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CASE REPORT - FUNCTIONAL

Microvascular decompression for typewriter tinnitus-case report

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Abstract Background: Microvascular decompression has been tested as a treatment for tinnitus. Methods: However, only a fraction of patients appear to benefit from surgery if the combination of findings such as paroxysmal vertigo, ABR changes and tinnitus is used to select patients for microvascular decompression. Results: Instead, a more specific syndrome of staccato or "typewriter" tinnitus, which is highly responsive to carbamazepine, was suggested to be caused by a neurovascular conflict. Conclusion: We present the first case of typewriter tinnitus with complete long-term symptom relief following microvascular decompression of the vestibulocochlear nerve. We suggest that this specific syndrome is caused by a neurovascular conflict and treatable by microvascular decompression.

Keywords Microvascular decompression · Surgical treatment · Tinnitus · Paroxysmal

Introduction

Microvascular decompression is a successful treatment of a number of compression syndromes that occur following a neurovascular conflict between offending vessels and the trigeminal, facial or glossopharyngeal nerves (reviewed in [13, 17, 20]) in syndromes that also respond to carbamzepine treatment. Jannetta suggested that arterial compression of the vestibulocochlear nerve complex is also a potential pathogenetic mechanism that may affect hearing and equilibrium [10]. He suggested microvascular decompression as a treatment for tinnitus and vertigo. The original selection criteria included unilateral tinnitus, positional aggravation and unilateral hearing deficit [5, 14]. ABR examinations and the occurrance of disabling positional vertigo were used to refine the surgical indications [4, 15]. Still, the benefit of surgery was moderate with improvement in a little more than half of the patients while complications occurred in a quarter of the patients [2, 7–9, 14]. Subsequently, most neurosurgeons are hesitant to treat tinnitus by microvascular decompression and decompression of the eighth nerve is infrequently performed; the selection criteria for treatment are not sufficiently specific.

Recently, a specific form of tinnitus, "typewriter tinnitus" or "staccato tinnitus," was found to be highly responsive to carbamazepine [3, 11, 16, 19]. The condition is characterized by attacks, either spontaneous or precipited by sounds or positioning. The attacks occur as paroxysmal staccato sounds, usually forming a crescendo before termination and described as "clatter," "machine gun," "coins in a can," "crackling" or "typewriter." Levine [11] emphasized the paroxysmal character of the syndrome, reminiscent of hemifacial spasm, tic douloureux and glossopharyngeal neuralgia. In addition, carbamazepine turned out to relieve typewriter tinnitus efficiently. These analogies led to the hypothesis that typewriter tinnitus was likely an effect of a neurovascular conflict, and Levine speculated that microvascular decompression would be effective.

As a surgical indication, tinnitus with positional vertigo is too wide, since many patients do not benefit [4, 5, 14, 15]. Typewriter tinnitus is a condition with strict diagnostic criteria [11] and may comprise a sufficiently specific indication. We have not come across any previously published reports of a

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patient who was operated on by microvascular decompression for the specific syndrome of typewriter tinnitus. This case report describes a patient with typewriter tinnitus who did not tolerate carbamazepine, although he responded, and underwent microvascular decompression with complete long-term symptom relief.

Case presentation

At 54 years of age, this male patient was diagnosed with paroxysmal unilateral staccato tinnitus related to microvascular compression (patient no. 1 in [3]). He had attacks of left-sided tinnitus with a vertical swaying sensation that lasted 10–15 s and were very frequent, sometimes occurring every minute. In between the attacks he had no audiovestibular symptoms. The attacks were spontaneous although more frequent in certain head positions. The tinnitus was described as "coins dropping into a tin can" and the attacks had a crescendo character. During the attacks he experienced oscillopsia and lost his balance. His caloric response was normal. His auditory brainstem response (ABR) and cervical vestibular evoked myogenic potentials (cVEMPs) were also normal with sytmmetric latencies. Further, there were no acoustic emissions during the attacks [3]. MRI revealed a neurovascular conflict between the AICA and the vestibulocochlear nerve on the affected (left) side (Fig. 1). Carbamazepine (100 mg twice daily) almost completely normalized the situation. He was, however, not comfortable with the medication and, although offered the possibility to try other medications, for example, gabapentin [18], he preferred a surgical procedure for relief.

Surgery was undertaken through a left-sided retromastoid craniotomy with the patient in a right lateral position. The vessels and VII–VIII nerve complex were identified and dissected. An AICA (anterior inferior cerebellar artery) loop was identified between the nerves, producing a grayish groove in both nerves (Fig. 2). The vessel and nerves were gently dissected and separated sharply; the vessel loop was mobilized laterally toward the brain stem. A miniature Teflon cushion was inserted between the nerves to provide space for the vessel, and the vessel was fixed in its new position with a Teflon loop glued to the petrous dura with fibrin glue (Fig. 3). Subsequently, the dura was closed in a watertight fashion with a fascial graft; the bone was repositioned and skin closed. The postoperative course was uneventful. The patient reported immediate symptom relief and was discharged home after 4 days.

At 6-week follow-up the patient reported sustained relief from the attacks. His hearing was unchanged, and the audiogram was identical to the preoperative one. At 4-year follow-up he remained symptom free with preserved hearing (Fig. 4). His caloric response at postoperative testing was also normal.

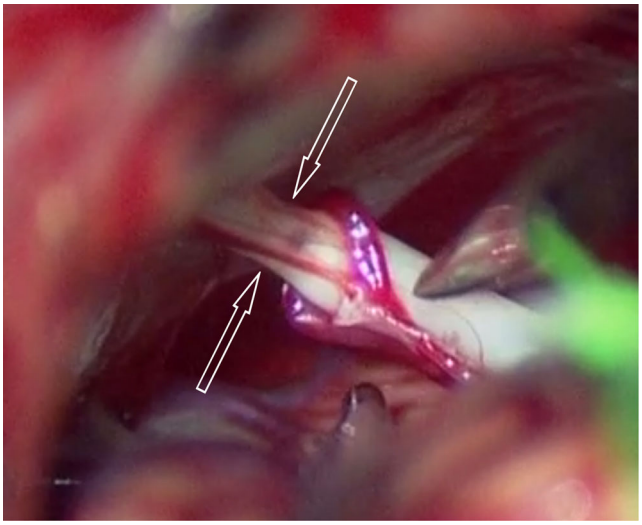

Fig. 2 A surgical photograph following initial mobilization showing the position of the AICA between the nerves and thinning of the VIIIth nerve (*arrows*) at the point of original neurovascular conflict

Fig. 1 Preoperative 3D T2 MRI showing the complex of Nn VII and VIII with a presumed symptomatic conflict with a loop of the anterior inferior cerebellar artery

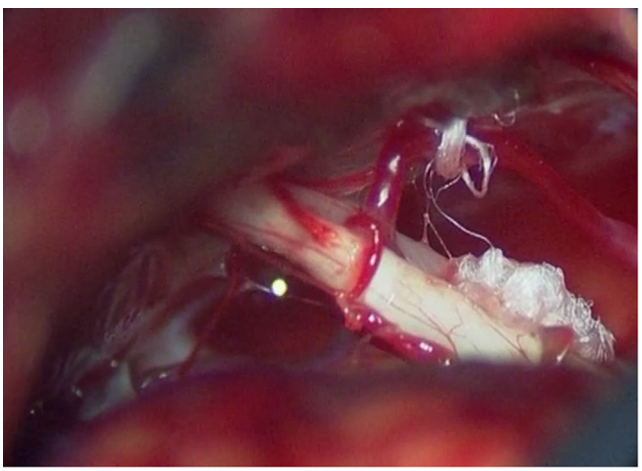

Fig. 3 The AICA loop has been mobilized, moved proximally and attached to the petrous bone via a Teflon loop. A Teflon cushion creates a space between the VIIth and VIIIth nerves

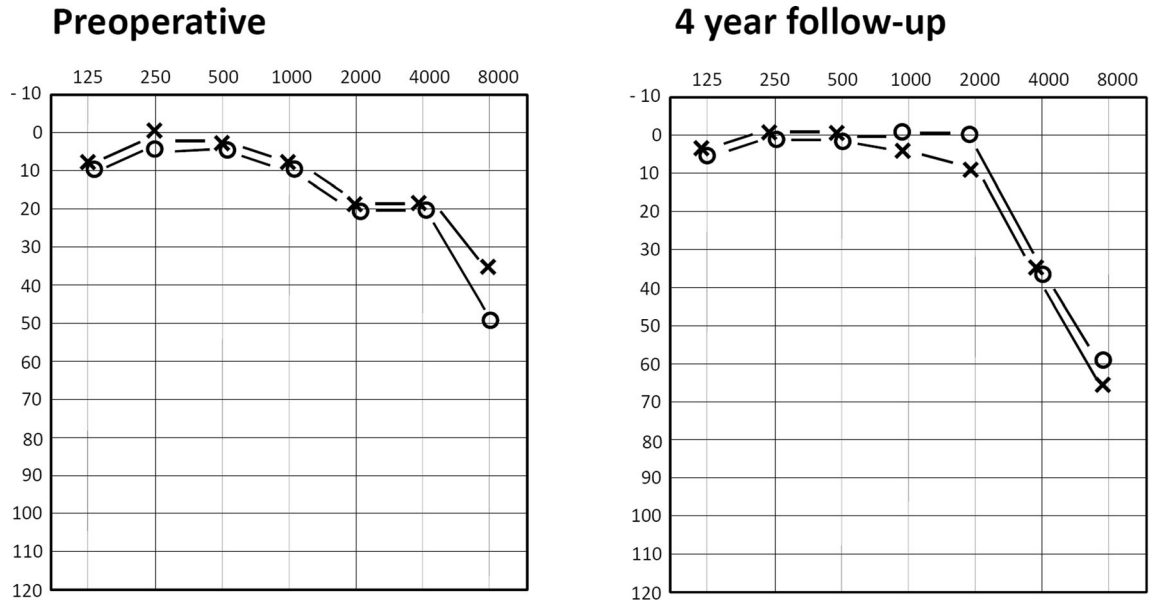

Fig. 4 Audiogram before surgery (*left*) and at the 4-year follow-up (*right*)

Discussion

This is to our knowledge the first reported patient who underwent microsurgical decompression for the specific syndrome of typewriter tinnitus. The potential specificity of the surgical indication was important, as it may finally provide an indication of tinnitus where successful surgical results can be expected in the majority of operated cases. Our patient experienced immediate symptom relief without a hearing deficit, a condition that was retained during long-term follow-up.

The syndrome, which comprises a well-defined subgroup of tinnitus patients, responds uniformly to carbamazepine. The analogy with tic douloureux and hemifacial spasm suggested that microvascular decompression would be effective; our case provided a proof of concept. With only one case, we lack evidence in an epidemiological sense. However, previous experience in the pathogenetic mechanisms of neurovascular conflicts and typewrite tinnitus suggests a coherent causal theory of typewriter tinnitus, a theory that was corroborated by the results in our patient. Intraoperatively, direct contact by the arterial loop and a discolored groove on the nerves indicated significant compression. Symptoms were relieved following the decompression. Another discriminative feature is the rapid response. Our patient improved immediately after surgery, and the response to carbamazepine is rapid in this patient group. In contrast, the general tinnitus patients that benefit from surgery improve only slowly [6]. We thus infer that the data can be generalized and suggest that microvascular decompression is an effective treatment for this category of patients. Additional cases may strengthen the finding. It is, however, scientifically more important to attempt to falsify the hypothesis by finding patients with typewriter tinnitus who may be unresponsive to carbamazepine or may not have a neurovascular conflict.

In this patient, surgery presented specific hazards. An AICA loop was positioned between the VIIth and VIIIth nerves, an anatomically common condition. It was thus not feasible to reposition the loop away from the nerves as is usually done in decompression of the Vth, VIIth and IXth nerves [13, 17, 20]. Instead, we gently separated the nerves, taking care not to lose the arachnoidal planes, pull on the nerves or bruise them. In this patient, the creation of a space between the nerves was successful, as was the relief of pressure from the arteries. We cannot know whether there would be safer or more effective surgical solutions, but consider that this technical approach can function. We did, however, consider that there were risks of injury to both nerves during the dissection.

The condition of typewriter tinnitus is rare, although many cases may go undetected; the syndrome has been described only recently [11]. How could tinnitus patients be identified who would potentially benefit from microvascular decompression of the N VIII nerve? Relying on MRI is questionable, because neurovascular conflicts are frequently imaged in asymptomatic cases [1]. Tinnitus is a very common symptom in the general population, often bilateral, but strictly unilateral tinnitus is also frequent. A history of repeated brief attacks of paroxysmal unilateral staccato tinnitus is, however, rare and does indeed suggest a typewriter tinnitus syndrome. Nevertheless, the key finding for identifying these patients is probably the prompt response, within a few days, even to a low dosage of carbamazepine. In contrast, carbamazepine is a medication that is not effective for the general population of tinnitus patients [8]. Finally, the effectiveness of carbamazepine provides support for the idea that typewriter tinnitus syndrome is a hyperactivity dysfunction symptom of N VIII. Another possibility, which we have not yet explored, is that

carbamazepin challenge in patients with other kinds of unilateral tinnitus may identify a subgroup of patients who may benefit from microvascular decompression. Endocochlear hydrops or myoclonic attacks of the stapedius could be considered as differential diagnoses to typewriter tinnitus. For several reasons, we consider typewriter tinnitus as a separate condition and inferred that our patient suffered from this condition rather than stapedius myoclonus or endocochlear hydrops. Acoustic emissions were not present during the attacks, as would have been expected from stapedius myoclonus. Furthermore, a patient with symptoms reminiscent of "typewriter tinnitus" was operated on with sectioning of the stapedius tendon, but without any symptom relief [12]. Endocoochlear hydrops, as with Menieres disease, would be expected to cause more long-lasting hearing deficit and vertigo attacks. This condition would not respond to carbamazepine.

Our report emphasizes that tinnitus is a symptom rather than one disease entity; refined diagnostic workup can identify patients with conditions that cause tinnitus where causal therapy is possible. We conclude that typewriter tinnitus is one such specific syndrome with stereotypic symptoms that are caused by a neurovascular conflict and have presented the first report of a patient who underwent surgical treatment with long-term cure for the condition.

The patient has consented to the submission of the case report for the journal.

Conflicts of interest All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements) or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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