Unilateral cochleo-vestibular nerve and internal auditory canal malformations

Jednostronne zaburzenia rozwojowe nerwu przedsionkowo-ślimakowego i przewodu słuchowego wewnętrznego – opis przypadku

Henryk Kaźmierczak, Wojciech Kaźmierczak, Katarzyna Pawlak-Osińska, Stanisław Osiński

Department of Otolaryngology and Laryngological Oncology with Audiology and Phoniatrics Division, Nicolaus Copenicus University Medical College, Toruń, Poland
Department of Sense Organ Research, Nicolaus Copenicus University Medical College, Toruń, Poland
Department of Pathophysiology of the Hearing and Balance Organs, Nicolaus Copenicus University Medical College, Toruń, Poland

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ABSTRACT: A case report of unusual one-sided developmental disorders of the vestibular and cochlear nerve and the internal auditory canal and difficulty of diagnostic process.

KEYWORDS: internal auditory canal malformations, unilateral cochleo-vestibular nerve malformations

STRESZCZENIE: Opis przypadku pacjentki z nietypowym jednostronnym zaburzeniem rozwojowym nerwu przedsionkowo-ślimakowego i przewodu słuchowego wewnętrznego oraz związane z tym problemy diagnostyczne.

SŁOWA KLUCZOWE: zaburzenia rozwojowe nerwu przedsionkowo-ślimakowego, zaburzenia rozwojowe przewodu słuchowego wewnętrznego

Congenital vestibular malformations are a rare cause of dysfunctions within this part of the balance system, and full diagnostics of these dysfunctions is difficult without imaging and genetic studies [1, 10]. Actual deficits in static and dynamic balance may be slight or indiscernible thanks to visual and proprioceptive compensation mechanisms [3, 6]. In addition, not all sensory cell islands may be affected by the developmental malformations of vestibular structures as is the case in Usher or Lange-Nielsen syndromes presenting with macular dysplasia alone or the Scheibe syndrome characterized by saccular dysplasia alone.

Most commonly, congenital malformations of peripheral structures are detected in patients examined for hearing loss, as various grades of cochlear structure dysplasia are more common

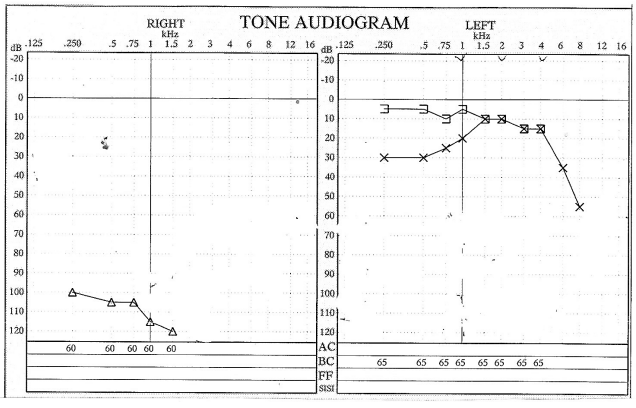

Fig. 1. Deep sensorineural hearing loss diagnosed on the right side in pure tone audiometry examination.

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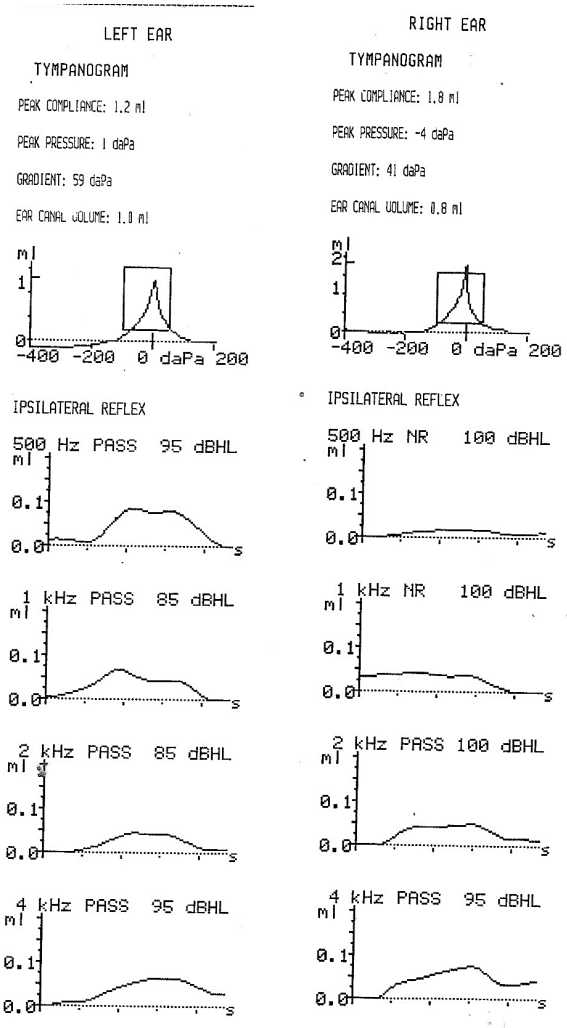

Fig. 2. Type A tympanogram of the right ear.

in various developmental disorder syndromes [11, 13]. What is important from the diagnostic standpoint, a significant correlation is observed between the hearing loss and hypo- or areflexia of ipsilateral labyrinth which, as mentioned before, is not brought into prominence due to the compensation mechanisms. Besides the aforementioned syndromes, vestibular pathologies are observed in DiGeorge syndrome, Cornelia de Lange syndrome, Marfan syndrome, Potter syndrome, Möbius syndrome, Arnold-Chiari malformation, Klippel-Feil syndrome, and trisomies 13, 18, 21, and 22 [11]. The anomalies may involve individual labyrinth structures or groups of more than one element, including isolated canals, vestibular aqueduct, the entire vestibule, or the vestibular nerve.

Concomitant abnormalities observed in other organs or systems, such as the organ of vision, the hormonal system, the

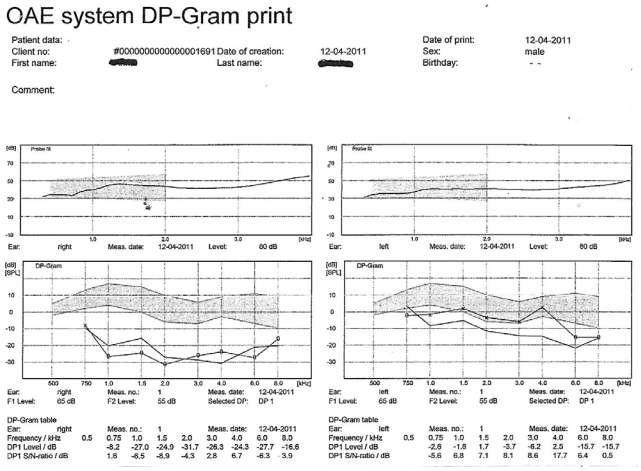

Fig. 3. Otoacoustic emission (OAE) tests revealed significant reduction in the amplitude of emissions for the right ear.

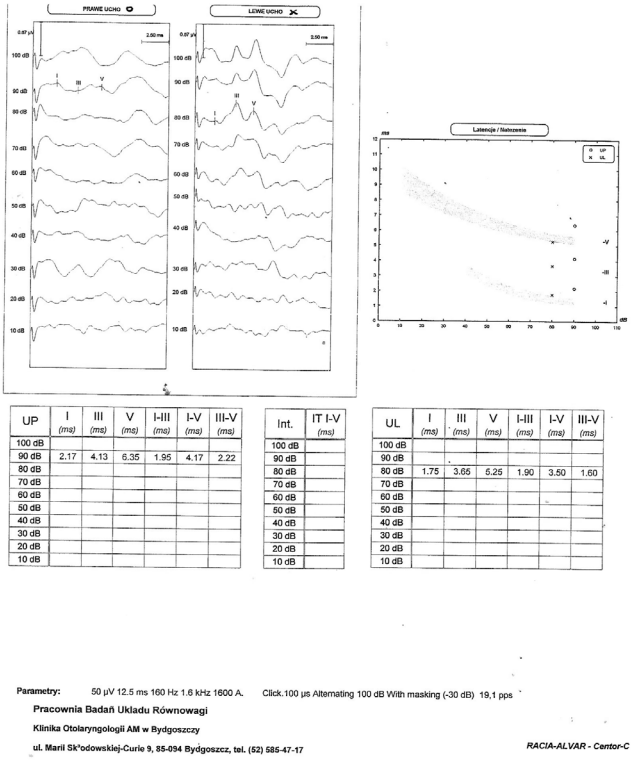

Fig. 4. Brainstem evoked response audiometry (BERA) examination revealed no clear response on the right side.

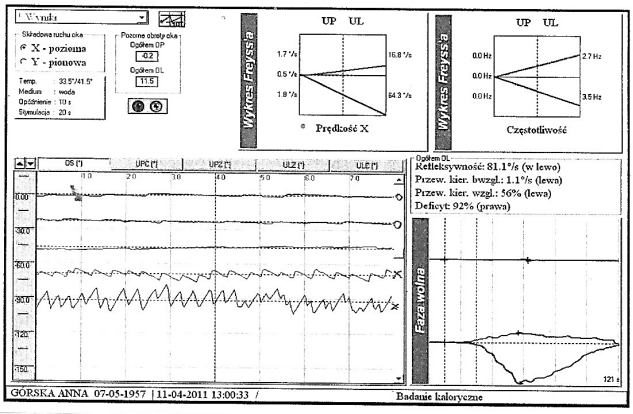

Fig. 5. Videonystagmographic (VNG) caloric testing of the examination of the labyrinths revealed right-sided canal paresis. Pseudoareflexia was excluded in Brünings tests.

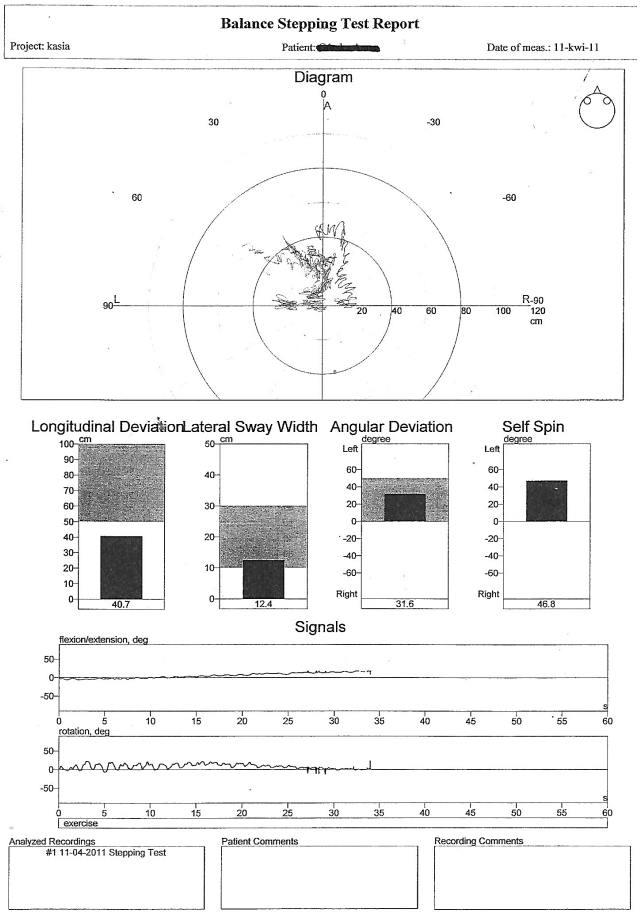

Fig. 6. Symmetrical rotatory nystagmus was observed in rotational chair tests. Vestibular evoked myogenic potentials (VEMP) evaluation revealed unremarkable morphology and correct latencies for P13/N23/P3 waves

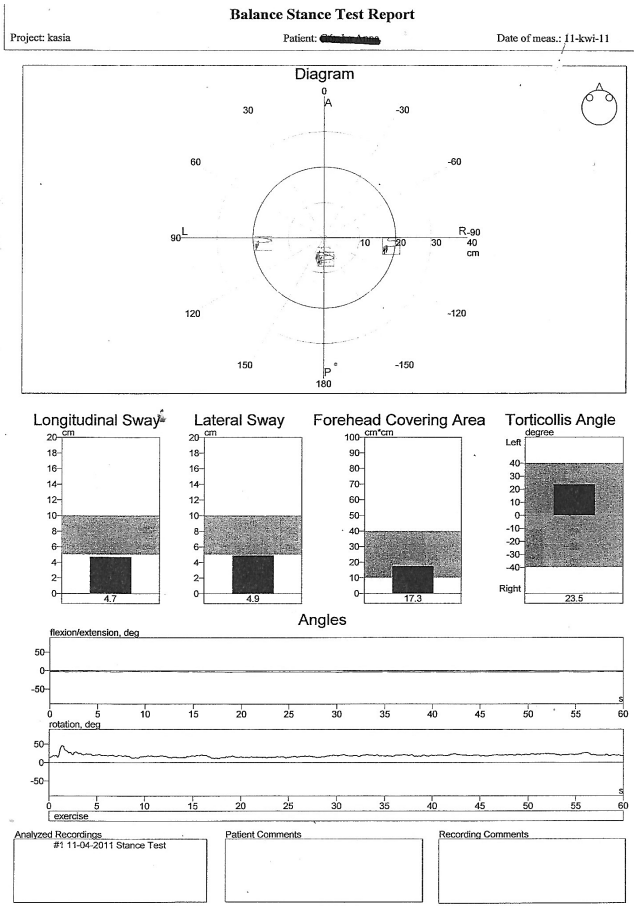

Fig. 7. Craniocorpographic (CCG) records of the Romberg test and the Unterberger tests were unremarkable.

cardiovascular system, the urinary system or the skin, may be of help in the diagnostic process [1,8,9,].

The objective of this study is to present a unilateral developmental malformation involving the vestibulocochlear nerve and the internal auditory canal.

CASE REPORT

Female patient G.A., aged 54, medical history no. 009834/11/564, was subjected to examination due to deep right-sided hearing loss and periodic balance disorders. The complaints had persisted since childhood. Medical history revealed no disorders of other sensory organs, viral diseases, injuries, intoxication, abnormalities in the cardiovascular, hormonal, urinary, skeletal

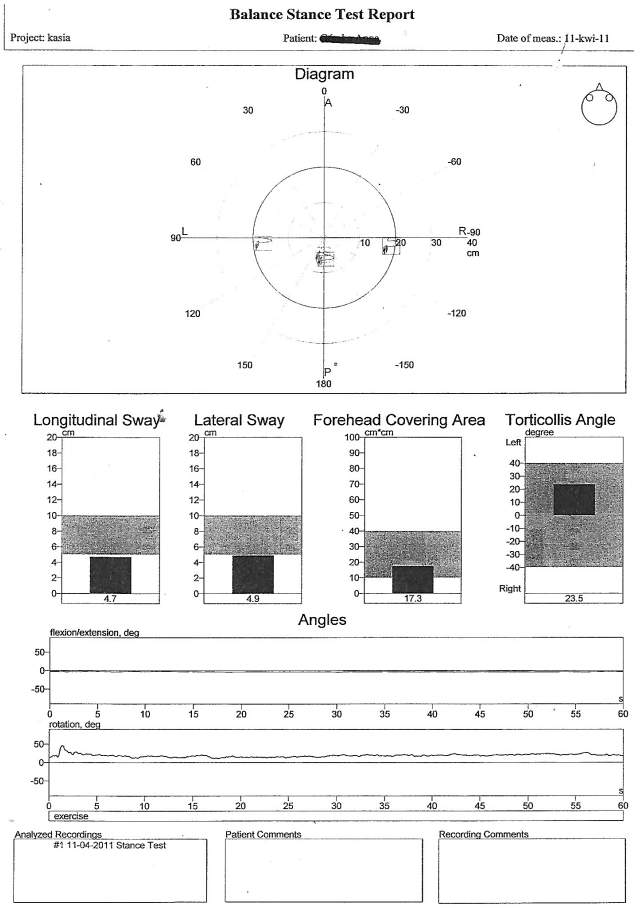

Fig. 8. Freyss stabilometry revealed normal Norre index and vhead/ vbody index values.

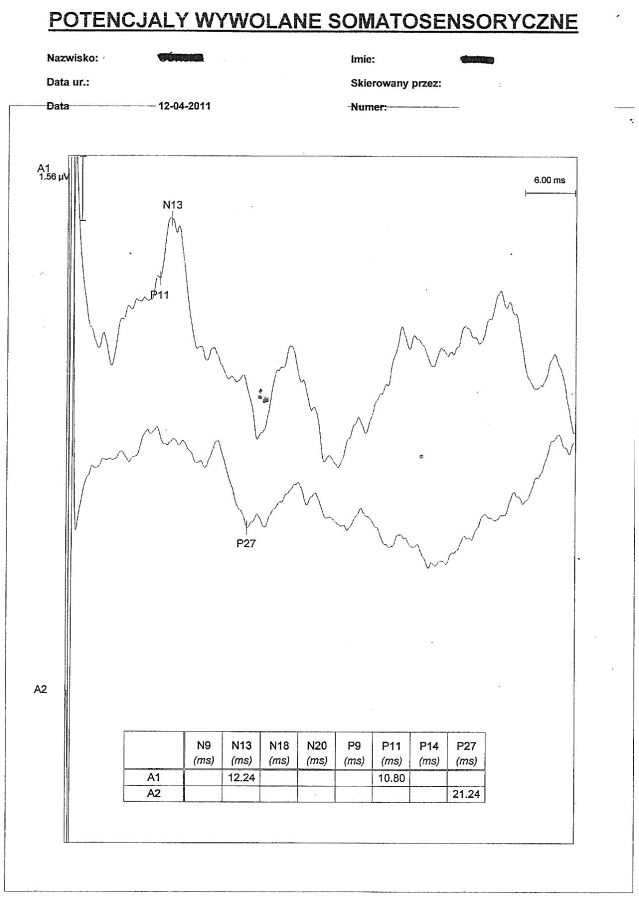

Fig. 9. Somatosensory evoked potential (SEP) tests revealed normal latencies for P11, N13, and P23 waves. No pathological lesions were found in the plain radiographic images of the spine.

system or skin function. Lateralization of hearing towards the contralateral ear was observed in the Weber test.

DISCUSSION

Embryonic development of the ear starts on gestation day 22 from the formation of otic placode on the lateral surface of the neural tube. Next, otocyst is formed and subsequently folded to develop into cochlear turn, utricle, and endolymphatic sac and duct. Semicircular canals are formed from the mesenchyme of the primordial vestibular part. Between weeks 16 and 22, the bone encasement of the labyrinth is formed [1]. Developmental disorders during embryogenesis may lead to aplasia or deformation of individual vestibulocochlear structures [12]. In their study involving 63 patients, Jackler et al. [5] observed 98 developmental malformations including, in the descending order of incidence, enlargement of the vestibule, the vestibular aqueduct, the lateral semicircular canal or the anterior semicircular canal. Malformations consisting in widened or narrowed internal auditory canal were the least common.

In our case, malformation of internal auditory canal and cranial nerve VII hypoplasia were detected during the diagnostic process taken up to identify the causes of a deep unilateral sensorineural hearing loss. Periodic vestibular disturbances experienced by the patient guided the diagnostic examination towards the assessment of the function and the structure of the labyrinth. Canal paresis was revealed by the caloric test, probably due to the distorted innervation of the crest structures as suggested by the MRI scan. On the other hand, VEMP evaluation was suggestive of the function of the right sac being maintained. The results of CCG, Freyss stabilometry, SEP, and rotating chair test confirmed the compensation of the right-sided vestibular deficit [3, 4]. The results of the hearing and vestibular function tests were suggestive of mal-

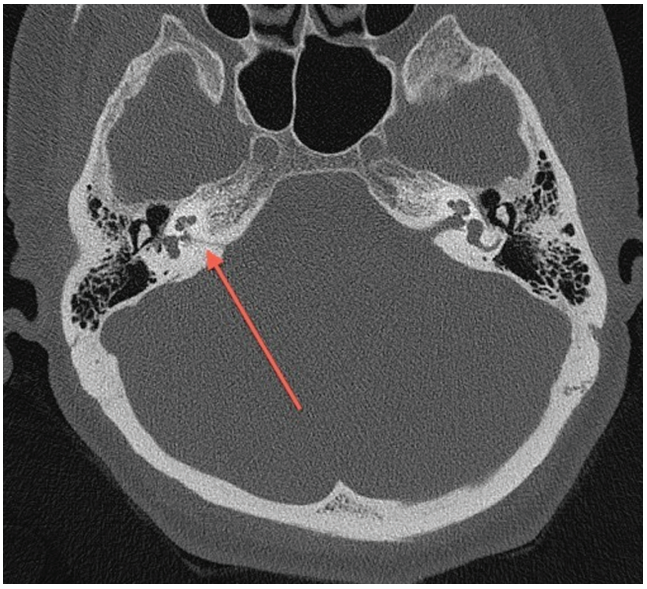

Fig. 10. CT-HRCT scan no. 618/11/CT1: pathological presentation of the internal auditory canal on the right. A narrow, bipartite canal, with the first canal, 1.8-2.3mm in diameter, being located along the course of the anterosuperior part of properly developed canal, and the second, fissure-like canal, 0.5mm in diameter, being located somewhat inferior and posterior to the first canal; both canals merge medially away from the vestibule. Also visible is the canal of the inferior vestibular nerve which innervates the saccular macula and the posterior ampullary crest; left temporal bone presentation unremarkable.

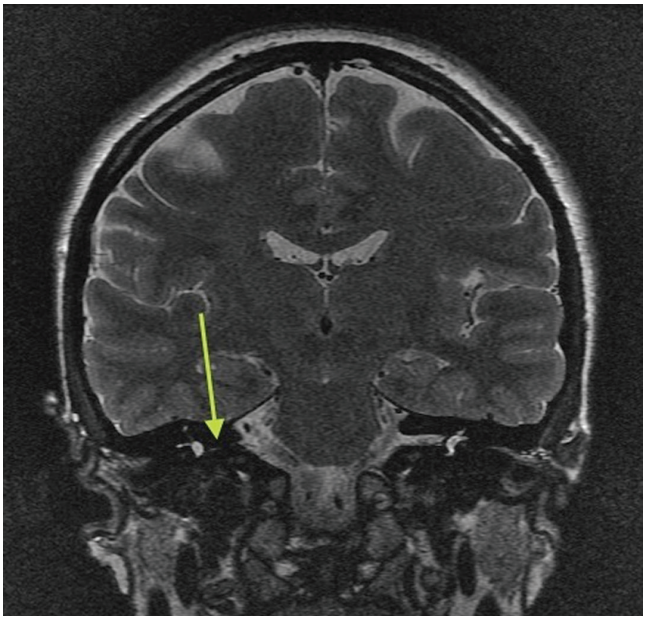

Fig. 11. MRI scan no. 1654/11/MR2: cranial nerve VIII on the right visible within the cerebellopontine angle; the nerve is narrower than the contralateral counterpart due to hypoplasia or aplasia or its cochlear components and the superior vestibular nerve; the ampullae, semicircular ducts, and cochlear turns bilaterally unremarkable; cranial nerve VII bilaterally unremarkable.

formation which could be classified as incomplete penetration trait (unilaterality, lack of complete deafness, maintained sac function) [7]. It was difficult to ascertain on the basis of clinical examination whether the developmental anomalies within this region involved, completely or partially, other structures of the inner ear (utricle, cochlear turn), and which epithelial elements were affected [5]. It is also difficult to classify malformations observed in the reported case as representative to any of the syndromes listed in the introduction. What could be determined, was that malformations involved the cranial nerve VII and the internal auditory canal, with no other organ or system-related deviations being confirmed. The case was somewhat similar to Mondini-like dysplasia with incomplete penetration [7, 12, 13].

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Corresponding author: Stanisław Osiński; Department of Pathophysiology of the Hearing and Balance Organs, Nicolaus Copenicus University Medical College ; e-mail: larystp14@gmail.com

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