congenital malformation syndromes [2]. LSCC aplasia is usually responsible for severe congenital nerve deafness. We report a case of isolated LSCC aplasia. Several functional investigations were performed to verify the correlation between the morphological lesion and the patient's symptoms.

2. Case report

A 20-year-old man presented with a three-year history of persistent right tinnitus with no associated vestibular symptoms, but had noticed recent onset of hearing loss, which was only disabling in noisy environments. He had no personal or family history and did not report any ototoxic drug use or sound trauma. Otoscopic examination was normal and Weber's test was lateralised to the left.

Videonystagmoscopy did not reveal any spontaneous nystagmus. Bone-conducted vibration at 100 Hz induced left horizontal-rotatory nystagmus after mastoid stimulation on each side.

Pure-tone audiometry revealed moderate sensorineural hearing loss on the right side, predominantly affecting high frequencies, with thresholds of 10 dB HL from 500 to 2000 Hz, 60 dB at 4000 Hz and 70 dB at 8000 Hz and a normal audiogram on the left side. An audiogram performed one year earlier showed better thresholds at 4000 and 8000 Hz, of 45 and 65 dB, respectively. Tympanometry was normal.

The radiological assessment comprised computed tomography (CT) and magnetic resonance imaging (MRI), which revealed isolated right LSCC aplasia. No signs of the lateral canal were detected, while the superior and posterior semicircular canals were normal. No cochlear malformation and no pathological contrast enhancement were observed. The internal auditory canal and the cerebellopontine angle were strictly normal (Fig. 1).

The videonystagmographic caloric test revealed right hyporeflexia with a 93% deficit with reflexivity of 8.8/s on the left and 1.0/s on the right after stimulation with cold water, and 12.7/s on the left and 0.2/s on the right after stimulation with hot water.

Auditory brainstem responses delivered reproducible waves, at normal latencies, shorter on the right side. The I–V latency interval was shorter on the right, compatible with an endocochlear recruitment phenomenon. Cochlear nerve and lateral lemniscus latencies

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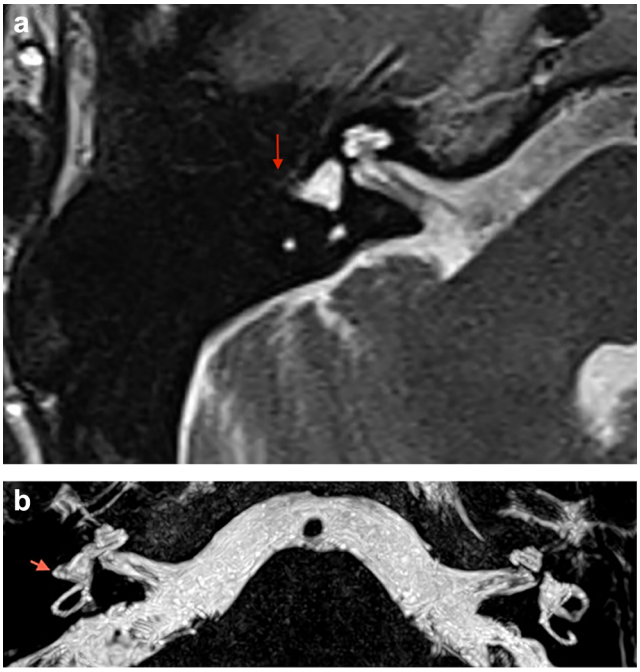

Fig. 1. a: magnetic resonance imaging, axial section, T2HR 3D sequence with MIP reconstruction; the red arrow indicates lateral semicircular canal aplasia with no associated cochlear or vestibular abnormality; b: magnetic resonance imaging, axial section, T2HR sequence; the red arrow indicates lateral semicircular canal aplasia.

were 1.90 and 5.80 ms at 80 dB on the right and 1.50 and 5.85 ms at 80 dB on the left (Fig. 2).

Vestibular evoked myogenic potentials showed good responses on the left and right sternocleidomastoid muscles after left-sided stimulation from 75 dB. No reproducible response was observed after right-sided stimulation up to 105 dB (Fig. 3).

3. Discussion

Embryologically, the superior semicircular canal is the first semicircular canal to develop, followed by the posterior semicircular canal and then the lateral semicircular canal during the 5th week

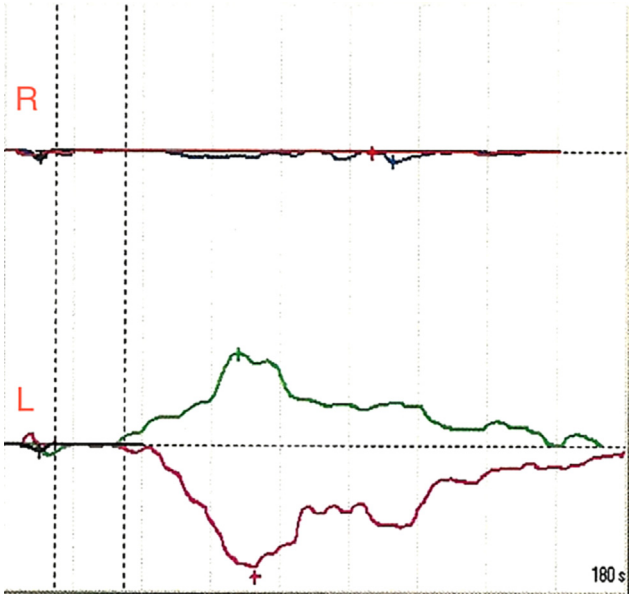

Fig. 2. Videonystagmographic caloric tests: right hyporeflexia with 93% deficit.

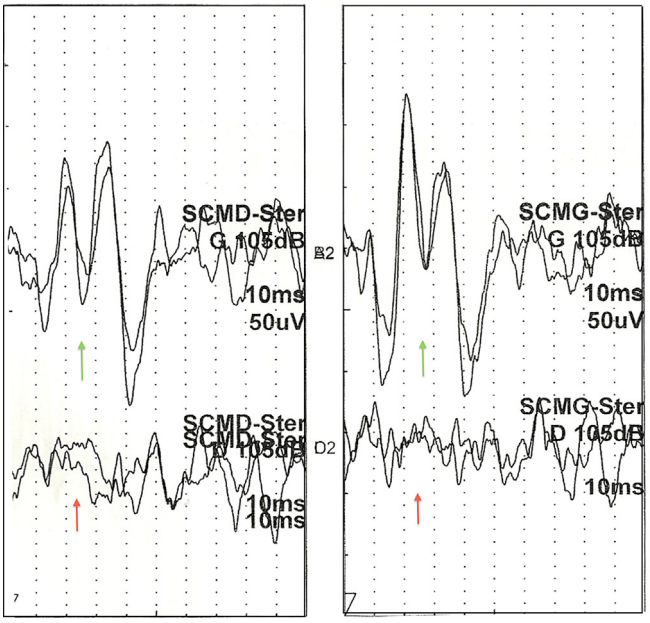

Fig. 3. Vestibular evoked myogenic potentials: good responses were observed for both sternocleidomastoid muscles after left-sided stimulation from 75 dB (green arrows), but no reproducible response was observed after right-sided stimulation up to 105 dB (red arrows).

of embryonic development. The LSCC is the last canal to be formed and the most frequently affected by aplasia. Abnormalities of the superior and posterior semicircular canals are therefore invariably associated with abnormalities of the LSCC, while isolated abnormalities of the LSCC can be observed [3]. The most common congenital abnormality is LSCC dilatation [4]. LSCC aplasia is defined as complete absence of development of the canal and is four times less frequent than LSCC dysplasia [5]. LSCC aplasia is part of several syndromes, such as CHARGE syndrome or Noonan's syndrome [2–4]. In the classification established by Sennaroglu and Saatci [6], semicircular canal aplasia was never detected in the absence of other malformations. Another retrospective study based on 16 patients [7] reported the largest series of LSCC malformations: 18% of these patients presented isolated LSCC dysplasia and 37% presented LSCC aplasia, but always in a context of congenital malformation syndromes. A literature search did not reveal any case of isolated LSCC aplasia.

Although this case of LSCC aplasia was morphologically isolated, it was nevertheless functionally associated with global vestibular, semicircular canal and otolithic deficits, although the patient did not report any vestibular symptoms. The otolithic deficit suggests a global anomaly of the membranous labyrinth; the absence of vestibular symptoms could be explained by a compensation phenomenon due to the congenital nature of this malformation.

This case of LSCC aplasia was associated with moderate sensorineural hearing loss. Sensorineural hearing loss is commonly observed in association with LSCC malformations, but no correlation has been established between the severity of the malformation and the audiometric deficit [7]. In a context of LSCC dysplasia, the severity of sensorineural hearing loss varies from normal hearing to severe sensorineural hearing loss [8,9], probably related to other abnormalities such as developmental disorders of the membranous labyrinth.

The results of audiometric and vestibular functional investigations demonstrated global labyrinthine deficiency with cochlear, semicircular canal and otolithic abnormalities, although the malformation appeared to be morphologically isolated. In the context

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of a diagnosis of isolated LSCC aplasia, functional investigations must be performed to detect more extensive anomalies of the labyrinth.

Disclosure of interest

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

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