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ORIGINAL ARTICLE

Audiovestibular findings in a branchio-oto syndrome patient with a *SIX1* mutation

YOSHIHIRO NOGUCHI, TAKU ITO, AYAKO NISHIO, KEIJI HONDA & KEN KITAMURA

Department of Otolaryngology, Tokyo Medical and Dental University, Tokyo, Japan

Abstract

Conclusion: A reported mutation in *SIX1* was identified in a patient with familial hearing loss (HL), a left preauricular pit, and bilateral enlarged vestibular aqueducts (EVA). Although the characteristic symptoms of EVA including fluctuating HL and repetitive vertigo were not seen in the patient, further studies are needed to clarify the association between EVA and such symptoms. *Objectives:* To study the audiovestibular functions, and to identify the causative gene in a patient with branchio-oto syndrome. *Methods:* We enrolled a 30-year-old female in whom HL was pointed out at the age of 6 years. She visited our department at the age of 21 years, and had not experienced any progression of her HL, tinnitus, or vertigo. Pure-tone audiograms showed bilateral moderate mixed HL with no apparent progression during a 9-year follow-up period. Audiovestibular examinations included distortion product otoacoustic emissions (DPOAEs), electrocochleography (ECochG), and electronystagmography (ENG). Direct sequencing was utilized to screen for *SIX1, EYA1, SLC26A4, GJB2,* and mitochondrial DNA *MTRNR1* including 1555 position. *Results:* The findings of DPOAEs, ECochG, and ENG indicated cochlear HL with no vestibular dysfunction. A previously reported mutation of a heterozygous c.386A > G (p.Y129C) in *SIX1* was detected. No mutation was identified in *EYA1, SLC26A4, GJB2,* or *MTRNR1.*

Keywords: *Hearing loss, deafness, vertigo, enlarged vestibular aqueduct, distortion-product otoacoustic emissions, electrocochleography, electronystagmography*

Introduction

Branchio-oto-renal (BOR) syndrome is an autosomal dominant disorder characterized by hearing loss (HL), branchial arch abnormalities, and renal anomalies. Although the penetration ratio is high, there is variability in the phenotypic appearance between and within families [1]. The most prevalent symptoms are HL (93%) and the presence of a preauricular pit (82%), followed by renal anomalies (67%), branchial fistulae (49%), pinna deformities (36%), external auditory canal stenosis (29%), preauricular tags (13%), and lacrimal duct aplasia (11%) [2]. HL can be caused by middle and/or inner ear abnormalities, and is categorized as mixed (52%), conductive (33%), or sensorineural (29%), with the severity ranging from mild to profound [2]. Branchio-oto (BO) syndrome that lacks renal anomalies is thought to be a phenotypic variant of the same disorder.

Three causative genes for BOR/BO syndrome, *EYA1*, *SIX1*, and *SIX5*, have been detected. Since Abdelhak et al. [3] discovered *EYA1* in 1997, more than 100 different mutations in *EYA1* have been reported as a cause of BOR/BO syndrome (designated as BOR1/BOS1) [1]. *EYA1* mutations are identified in approximately 40% of patients who meet the diagnostic criteria for BOR syndrome [4]. Recently, mutations in two *SIX* genes, *SIX1* and *SIX5*, were identified to be responsible for BOR/BO syndrome (BOS3 and BOR2, respectively) [5,6]. The mammalian *Six* gene family comprises six members (*Six1–6*), and their products share two highly conserved domains, a Six domain and a homeodomain [1]. *Six* genes are widely co-expressed with *Eya* and other

Correspondence: Ken Kitamura MD, Department of Otolaryngology, Tokyo Medical and Dental University, 1-5-45 Yushima, Bunkyo-ku, Tokyo 113-8519, Japan. Fax: +81 3 5803 0146. E-mail: kitamura.oto@tmd.ac.jp

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