3-Tesla magnetic resonance imaging reveals vasculitis-caused otitis media in a patient with giant cell arteritis

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Abstract

Giant cell arteritis (GCA), a medium to large-vessel vasculitis, has a broad spectrum of disease manifestations, including hearing loss. In addition to tissue histology, clinical correlation of symptoms and vascular imaging is optimal for a definitive diagnosis of GCA. Here, we describe an extremely rare case of GCA with hearing loss and otitis media, wherein contrast-enhanced 3D-magnetic resonance imaging was acquired at 3-Tesla for assessment of inflammatory involvement of cranial arteries. Spin-echo imaging demonstrated mural thickening and contrast enhancement of the medial meningeal artery in addition to the superficial temporal artery on the left side. Simultaneously, gradient-echo imaging revealed intense enhancement in the fallopian canal and eustachian tube on the left side, which was associated with non-enhanced effusion in the mastoid cavity. The findings suggest a broad distribution of arterial inflammation in the temporal bone, including small vessel vasculitis, which may cause otologic manifestations in cranial GCA.

Introduction

Giant cell arteritis (GCA) is the most common type of systemic vasculitis, mainly affecting medium- and large-sized vessels. Common manifestations of GCA at presentation include headache, jaw claudication, scalp tenderness, polymyalgia rheumatica (PMR), and hearing loss [1]. In addition, visual manifestations and cerebrovascular accidents are less frequent but clinically important because they potentially leave permanent disability. Due to the low sensitivity of temporal artery biopsy, ranging from 70% to less than 90% [1], vascular imaging methods, including contrast-enhanced magnetic resonance imaging (MRI) at 3-Tesla, have an increasing role in the diagnosis of GCA [2,3].

Previous studies have demonstrated that otologic involvement is rather common in GCA patients, with an incidence ranging from 25% [4] to 53% [5]. Sensorineural hearing loss is predominant and usually concomitant with other ischemic symptoms on the ipsilateral side, including ocular involvement, eye pain, and tenderness of the temporal artery [4,6]. On the other hand, otitis media is extremely rare, and only one report has been published on this topic in literature [7].

In this report, we present findings in a patient with cranial GCA and otitis media. Using a novel imaging protocol that we have developed recently [8], we demonstrate the involvement of small vessel vasculitis in the temporal bone, which may have caused the development of otitis media in this patient.

Case report

A 56-year-old man presented with diplopia, left-sided severe temporal headache, and ipsilateral hearing loss for two months. He had a medical history of diplopia at the age of 40 years, which resolved without treatment. Antiepileptic and anti-coagulant therapy were used for treatment of epilepsy and transient ischemic attack due to occlusion of the left internal carotid artery, respectively, at 42 years of age. Examination of the patient at the time of presentation revealed otitis media with effusion (Figure 1) and a conductive
hearing loss with a large air-bone gap (Figure 2(A)) in the left ear. Furthermore, severe tenderness on palpation was present along the tortuous course of the superficial temporal artery (STA) on the left side. Ophthalmologic examination revealed bilateral abducens nerve paralysis and normal vision. No signs of PMR or extracranial involvement were found. C-reactive protein was elevated to 4.1 mg/dL, and erythrocyte sedimentation rate was 40 mm/h. Antineutrophil cytoplasmic antibodies (ANCA) were absent. The patient was referred for a contrast-enhanced, T1-weighted, 3 D-MRI at 3-Tesla for assessment of inflammatory involvement of the cranial arteries. Fast spin-echo (SE) imaging with fat suppression demonstrated mural inflammation of the STA on the left side (Figure 3(A,B)) with normal findings on the right side (Figure 3(C)), and thromboembolism and wall thickening of the left internal carotid artery (Figure 3(D)). Simultaneously, fast-spoiled gradient-echo (GRE) imaging revealed intense enhancement of the tympanic (Figure 4(A)) and mastoid (Figure 4(B)) segments of the left fallopian canal. Moreover, mural inflammation of the medial meningeal artery (MMA) was evident at the foramen spinosum on SE imaging (Figure 5(A)), which was accompanied by inflammation in the surrounding tissues, including the eustachian tube, on GRE imaging (Figure 5(B)) and simple effusion in the tympanic cleft on T2-weighted MRI (Figure 5(C)). Temporal artery biopsy specimens showed only mural inflammation without granulomatous formation. The patient was clinically diagnosed with cranial GCA and was prescribed 40 mg prednisolone per day, which resulted in a prompt recovery from headache and resolution of hearing loss on audiometry (Figure 2(B)). However, with a gradual attempted decrease in the dose of prednisolone to 25 mg, he experienced a relapse of headache. At the time of the study, the patient was being administered 25 mg prednisolone combined with methotrexate.

Discussion

We report an extremely rare case of cranial GCA that presented otitis media as one of the clinical symptoms. The findings of imaging techniques revealed increased enhancement in the fallopian canal and
mural inflammation of the MMA, which suggested that vasculitis in the temporal bone and helped in the diagnosis of GCA in this patient. The result is important because this is the first report showing radiological evidence of vasculitis causing otologic manifestations in GCA.

Despite the negative results of histopathology of temporal artery biopsy, the diagnosis of this patient was supported by several clinical manifestations. At presentation, he fulfilled three of the five criteria of the 1990 ACR classification criteria for GCA [9], that is, the patient was 50 years or older at the time of onset of the disease, there was evidence of new onset of localized headache, and there was tenderness of the temporal artery. Mural inflammation of the STA was consistent with the symptoms. In addition, the patient showed diplopia due to bilateral abducens nerve paralysis, which is recognized as one of the key symptoms of GCA [10]. Furthermore, he developed epilepsy and transient ischemic attack as the initial symptoms, which were believed to have developed due to left internal carotid artery occlusion. Previous studies have reported that carotid artery lesion is found in 41% patients with GCA [11] and ischemic stroke occurs in 2–16% [12].

Otitis media as an otologic manifestation of GCA is extremely rare, and only one case has been reported in the literature [7]. However, atypical presentations for GCA, including conjunctivitis and lung involvement, suggest that overlap with granulomatosis with polyangiitis. The present case is the first report of GCA with otitis media that was concurrent with temporal headache in terms of both timing and location, and had a negative serological examination of ANCA.

Our results suggest that two possible etiopathogenesis of otologic symptoms in GCA. One possibility is small vessel vasculitis in the fallopian canal. Our recent research shows that intense enhancement of the fallopian canal on contrast-enhanced 3D-GRE...
imaging acquired at 3-Tesla can be a marker for vasculitis in the temporal bone in ANCA-associated vasculitis with otologic manifestations [8]. Two small arteries inside the fallopian canal, the stylomastoid artery and superficial petrosal artery, are the main blood supply for the tympanic and mastoid cavities, and vasculitis of these arteries leads to otitis media. Small-vessel vasculitis occasionally occurs in association with both clinically diagnosed and biopsy-proven GCA [13,14]. On the other hand, our results did not show any enhanced lesions suggestive of inflammation in the tympanic and mastoid cavities, as was found in ANCA-associated vasculitis [8].

Another possible explanation for otologic involvement in GCA is eustachian tube dysfunction secondary to vasculitis of the MMA. Our results clearly showed mural inflammation of the MMA at the foramen spinosum, which was accompanied by contrast enhancement in the eustachian tube. The MMA is a middle-size artery with an average diameter of 2.1 mm at the foramen spinosum [15] and is one of the main blood supplies for the eustachian tube [16]. Intracranial involvement of the MMA is occasionally found in GCA patients [17]. However, eustachian tube dysfunction in GCA has not been reported in the literature. Furthermore, the relationship between vasculitis of the MMA and vasculitis in the fallopian canal is unclear.

Small-vessel involvement in GCA is considered to be associated with severity of disease activity [14]. Moreover, it is reported that severe symptoms, including headache, visual loss, and central nervous system involvement, are more frequent in GCA patients with hearing loss [4]. Uncontrolled headache with a gradual decrease in dosage of prednisolone in our patient was consistent with these facts. Further research is necessary to elucidate the clinical implications of otologic involvement in GCA.

Acknowledgments
The authors thank Editage (www.editage.com) for English language editing. This work was supported by JSPS KAKENHI Grant Numbers 16K11174 and 20K09727.

Consent
Written and informed consent was obtained from the patient regarding the use of his clinical findings and reports of the investigations that were conducted.

Disclosure statement
No potential conflict of interest was reported by the authors.

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