Rare Pulsatile Tinnitus in Children Treated with Transcatheter Coil Embolization: A Case Report

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Tinnitus is the perception of abnormal ear or head noises without any external sounds. The noises might be high pitched or comprise ringing, clicking, or buzzing. Pulsatile tinnitus is a kind of rhythmical tinnitus that usually occurs at the same rate as the heartbeat and is rare in children. Herein, we present a 4-year-old girl who had an abnormal audible sound in the right ear during sleep for a year. We confirmed a simple congenital right external carotid artery to jugular vein fistula that produced pulsatile tinnitus and was successfully treated with coil embolization. Pulsatile tinnitus resulting from a congenital external carotid arteriovenous fistula in children is rare and deserves precise evaluation, as it can be effectively treated with transcatheter coil embolization.

Key words: Pulsatile tinnitus, Arteriovenous fistula, Transcatheter coil embolization

INTRODUCTION

Tinnitus is the perception of sounds such as buzzing, ringing, roaring, clicking, pulsations, and other noises in the ear or head occurring without an outside acoustic stimulus.[¹,²,³] Tinnitus is a symptom, not a disease; as such, it can arise as a result of inappropriate activity at any point in the auditory pathway. In a recent study conducted in the United States, about 50 million individuals (aged 20 years or older) reported having tinnitus over the last 12 months, with 16 million reporting that their tinnitus occurred at least once daily.[²] The most common cause of tinnitus involves noise exposure. Other causes of tinnitus include a variety of pathologies, illnesses, medications, allergies, dietary changes, stresses, and traumatic events.[³]

Pulsatile tinnitus is a type of noise in the ear that is perceived as a rhythmic pulsation that is often timed with the heartbeat.[⁴,⁵,⁶] It is often referred to as vascular tinnitus because for the majority of cases, it is related to blood flow disturbances in the head or neck region. This results from either increased blood flow or a narrowing of the blood vessel opening, both of which result from turbulent
blood flow that can be heard in the ears.

Arteriovenous fistulas (AVFs) in the head and neck region are rare. The majority of AVFs involving the major neck vessels are usually secondary to blunt or penetrating trauma.[7] Congenital AVFs between the external carotid artery and jugular vein(s) appear to be relatively rare in medical literature. Direct surgical ligation of an AVF is often curative. In surgically difficult cases involving the delicate head and neck anatomy, and multiple arteriovenous communications or collaterals, tranarterial embolization provides another treatment choice.[8,9]

Herein, we present a girl who had a rare congenital right external carotid AVF and who had a long period of night tinnitus and sleeplessness. The fistula was treated successfully with coil occlusion. Moreover, pulsatile tinnitus resulting from congenital carotid arterial AVFs in children is remarkably rare and can be effectively treated with a transcatheter coil technique.

**CASE REPORT**

A 4-year-old girl was in good health, without any traumatic neck history. However, she began to hear an abnormal sound in her right ear at the age of 3. The sound was so unusual that it could only be heard during quiet sleep and disappeared after waking up. The terrifying sound lasted for almost a year, resulting in insomnia and psychomotor behavior. Because there were no positive findings during any investigation, the girl was even considered to have a psychiatric illness.

A physical examination in our hospital revealed a pulsatile swelling in her neck, a thrill, and a grade II-III/VI bruit near the mandibular angle anterior to the ipsilateral ear. Cardiac auscultation revealed a grade I-II/VI systolic murmur over the left main stem bronchus. Her blood pressure, chest roentgenogram, and ECG were normal. All neurological and hearing function tests were negative. Further, all laboratory investigations were unremarkable. Transthoracic echocardiography revealed a normal intracardiac structure, normal aortic branches, and intact wall motion.

Arterial phase magnetic resonance angiography (MRA) of the head and neck eventually revealed a remarkably engorged right common carotid artery (RCCA) and an engorged right external carotid artery (RECA), as well as abnormal delineation between the right external jugular vein (REJV) and right internal jugular vein (RIJV). After MRA, interventional catheterization was performed. A right common carotid angiogram revealed a dilated RCCA, dilated RECA, and normal right internal carotid artery, as well as rapid opacification of the REJV and RIJV (Fig. 1A). A selective right external carotid angiogram clearly delineated a simple segmental fistula at the distal end of the RECA that supplied the REJV and RIJV (Fig. 1B). The RECA was engorged, with a maximum diameter of approximately 8–10 mm, but was connected to a single fistula, with the narrowest diameter of 2-3 mm. A single feeding artery with a narrow fistula that had no other major arterial collaterals near the lesion, and an accessible route for catheter manipulation helped guarantee that the patient would be a preferred and appropriate candidate for coil occlusion, as opposed to conventional
Pediatric pulsatile tinnitus

Two percutaneous transvascular coils (4 mm) were carefully deployed in the distal end of the RECA to occlude the fistula. A post-coil angiogram revealed the complete occlusion of the lesion, without any residual shunts (Fig. 1C, Fig. 1D). The patient remained well with complete symptom resolution. The post-coil period was uneventful without any complications or recurrences.

Figure 1. (A) A right common carotid angiogram delineating an engorged right common carotid artery (white arrow), a tortuous and dilated right external carotid artery, and a normal right internal carotid artery, as well as early opacification of the right jugular vein (dashed arrow). (B) Selective right external carotid angiography clearly revealed a simple, narrowed segmental fistula (white arrow) that was connected to the jugular vein (dashed arrow). (C) A post-coil right external carotid artery angiogram revealed the total occlusion of the fistula with 2 coils (white arrow), without any residual shunts or any right jugular vein disappearances. (D) A post-coil common carotid angiogram revealed the coils (white arrow) in situ, as well as a normal carotid artery distribution, with no early opacification of the right jugular vein.
DISCUSSION

Tinnitus is a persistent or intermittent noise that patients perceived to be heard in the ear. The prevalence of tinnitus increases with age.\[^3\] According to the American National Institute on Deafness and Other Communication Disorders (NIDCD), almost 12% of men who are 65 to 74 years of age are affected by tinnitus. Tinnitus is a symptom that is rarely mentioned by children but is frequently obtained after a careful history. Children’s ability to describe tinnitus depends on their verbal ability and experience. The reported prevalence of tinnitus in the United States varies from 7% for 5 to 17 year olds to 34% for 6 to 16 year olds, both in children without any type of hearing loss, and is reported for up to 66% of secondary school children with moderate to severe hearing loss.\[^2\]

Tinnitus may be either objective (i.e., able to be heard by the examiner using auscultation) or subjective (i.e., unable to be heard by the examiner). Objective tinnitus in children is usually pulsatile and of vascular origin.\[^4\] In children, the most common cause of pulsatile tinnitus is turbulent blood flow through normal vessels and vascular anomalies such as an aberrant carotid artery in the middle ear that produces a bruit that can be heard by the patient.\[^4\] The sound is most prominent at night when competing noises are absent.

Pulsatile tinnitus with normal otoscopic findings is a common diagnostic dilemma\[^5\]. The differential diagnosis is diverse and includes hydrocephalus, intracranial hypertension, atherosclerosis, and valvular heart disease; further, the condition could occur because of serious vascular malformations such as aneurysms or AVFs.\[^5\] A thorough clinical history and physical examination could help direct the evaluation and management of patients with pulsatile tinnitus who have normal otoscopic findings. Considering the variety of vascular etiologies, pulsatile tinnitus is best evaluated using computed tomography (CT) or MRA.\[^10,11\] In the absence of objective pulsatile tinnitus, MRA is an appropriate initial diagnostic step.\[^6\] If a patient has an objective bruit, the clinician may choose angiography to ensure that pathological vascular abnormalities are not missed. Angiography remains the diagnostic gold standard, because of its high sensitivity and specificity.\[^5,11\]

Pediatric AVFs in the head and neck that produce pulsatile tinnitus are rare. An AVF is 20 times more common in the intracranial vasculature than in the external carotid artery.\[^7\] AVFs involving the great vessels of the neck are usually secondary to penetrating trauma such as missile injuries, stab wounds, or even iatrogenic causes.\[^8\] Congenital AVFs are remarkably rare and usually appear early in life.\[^7,8,12,13\] Pulsatile tinnitus accompanied by an AVF is so unusual that it can be found in only 3% of all fistulous cases and might be an initial clue to suggest a congenital origin.\[^10\] The most common symptoms of congenital neck AVFs include a bruit (83%), a pulsatile mass (67%), and pain (50%). If untreated, a congenital AVF may lead to a number of well-known complications such as cardiac failure, rupture, or emboli. Symptomatic or complicated congenital AVFs that have pain, hemorrhaging, pressure symptoms, ischemic ulceration, or even congestive cardiac failure might necessitate surgi-
cal excision or selective embolization.

In the past, treatment of head and neck AVFs was primarily reliant on surgical excision or ligation of the feeding arteries. For small and superficial lesions, excision alone might be possible, with a lower risk. For more extensive, multi-collateral vascular abnormalities and intracranial lesions, the treatment of choice might involve combination therapy comprising preoperative angiography with selective embolization, followed by definitive resection. Large-scale surgical exploration or craniotomy may be necessary to expose extensive lesions and the adjacent vascular anomalies. More complicated procedures and subsequent bleeding problems incur increased risks. Complications from surgery may include various cranial nerve deficits, vertebral nerve deficits, brain damage, troublesome bleeding, embolic occlusion, hemiplegia, and death.

Conservative treatment with selective embolization and preservation of the feeding artery is an alternative to surgical intervention. With a more isthmus-like single fistula, transcathetercoil embolization was considered to be effective and safe. A detachable balloon is the preferred approach for the occlusion of AVFs that do not interfere with the surrounding normal vasculature and that maintain the patency of the remaining feeding vessel branches. Small diameter coils are especially useful if it is impossible to pass a balloon through the narrow orifice of the fistula or if the venous portion of the fistula is not large enough to inflate the balloon. Finally, after intervention, most patients underwent CT or MRA to assess possible postembolization ischemia, vasogenic edema, infarct, hemorrhage, or coil dislodgement.

Our case had a rare congenital right external carotid artery fistula that presented with pulsatile tinnitus, a carotid bruit, and thrills, as well as nightmares. After precise evaluation, the fistula was completely occluded with 2 coils. Such a simple, narrow fistula that did not involve the distal RECA branches guaranteed the success of coil occlusion and had a low complication risk.

REFERENCES


罕見兒童脈動性耳鳴使用心導管螺旋線圈栓塞術治療成功－病例報告

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我們發表一個四歲的女生，初始的症狀表現出少見的夜間睡眠時右耳單側耳鳴，耳鳴怪異白天清醒時消失，持續了一年之久，導致病患失眠與夜間驚恐不安。經頭頸部磁振血管攝影與心導管攝影檢查後，證實其有先天性右外頸動脈瘻管，瘻管連接右側外頸與內頸靜脈。此先天頸部動靜脈瘻管經由心導管螺旋線圈方式栓塞，術後狀況良好，病患夜間症狀痊癒，無併發症發生。耳鳴是個症狀，不是疾病，每個人在某個時間或情況下都曾感覺，目前缺乏全球耳鳴發生率的統計，在一些報告裡耳鳴在聽力正常小孩發生率約6~36%，而在聽力障礙的小孩上有超過一半的人感到耳鳴。若耳鳴出現規律、且跟自己的心跳一致的話被為脈動性耳鳴(pulsatile tinnitus)。脈動性耳鳴形成原因很多，大多與頭頸部靠近聽覺器官的血管或血流異常有關，它罕見在小孩身上，只佔所有耳鳴的百分之3。

因病患的單側耳鳴僅出現在深夜睡眠時，導致驚恐失眠，經過反覆耳鼻喉科與神經科檢查並無發現相關的腦神經與聽覺器官的障礙缺陷，因而被誤認為身心症候群而受精神科藥物治療。經診斷後，確定是一罕見的兒童先天性右側外頸動脈瘻管導致此夜間耳鳴，完成了心導管的螺旋線圈栓塞治療，術後長期症狀痊癒。過去的文獻裡，僅有少數報告發表兒童脈動性耳鳴導因於先天性動脈瘻管，並成功藉由栓塞治療，對符合適應症的病人，栓塞治療具有良好的治療效果，提供了傳統外科手術治療瘻管之外的另一項選擇。

關鍵字：兒童脈動性耳鳴，先天性動脈瘻管，心導管螺旋線圈栓塞術