

Case Report

“Emergency” Cochlear Implantation in Labyrinthitis Ossificans Secondary to Polyarteritis Nodosa: How to Face a Rare Entity

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Polyarteritis nodosa (PAN) is a systemic vasculitis affecting the small- and medium-sized arteries that may present with hearing impairment. In rare cases, PAN may be associated with progressive labyrinthitis ossificans (LO), an otologic emergency requiring early cochlear implantation (CI) to restore hearing before the complete, irreversible cochlear ossification. We report the first case in the literature of a patient affected by PAN with bilateral sudden sensorineural hearing loss and rapid LO who underwent “emergency” bilateral simultaneous CI. This case report emphasizes the importance of an early audiological evaluation in patients with PAN when LO is suspected. Multidisciplinary approach is mandatory when facing organ-specific manifestations in patients with PAN. Detailed discussion is provided with particular regard to clinical and radiological presentation as well as CI outcomes in such a rare and challenging case.

KEYWORDS: Polyarteritis nodosa, cochlear implant, sudden sensorineural hearing loss, labyrinthitis ossificans

INTRODUCTION

Polyarteritis nodosa (PAN), first described by Kussmaul and Maier ^[1] in 1866, is a systemic vasculitis affecting the small- and medium-sized arteries. PAN has a prevalence of 6.3/100,000 and an annual incidence of 0.7/100,000; clinical manifestations of the disease mainly occur in the 5th and 6th decades of life ^[2]. The pathogenesis of PAN remains unclear, although the clinical response to immunosuppressive therapy suggests an active pathogenic role by immunological mechanisms. The disease is characterized by a multi-organ involvement, particularly kidneys (70%), skeletal muscles (60%), skin (40%), heart (35%), and central nervous system (25%). Facial nerve weakness and hearing impairment have been reported in patients with PAN, and, in rare cases, they may represent early manifestations of the disease ^[2]. Hearing loss is usually bilateral and symmetrical; it is mainly sensorineural with sudden or progressive course although cases of mixed or conductive hearing loss have been described ^[2]. Progressive labyrinthitis ossificans (LO), a reactive response to the inflammatory vasculitis typical of PAN that can determine the formation of pathological fibrous tissue or new-bone formation in the inner ear, can be found in patients with PAN and other audiological symptoms ^[3].

We report the case of a patient affected by PAN with bilateral sudden sensorineural hearing loss and rapid LO who underwent bilateral simultaneous cochlear implantation (CI) within a short time from diagnosis to prevent ongoing fibrotic degeneration of the labyrinth. To the best of our knowledge, this is the first case of emergency CI due to a rapid progressive LO secondary to PAN. Clinical presentation and diagnostic and therapeutic issues will be discussed, focusing on the key role played by the multidisciplinary approach in such a rare and challenging case.

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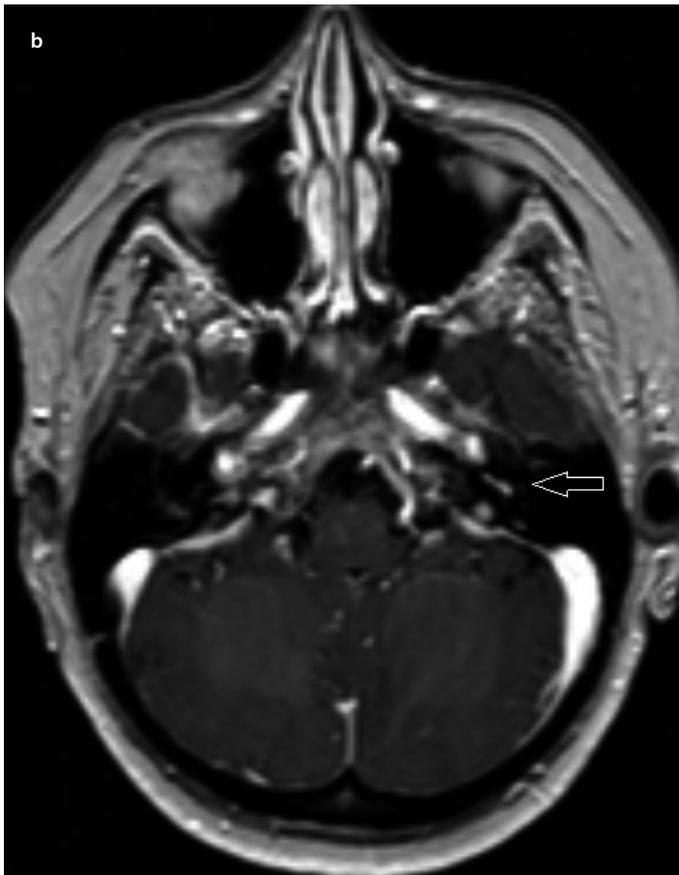
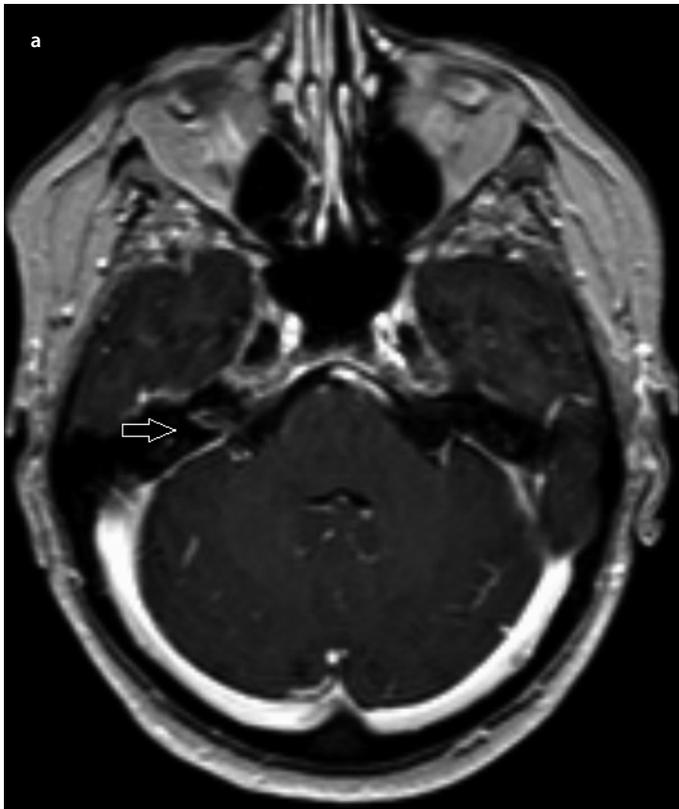


Figure 1. a, b. Axial T1-weighted MRI shows contrast enhancement of the (a) right internal auditory canal and (b) basal turn of the left cochlea.

CASE PRESENTATION

A 53-year-old woman was admitted to the neurology clinic of a local hospital reporting sudden onset of right facial nerve palsy, pain and stiff neck, sudden bilateral hearing loss, and dizziness. On presentation, blood examination showed neutrophil leukocytosis (13,600/mcL total white blood cell with 76% of neutrophils), thrombocytosis (524,000/mcL platelets), and high levels of inflammatory markers (erythrocyte sedimentation rate and C-reactive protein were 108 mm/h and 13.3 mg/dL, respectively). Urine, blood, and cerebrospinal fluid cultural examinations were unremarkable, and the autoimmune blood panel was negative. Magnetic resonance imaging (MRI-Intera® 1.5 T; Philips Medical Systems, Amsterdam, Netherlands) showed contrast enhancement of the meninges in the subtentorial region, on the right acoustic-facial bundle and on the left basal turn of the cochlea; no evidence of white matter hyperintensities, characteristics of cerebral vascular disease, was observed (Figure 1). The patient was treated for 3 consecutive weeks with high oral corticosteroid therapy with prednisone, 1 mg/kg/day (Deltacortene®, Bruno Farmaceutici S.p.A., Rome, Italy); however, no substantial improvement of the clinical symptoms was observed. Shortly after, the patient developed livedo reticularis of the extremities and acrocyanosis suggestive of peripheral vascular involvement. The patient was hospitalized in the rheumatology clinic of our institution; angiographic studies revealed multiple aneurysms of medium-sized visceral arteries and occlusions/stenosis of the tibial arteries. A diagnosis of PAN was made according to the criteria of the American College of Rheumatology [4]. The patient was positive to hepatitis C virus (HCV); an association between HCV and PAN has been previously reported, and it often leads to a more severe and acute clinical presentation and a higher rate of clinical remission [5]. Owing to the severe and worsening bilateral hearing impairment reported by the patient, an otolaryngology examination was performed. Otomicroscopy examination and tympanometry were bilaterally normal. Au-

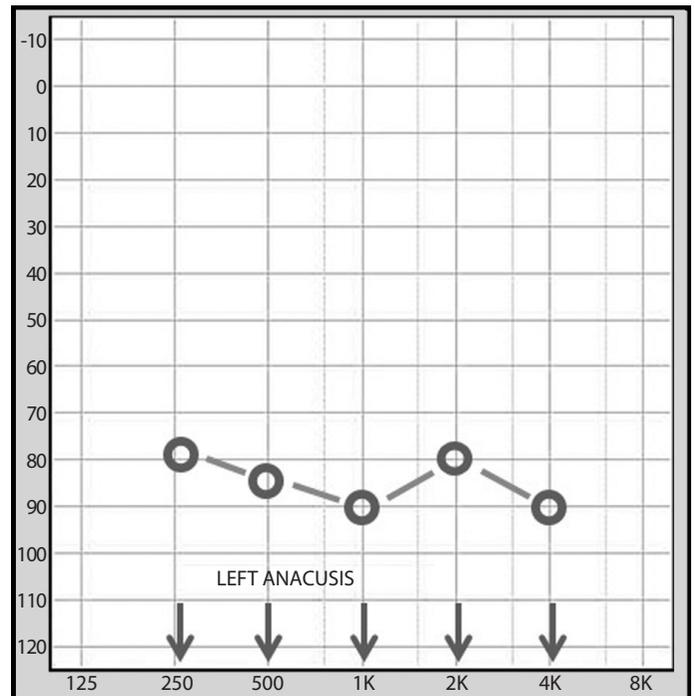


Figure 2. Preoperative pure-tone audiometry showing left anacusis and right severe hearing loss.

diometry showed left anacusis and right-sided severe hearing loss with pure-tone average (PTA) in the 0.5-2 kHz frequency range of 85 dB hearing level (HL), without effective discrimination at speech audiometry (Figure 2). Bed-side vestibular examination did not reveal spontaneous or evoked nystagmus, the head-shaking test was negative, and the Halmagyi test was bilaterally positive. Bilateral vestibular areflexia (Dandy's syndrome) was found at caloric stimulation. The patient underwent computed tomography (CT—Aquilion One®; Toshiba Medical Systems Corporation, Tokyo, Japan) and MRI scans (Magnetom Symphony®1.5T; Siemens Healthineers, Munich, Germany) to investigate inner ear involvement. Approximately 45 days after the clinical onset, the CT and MRI findings were consistent with a fibrous process involving the basal turn of the left cochlea and the vestibular portion of the labyrinth bilaterally (Figure 3). A simultaneous bilateral CI (Digisonic SP®; Oticon Medical, Smorum, Denmark)

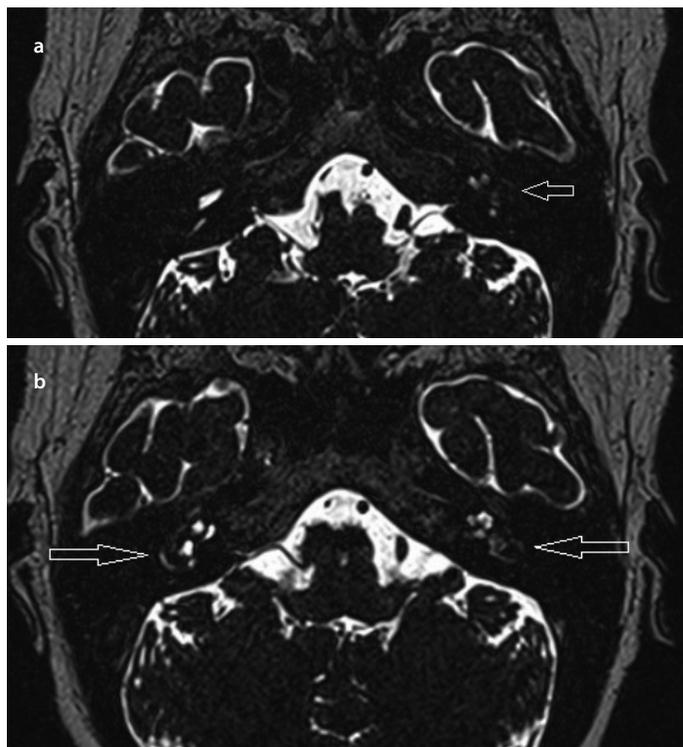


Figure 3. a, b. Axial T2-weighted MRI outlines the loss of signal of the (a) left basal turn of the cochlea and (b) posterior labyrinth bilaterally, consistent with fibrous degeneration.

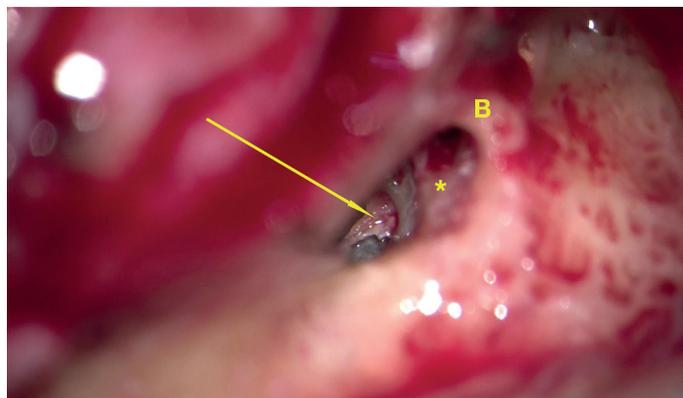


Figure 4. Intraoperative view of the posterior tympanotomy (left ear). Fibrous tissue through round window cochleostomy (arrow). B: buttress. *Pyramidal process with the stapedial tendon.

was immediately attempted to prevent the progression of inner ear fibrous-ossification according to current recommendations [6]. The right CI was performed uneventfully with the complete insertion of all electrodes (20 out of 20). The surgical procedure on the left side was complicated by the intraoperative finding of fibrous tissue obliterating the round window region and the basal turn of the cochlea limiting the complete insertion of the CI array (2 excluded electrodes out of 20) (Figure 4). The patient was discharged approximately 4 weeks after the admission with chronic immunosuppressive therapy with prednisone (Deltacortene®) and methotrexate (Metotrexato Sandoz®, Sandoz Industrial Products S.p.A., Holzkirchen, Germany). On 18months of follow-up, the patient showed complete facial nerve recovery and significantly improved hearing performances, with a PTA of 40 dB HL on the right side (speech recognition score in quiet: 80% at 70 dB HL) and 60 dB HL on the left side (no speech recognition score), with binaural speech recognition score in quiet of 90% at 70 dB HL. The worse audiological performance on the left side resulted by the progressive inactivation of 15 electrodes due to the increasing electric impedance probably secondary to the ongoing inner ear degeneration (Figure 5). All of the described diagnostic and therapeutic procedures were performed after obtaining a signed informed consent by the patient.

DISCUSSION

Labyrinthitis ossificans (LO) represents an otologic emergency requiring early CI to restore hearing before the complete, irreversible cochlear ossification [7]. The etiology of LO comprises infectious, traumatic, inflammatory, dystrophic, and tumoral processes. Even if meningitis represents the most common cause of LO, Quaranta et al. [8] found LO as a nearly-constant consequence of inner ear involvement due to unspecified arteritis. Nevertheless, PAN represents one of the most common non-organ-specific autoimmune diseases that involve the inner ear [9]. Human temporal bone studies outlined the histopathological alterations occurring in PAN supporting the devel-

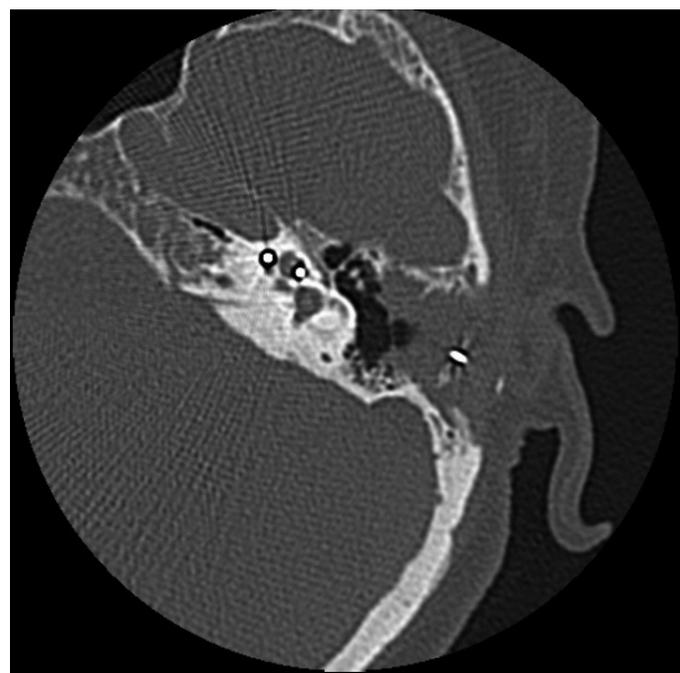


Figure 5. CT scan of the left ear shows the progression of LO in the left ear at 6months of follow-up. It is possible to appreciate the advanced ossification of the lateral semicircular canal.

opment of LO ^[10]. We report the first case in the literature of rapidly progressive LO refractory to medical treatment that led to an emergency CI at the clinical onset of the disease. The typical clinical presentation of patients with PAN is unlikely to involve the otolaryngologist at diagnosis. In this case, the diagnostic delay for the otologic aspects was due to a lack of multidisciplinary discussion during the first days of hospitalization. However, the prompt rheumatologic attention to the audiological issue prevented an irreversible deafness condition due to LO. The effectiveness of CI in patients with PAN has been previously reported in one case of slowly progressive hearing loss developed over a period of 4 years, without radiological and intraoperative evidence of LO ^[10]. The audiological outcomes found in our case pointed out a dramatic improvement of hearing function on the right side, with poor results on the contralateral side. The limited benefits on the left ear might be explained by the partial CI insertion and by the subsequent inactivation of the electrodes due to the advanced degree of LO.

CONCLUSION

Our case report emphasizes the importance of a multidisciplinary approach at the onset of organ-specific manifestations in patients with PAN. Specifically, LO may be suspected in case of sudden hearing loss unresponsive to medical therapy, and “emergency” CI should follow when radiological signs of LO are found. All physicians involved in PAN management should be aware that “timing” represents the key element influencing the CI outcomes.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

Peer-review: Externally peer-reviewed.

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