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CLINICAL RECORD

Sudden sensorineural hearing loss as the first manifestation of chronic myeloid leukaemia: case report

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Abstract

Background: Sudden sensorineural hearing loss rarely occurs in patients with chronic myeloid leukaemia.

Case report: We present a case report of a patient who presented with sudden sensorineural hearing loss as the first manifestation of chronic myeloid leukaemia, and review the mechanisms responsible for sudden sensorineural hearing loss in leukaemic patients.

Results: A 31-year-old female presented to our clinic with unilateral sudden sensorineural hearing loss and tinnitus. Pure tone audiometry revealed profound sensorineural hearing loss in the left ear at all frequencies. During an investigation into her hearing loss, the patient was found to have chronic myeloid leukaemia.

Conclusion: Every case of sudden sensorineural hearing loss must be carefully evaluated, and haematological disorders must be considered in the differential diagnosis of sudden hearing loss.

Key words: Hearing Loss, Sudden; Tinnitus; Leukaemia, Chronic Myeloid; Hyperbaric Oxygenation

Introduction

Chronic myeloid leukaemia is a myeloproliferative disease resulting from the clonal expansion of haematopoietic progenitor cells. It is clinically characterised by myeloid hyperplasia, leukocytosis with basophilia, and splenomegaly.[1] Common symptoms are bone pain, weight loss, excess sweating, fatigue, and early satiety and abdominal discomfort related to splenomegaly. Less common features relate to granulocyte or platelet dysfunction associated with infection, thrombosis or bleeding. Patients occasionally present with leukostatic manifestations such as mental status changes, headache, visual disturbance, stroke, cerebellar signs and priapism. Acute or chronic myeloid leukaemia rarely affects the ear. Sudden sensorineural hearing loss (SNHL) is very rarely the first manifestation of haematological diseases. We report our experience of a 31-year-old female with unilateral sudden SNHL as the first sign of chronic myeloid leukaemia.

Case report

A 31-year-old female presented to our clinic with unilateral sudden hearing loss, tinnitus, vertigo and nausea for 12 hours. Hearing loss was described as sudden onset and was reported to impair social activity. It was not associated with otalgia or otorrhoea. No previous history of otological trauma, drug intake, noise exposure or upper airway infection was noted. The patient had no history of systemic infection or surgery, and nystagmus was not observed. Systemic physical examination revealed a moderately enlarged spleen and mild hepatomegaly. Examinations showed that mental function, the motor system, the sensory system and all cranial nerves except for the VIIIth were normal.

Both the external auditory canal and tympanic membranes were assessed as normal. The response to the Weber tuning fork test was lateralised to the right side. An audiometric examination showed profound SNHL in the left ear; an audiogram of the right ear was normal (10 dB HL; Figure 1). Magnetic resonance imaging of the temporal bone was normal (Figure 2).

Laboratory investigations revealed profound leukocytosis (white blood cell count 264×10^9/l) with mild anaemia (haemoglobin level 98 g/l). Coagulation tests were normal. The patient was referred to the haematology department for investigation of the leukocytosis. Abundant granulocytic cells at all levels of maturity were seen by microscopic examination of a peripheral blood smear. The differential white blood cell counts were 38 per cent neutrophils, 30 per cent band cells, 10 per cent metamyelocytes, 5 per cent eosinophils, 5 per cent myelocytes, 3 per cent promyelocytes, 2 per cent lymphocytes, 2 per cent monocytes and no basophils. Bone marrow aspiration and a biopsy were carried out to provide material for the pathological and cytogenetic investigation of a suspected myeloproliferative disorder. Bone marrow aspiration findings suggested chronic lymphocytic leukaemia with myeloid hyperplasia, increased promyelocytes and 3 per cent blast cells. In addition, chromosomal analysis was positive for the t(9; 22) translocation. Fluorescence in situ hybridisation identified the BCR–ABL gene fusion. Based on these findings, the patient was diagnosed with chronic lymphocytic leukaemia.

To treat deafness, an intratympanic steroid (8 mg dexamethasone in a 1–2 ml volume) was administered into the left ear under local anaesthesia six times in one month.

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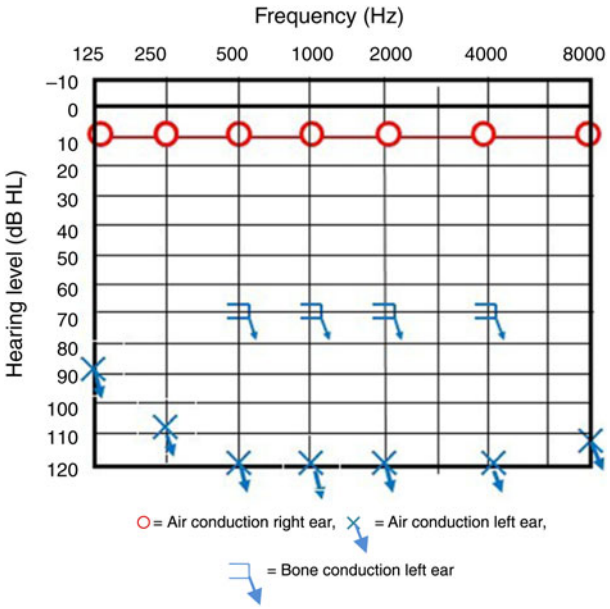

FIG. 1
Pure tone audiometry showing total sensorineural hearing loss in the left ear and normal hearing in the right ear

Hyperbaric oxygen therapy was administered 10 times and an intravenous injection of 25 mg *Ginkgo biloba* extract was prescribed once per day for two weeks to treat sudden SNHL. There was no clinical or audiological change in the patient's hearing on the left side six months after her initial visit. Although the intensity of tinnitus decreased, it has continued. Hydroxyurea therapy was started by the haematology department to reduce the leukocyte count, and symptoms associated with leukostasis disappeared gradually. After hydroxyurea therapy, 0.4–0.8 g/day imatinib was added to a chemotherapy regimen for 29 days. After one month of therapy, the total leukocyte count was within the normal range, and this continued after six months of follow up. Currently, the patient continues to take 0.4 g/day imatinib.

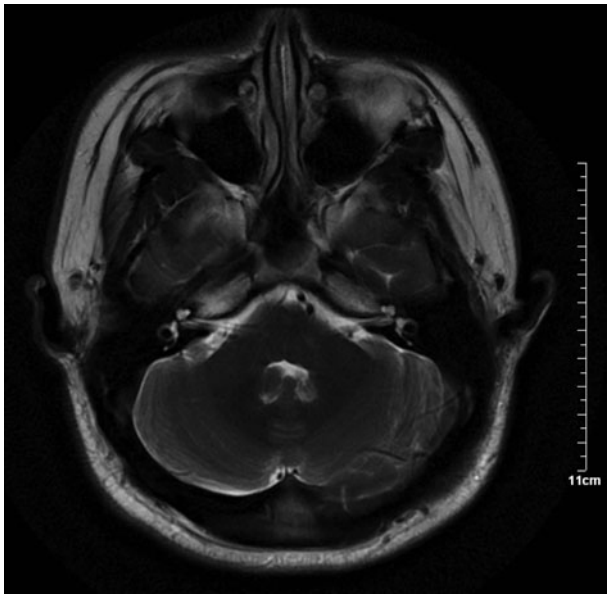

FIG. 2
Magnetic resonance image of the temporal bone showing an absence of cochlear pathology

Discussion

Sudden SNHL is defined as SNHL of at least 30 dB over three consecutive frequencies in the 0.125–8 kHz range compared with the contralateral side and that occurs over 3 days. The cause of sudden hearing loss is usually unknown. However, the cause of sudden SNHL can sometimes be identified, and a specific treatment initiated. The aetiology of sudden SNHL is classified into eight categories: viral and infectious; autoimmune; traumatic; vascular; neurological; tumoural; ototoxic; and pressure related.[2] Sudden SNHL can present as an isolated problem, as the presenting symptom of systemic disease or during the course of an established disease.

Deafness in leukaemia was first described by Donne in 1844.[3] Otological findings, for instance, sudden hearing loss, vertigo, tinnitus, facial weakness and infection, have been described in 16–40 per cent of leukaemic patients.[4,5] Sudden-onset deafness is a rare clinical manifestation of chronic myeloid leukaemia.[6] However, sudden SNHL and tinnitus in the left ear were the first findings associated with chronic myeloid leukaemia in our patient.

The pathogenesis of hearing loss in leukaemia patients is complex. Studies have revealed four main categories of histopathological changes to the temporal bones: leukaemic infiltration; haemorrhage; infection; and hyperviscosity.[7–9] However, the biological mechanism driving leukostasis remains unclear. It was traditionally thought to be related to leukaemic cell 'overcrowding' in the capillaries comprising the microcirculation. Such overcrowding could lead to the formation of small leukocytic aggregates and thrombus, resulting in infarction in different tissues. The inner ear is supplied by the labyrinthine artery (a branch of the anterior inferior cerebellar artery), the small diameter and tortuosity of which predispose it to the effects of hyperviscosity.[6] Some investigations have indicated that leukostasis might also result from endothelial damage.[10,11]

- **Sudden sensorineural hearing loss (SNHL) is a syndrome but not a diagnosis**
- **Every case of sudden SNHL must be carefully evaluated**
- **We report a rare case of sudden SNHL as the initial manifestation of chronic myeloid leukaemia**
- **Haematological disorders should be considered in the differential diagnosis of sudden hearing loss**

Our patient presented with sudden SNHL as a clinical manifestation of hyperleukocytosis of chronic myeloid leukaemia. Most symptoms associated with leukostasis, such as enlarged spleen and mild hepatomegaly, are reversible following a rapid reduction in the leukocyte count. However, there was no change in the patient's hearing on the left side following combined treatment with intratympanic steroid, hyperbaric oxygen and *Ginkgo biloba* extract. This finding suggests that her deafness was probably caused by leukostasis with occlusion of the labyrinthine artery, resulting in irreversible hearing loss despite a rapid reduction in the leukocyte count. Corticoid administration to treat a haematological disorder can trigger a leukaemoid reaction.[12] However, local

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steroid therapy did not adversely influence the progression of chronic myeloid leukaemia.

In some cases, sudden SNHL in chronic myeloid leukaemia may be improved by leukapheresis and chemotherapy. This has been achieved in some hyperviscosity syndrome patients, suggesting that this type of deafness is reversible.[1,13] Unfortunately, we did not observe an improvement in our patient's hearing. In this case, hearing loss was probably caused by hyperleukocytosis with leukostasis and occlusion of the labyrinthine and other small arteries of the vertebrobasilar area.

Conclusion

This study reports a patient who presented with unilateral sudden SNHL as the first manifestation of chronic myeloid leukaemia. The existence of such cases indicates the importance of identifying possible underlying diseases before diagnosing idiopathic sudden deafness. Sudden SNHL may occur as a complication of haematological malignancies such as chronic myeloid leukaemia. In this case, the underlying disease was controlled but with minimal improvement in hearing.

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