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CASE REPORT

Persistent Stapedial Artery: Computed Tomography and Magnetic Resonance Imaging Features

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ABSTRACT

Persistent stapedial artery is a rare congenital anomaly that has significant implications for middle ear operations. This report presents the imaging features — computed tomography and magnetic resonance imaging — of persistent stapedial artery in a child with Noonan syndrome. The clinical issues associated with this rare condition are also discussed.

Key Words: *Arteries; Cochlear implants; Magnetic resonance imaging; Tinnitus; Tomography, X-ray computed*

中文摘要

永存鐙骨動脈—於電腦斷層掃描及磁力共振之影像特徵

郭啟欣、羅尚銘、曾子勤、范子和、關鼎樂

永存鐙骨動脈是一種罕見的先天性血管變異。此狀況會對中耳的手術做成障礙。本文章將闡述一出現於努南氏症候群的永存鐙骨動脈病例，並就其臨床及放射醫學影像之特徵作討論。

INTRODUCTION

Stapedial artery is transiently present during foetal development and normally involutes in the third foetal month. Failure of its involution leads to persistent stapedial artery. The imaging features and clinical impact of this rare condition are presented in this report.

CASE REPORT

A 3-year-old girl with Noonan syndrome and right-sided profound deafness was due to undergo right cochlea implant surgery. Preoperative computed tomography (CT) scan of the temporal bone was performed and no significant abnormality was reported.

During the cochlea implant operation, the surgeons encountered a pulsating structure that passed over the round window and extended upwards to the oval window. The operation was therefore abandoned.

Review of the CT scan of the temporal bone showed enlargement of the tympanic segment of the right facial nerve canal and absence of the right foramen spinosum (Figure 1). Right persistent stapedial artery was suspected. Magnetic resonance imaging (MRI) was performed and time-of-flight MR angiogram demonstrated abnormal origin of the right middle meningeal artery, which arose from the persistent stapedial artery (Figure 2).

DISCUSSION

Persistent stapedial artery is a rare condition with a quoted prevalence of 0.48%.[1] There is an association with the aberrant internal carotid artery and other middle ear

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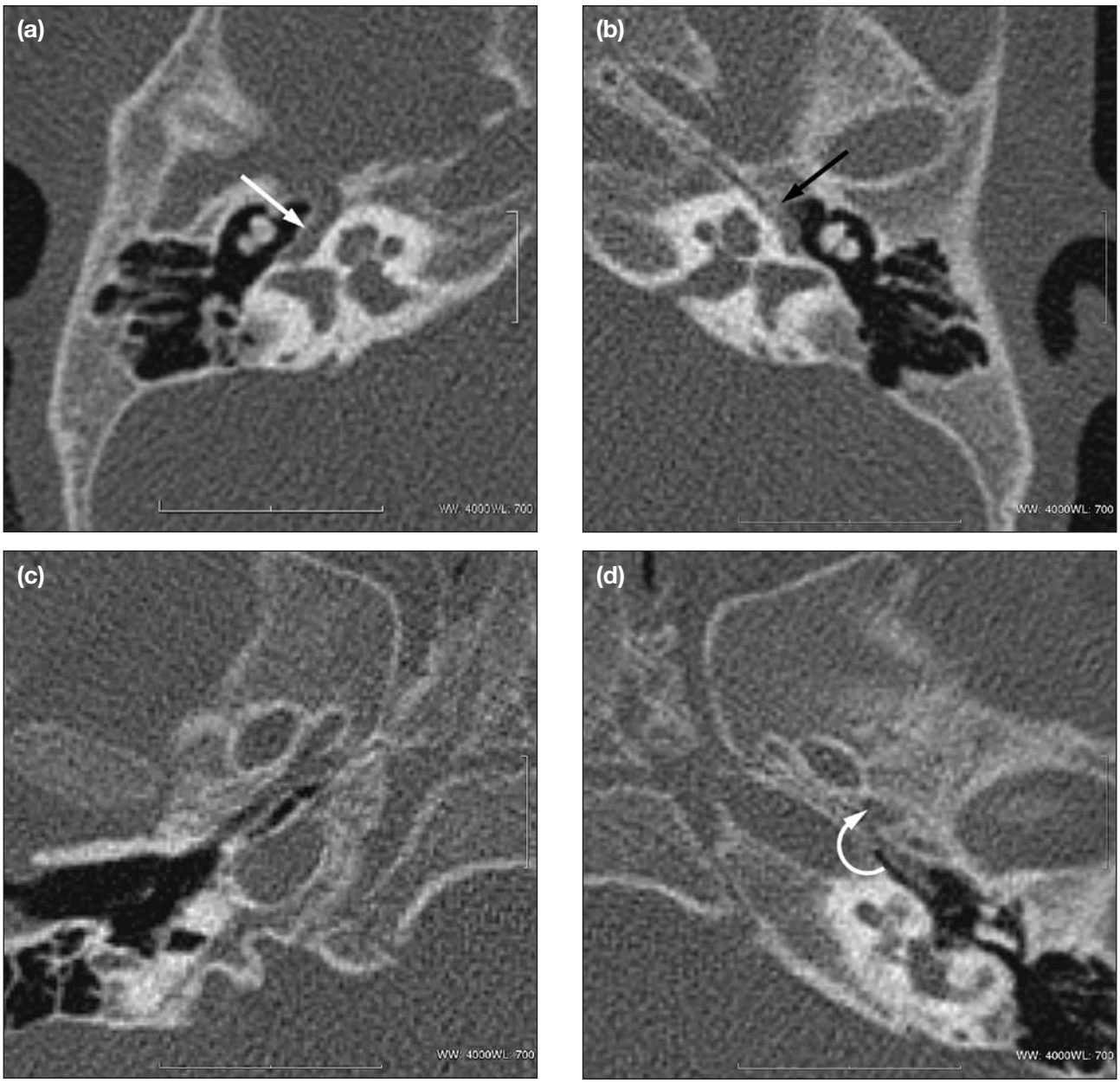

Figure 1. Axial computed tomography images of (a and b) the temporal bone at the level of the cochlea showing widening of the right facial nerve canal (white arrow) compared with the left facial nerve canal (black arrow); and (c and d) the skull base level showing absence of the right foramen spinosum, compared with the normal left foramen spinosum (curved white arrow).

abnormalities, especially involving the stapes and facial nerve.[2-4] Persistent stapedial artery may also be present in patients with trisomy 13, 15, and 21, thalidomide deformities, second brachial arch anomalies, congenital immunodeficiency, neurofibromatosis, Paget's disease, and otosclerosis.[2-4] The prevalence of persistent stapedial artery in Noonan syndrome is unknown. No reported case of persistent stapedial artery in patients with Noonan syndrome was found during a literature search.

Early in foetal life, the stapedial artery connects the branches of the future external carotid artery to the internal carotid artery and it normally involutes at the third foetal month.[2-5] If the stapedial artery persists in post-natal life, the middle meningeal artery would be supplied by the persistent stapedial artery. A persistent stapedial artery typically arises from the petrous internal carotid artery, enters the hypotympanum, passes cephalad through the obturator foramen of stapes, and then joins the facial nerve canal. The artery usually leaves the facial nerve canal just before the geniculate ganglion and travels anterosuperiorly in the extra-dural space of the middle cranial fossa, becoming the middle meningeal artery.

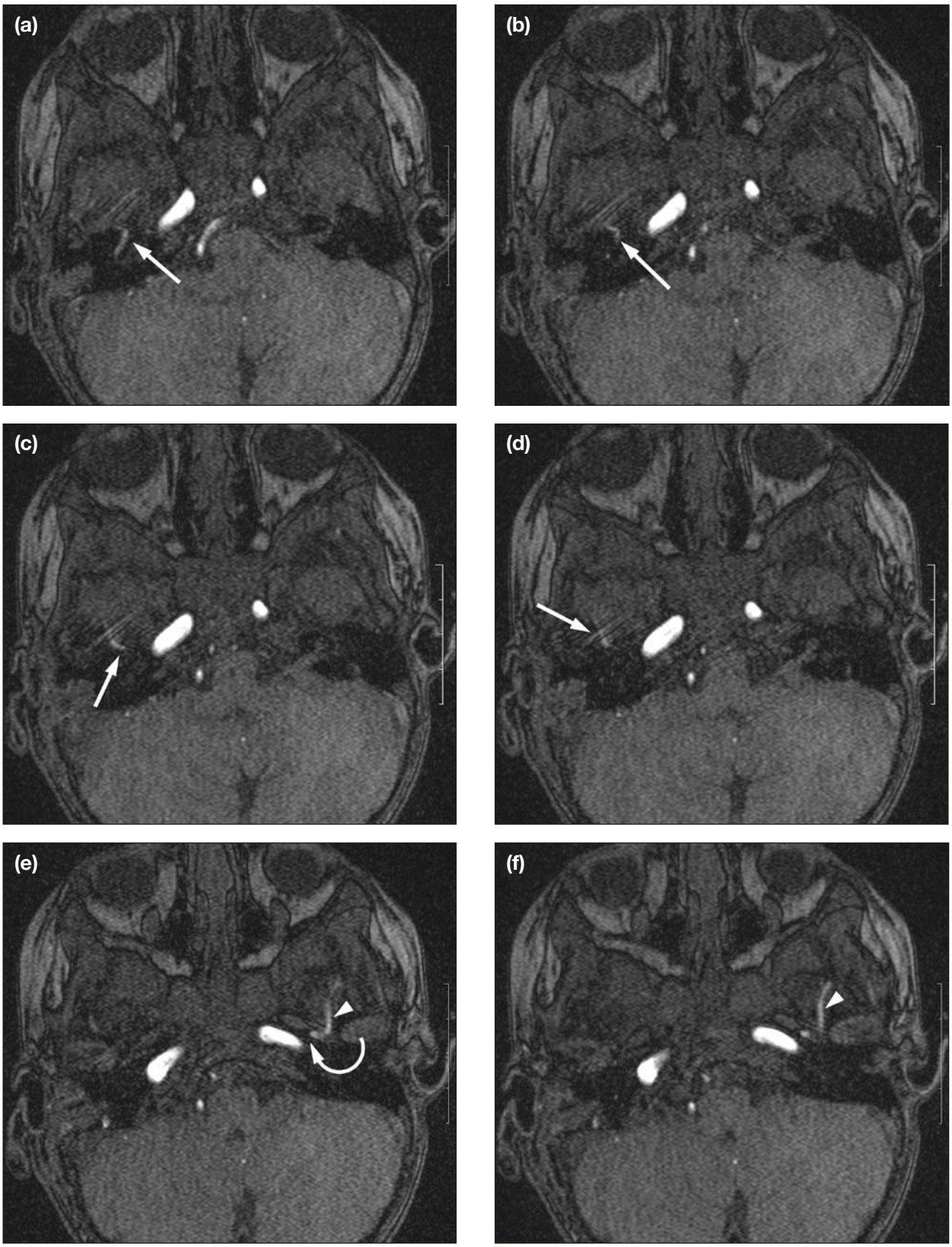

Figure 2. Time-of-flight magnetic resonance angiogram demonstrating (a to d) abnormal origination of the right middle meningeal artery (arrow) from the persistent stapedial artery; and (e and f) the normal course of the left middle meningeal artery (arrowhead) passing through the normal left foramen spinosum (curved arrow).

Clinically, patients with persistent stapedial artery are often asymptomatic and the condition is detected as an incidental finding on imaging.[2-5] Patients with this condition can also present with symptoms of pulsatile tinnitus and conductive hearing loss,[2-5] which is related to ankylosis of the stapes.[4] A persistent stapedial artery may also be encountered as a pulsatile mass during middle ear surgery.[2-5] Persistent stapedial artery is known to complicate stapedectomy and cholesteatoma resection, and can prevent cochlea implantation.[3] Therefore, it is important to look for this particular vascular anomaly before middle ear operations.

Persistent stapedial artery can be demonstrated on plain CT scan of the temporal bone. However, the vascular anomaly is so small that the vessel may not be identified. Two secondary radiological signs help to attract radiologists' attention and improve the detection of this subtle condition. First, as the persistent stapedial artery passes along the facial nerve canal, the tympanic segment of the facial nerve canal appears enlarged. Watanabe et al found that the normal mean diameter of the tympanic segment of the facial nerve canal was 1.201 ± 0.187 mm.[6] There is occasionally a separate canal parallel to the facial nerve in patients with persistent stapedial artery. Differential diagnosis of an enlarged facial nerve canal includes tumour mass such as facial nerve schwannoma, which may cause more focal enlargement. Second, aberrant origination of the middle meningeal artery from the persistent stapedial artery leads to absence of the foramen spinosum, which should be present when the middle meningeal artery arises from the internal maxillary artery as in the normal anatomy. Absence of the foramen spinosum is also a non-specific sign and occurs in up to 3% of the population as a normal variant, particularly when the middle meningeal artery originates from the ophthalmic artery.[4] Nevertheless, when both radiological signs are detected, the diagnosis of persistent stapedial artery has to be considered. Other useful radiological signs of persistent stapedial artery on plain CT scan of the temporal bone include visualisation of a small canaliculus leaving the carotid canal and a curvilinear structure passing over the cochlea promontory.

Persistent stapedial artery can also be diagnosed by time-of-flight MR angiogram.[5] As demonstrated in this patient, MR angiogram clearly reveals the course of the persistent stapedial artery and middle meningeal artery. However, there were no other specific clues detected on the remaining standard MRI sequences for this patient. No specific discussion of the features of persistent stapedial artery on routine MRI sequences of the internal auditory meatus or temporal bone was found during a literature search. To the authors' knowledge, time-of-flight MR angiogram may be the best way to demonstrate persistent stapedial artery during MRI study. Conventional angiogram is only rarely performed for symptomatic patients with persistent stapedial artery when ligation is considered. The purpose is to ensure that this is not an end artery and does not contribute a substantial blood supply to important tissues.[5]

In conclusion, persistent stapedial artery is a rare vascular anomaly that can have important consequences on middle ear surgery. Recognition of this condition by imaging can prevent unnecessary middle ear operations and potential complications.

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