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Case Report

Asymptomatic unilateral lateral semicircular canal aplasia associated with a dilated vestibule

Nuno Medeiros*, Pedro Valente, Artur Condé, João Larangeiro

Department of Otorhinolaryngology, Centro Hospitalar Vila Nova de Gaia, Portugal

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***Correspondence:**
Dr. Nuno Medeiros,
E-mail: nuno.medeiros.oliveira@chvng.min-saude.pt

ABSTRACT

Lateral semicircular canal aplasia is a rare form of vestibular dysplasia. This is the last canal to be formed during the fetal period, becoming susceptible to isolated malformations. A 17-year-old male patient performed a magnetic resonance with a subsequent incidental finding of isolated, unilateral, lateral SCS aplasia associated with a dilated vestibule. He was asymptomatic and physical examination was unremarkable. Videonistagmography showed a diminished ipsilateral caloric response and ipsilateral v-HIT gains were normal. To the best of our knowledge, this is the first non-syndromic case of aplasia of the lateral SCS with no vestibular or audiological symptoms. This absence could be explained by compensation due to the congenital nature of this malformation.

Keywords: Aplasia, Semicircular canal, Congenital

INTRODUCTION

Inner ear abnormalities have been extensively recognized since the appearing of imaging modalities as the CT scan or magnetic resonance imaging (MRI), and various classification systems have been proposed.[1-3] They may affect the vestibular and cochlear apparatus simultaneously, but they may also appear isolated.[4] Epidemiologically speaking, mild hypoplasia of a semicircular canal is fairly common, while complete aplasia is an extremely rare finding.[5]

From an embryological perspective, it is now accepted that malformations arise after the arrest of the inner ear development during a specific point in its formation. As the LSC is the last canal to be formed during the fetal period it becomes, therefore, susceptible to isolated dysplasia. It is also recognized that the vestibular apparatus is phylogenetically older than the cochlear one.[6] That would suggest that vestibular development anomalies should be present in concomitance with cochlear anomalies. This is not always the case. One possible explanation is that there are several genes specific to each one of these structures, although the embryophatogenesis of this finding is still up to much debate.[7]

Some syndromes, like CHARGE syndrome, have been associated with inner ear abnormalities, but these may also be found in non-syndromic patients.[8] Usually, there are cochlear and/or vestibular complaints, albeit not always promptly recognized. We report our experience with a patient with unilateral LSC aplasia associated with ipsilateral dilation of the vestibule, and without vestibular or cochlear symptoms. A review of the literature and speculative considerations are also given.

CASE REPORT

A 17 years old Caucasian male patient with a diagnosis of epilepsy performed an MRI with a subsequent incidental finding of unilateral lateral SCS aplasia associated with a dilated vestibule.

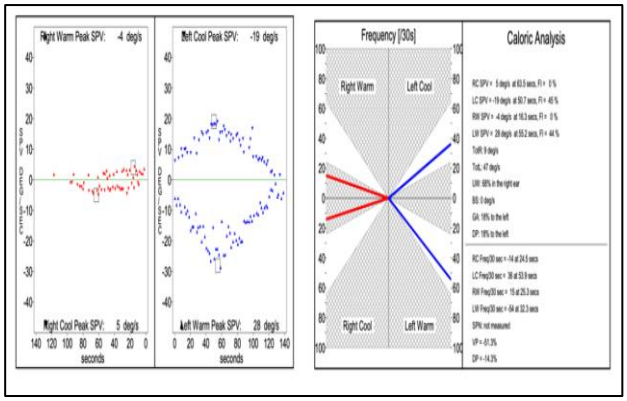

Figure 1: Videonistagmography showing asymmetry in caloric bithermal testing.

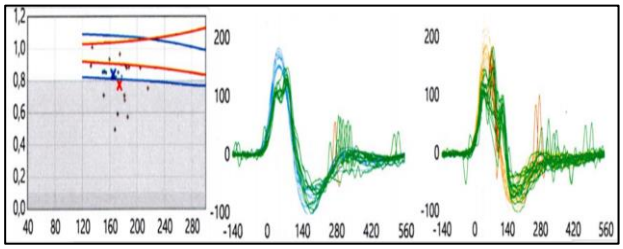

Figure 2: Video-head impulse test showing normal gain values with abnormal curve configuration. 1st image: right ear (red) and left ear (blue); 2nd image: left ear v-HIT; 3rd image: right ear v-HIT.

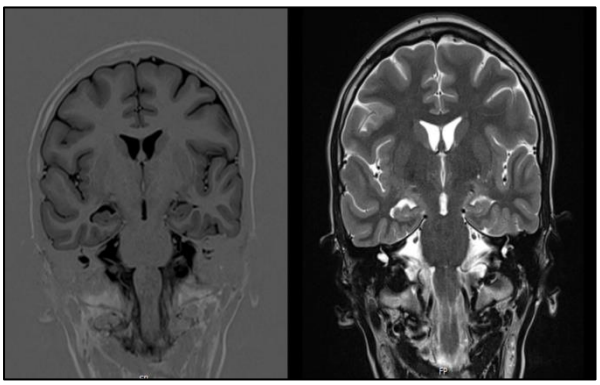

Figure 3: Cranioencephalic MRI showing aplasia of lateral SCC with vestibule dilation.

He was therefore sent to our neuro-otology consultation. No sensation of imbalance or vertigo was mentioned. The teenager was described as a normal kid, although sometimes clumsy and uncoordinated for sports. Hearing loss or tinnitus were not present. The patient's history was negative for other diseases, medication. Family history was also unremarkable.

Vestibular examination showed no resting or gaze-evoked nystagmus, and no overt saccade was present in the head impulse test. Videonystagmography with bithermal caloric test demonstrated an asymmetric vestibular response and visual-ocular control within normal ranges (Figure 1) Video Head Impulse Test was also performed, with gains above 0.8 bilaterally. The ipsilateral curve had a slightly abnormal configuration, with an indentation (Figure 2). Audiogram and impedanciometry were normal.

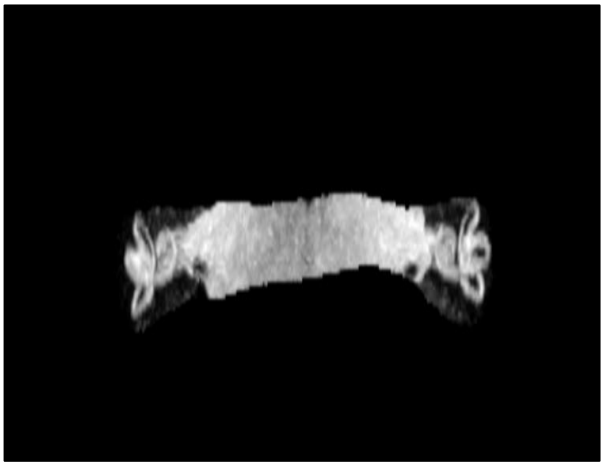

Figure 4: 3D reconstruction showing aplasia of lateral SCC with vestibule dilation.

MRI and CT evaluation, with native images and 3D reconstruction, showed no abnormalities in the right side. (Figure 3)

DISCUSSION

LSC dysplasia is one of the most common malformations in the inner ear, although the reported frequency varies with the severity of the finding. They are related to interruptions in the early embryogenic period (between the 5th-6th week of fetal development). The pars superior forms the utricle and semicircular canals, while the pars inferior becomes the saccule and cochlea.[9] Aplasia of one SCC is considered a rare finding, compared with hypoplasia or mild dysplasia.[8]

Previously, it was believed that a simple arrest in development would explain all of the inner ear abnormalities. Yet, this has been systematically challenged. Chronologically, this would not explain isolated SCS malformations, as the membranous labyrinth forms before the complete formation of the cochlea. On these grounds, we may assume that aberrations in specific vestibular genes may be one possible explanation for these isolated findings.[8,10]

LSC aplasia may be isolated or associated with syndromes like Goldenhar syndrome, CHARGE syndrome, trisomy 8 or 11.[11] The syndromic variety is more common. Published case series reports state that the percentage of dysplasia in CHARGE syndrome patients is reported in the majority, with most having no canal formation and a small subset having them partially developed.[12,13]

Associated hearing-loss is reported as present in the majority of LSC dysplasia cases. Venkatasami et al described this finding in 61% of the unilateral dysplasia and 81% if bilateral dysplasia.[14] Compared to other smaller, previous case-series, this corresponds to a higher percentage of normal hearing thresholds than what was previously described.[15] This hearing loss is more often SNHL, although mixed and CHL can be present. They also reported a surprising finding of contralateral hearing loss in unilateral dysplasia cases, proving the need for an extensive and throughout study of these incidental findings.[14]

Tests of vestibular function did not show a complete absence of response in this case. It is well documented that plasticity of the nervous system can confound the identification of vestibular dysfunction. Neural changes appear mainly when the changes are congenital or appear early in life, and these may alter the expected results on traditional testing methods, masking the deficit.[16]

Considering the vestibular aqueduct enlargement, this is also a common radiological finding, often associated with SNHL, the most common symptom arising from this change.[17] Many case reports reveal no videonistagmography/eletronistagmography alterations, with no asymmetry in caloric response.

To the best of our knowledge, no study in the literature has reported cases of asymptomatic unilateral LSC aplasia with vestibule dilation. Concerning the pathophysiology of this condition, we hypothesized that central compensation was enough, in this case, to maintain vestibular function within normal levels, being compensated by the contralateral side. We postulate these findings may be more common than previously thought, and sometimes detected only incidentally, as with this case.

CONCLUSION

Isolated unilateral LSC has been little described, being rare and seldomly asymptomatic. As this was an incidental finding, one may presume its true incidence is currently undervalued. The absence of vestibular symptoms shows possible complete central compensation. In cases like this one, a syndromic association should be extensively investigated.

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