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Sudden hearing loss in chronic myelogenous leukaemia implicating the hyperviscosity syndrome

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Abstract

Sudden sensorineural hearing loss that presents as the initial sign of haematological disease is very rare. Chronic myelogenous leukaemia has been implicated as a causative factor of sudden sensorineural hearing loss.

A 49-year-old male presented with unilateral sudden sensorineural hearing loss. The patient was found to have chronic myelogenous leukaemia during a work-up for his hearing loss. We present a case of a chronic myelogenous leukaemia patient whose first manifestation was sudden sensorineural hearing loss. We presume that cochlear vessel occlusion as a result of elevated blood viscosity was responsible for this patient's hearing loss. Early onset of sudden deafness in a chronic myelogenous leukaemia patient may be due to the hyperviscosity syndrome and it may be possible to reverse hearing loss through early leukapheresis.

Key words: Hearing Loss, Sensorineural; Blood Viscosity; Leukaemia, Myeloid, Chronic

Introduction

The incidence of acute-onset sensorineural hearing loss as the first sign of leukaemia is extremely rare. A variety of haematologic diseases such as leukaemia, multiple myeloma,[1] and Waldenstrom's macroglobulinaemia[2] are among the causes of sudden-onset deafness. Hyperviscosity syndrome may be responsible for neurological signs and symptoms attributable to elevated blood viscosity.[3] It is presumed that the abnormally elevated cytocrit causes partial occlusion of the vessels supplying the cochlea causing ischaemia of the cochlea, resulting in hearing loss. Although it has been suggested that lowering the blood viscosity by lowering the cytocrit is the most important therapeutic modality,[4] reversal of sensorineural hearing loss in leukaemic patients has rarely been reported.

In this report we describe a patient with chronic myelogenous leukaemia whose initial symptom was sudden unilateral hearing impairment, which resolved after leukapheresis and chemotherapy without steroids. Therefore, hyperviscosity syndrome as a result of increased cytocrit may be an important cause of sudden sensorineural hearing loss in CML patients, and it may be possible to reverse the hearing loss through early leukapheresis.

Case report

A 49-year-old male patient presented with right-sided hearing loss of one day's duration accompanied by right-sided tinnitus and ear fullness, and generalized weakness. Past history and family history were unremarkable. On physical examination, both tympanic membranes were intact, and both external auditory canals were clear. Results from other otolaryngological examinations were unremarkable. Audiological evaluation revealed an air conduction threshold of 69 dB, and bone conduction threshold of 56 dB (Figure 1).

The patient was admitted because of the sudden sensorineural hearing loss. Computed tomography (CT) of the temporal bone and brain magnetic resonance imaging (MRI) were normal (Figure 2). Speech reception thresholds were 75 dB on the right, and 25 dB on the left. Speech discrimination was 85 per cent on the right and 100 per cent on the left. The Weber test lateralized to the left and both ears were positive on the Rinne test.

An initial laboratory examination revealed a WBC count of 4.851×10^{11}/L, with seven per cent segmented neutrophils, 20 per cent banded neutrophils, 0 per cent lymphocytes, 0 per cent monocytes, 0 per cent eosinophils, one per cent basophils, 57 per cent metamyelocytes, 11 per cent myelocytes, one per cent promyelocytes, and three per cent blasts. Haemoglobin was 11.0 g/dl, haematocrit was 32.5 per cent and platelet count was 3.65×10^{11}/L. Coagulation studies showed activated partial thromboplas-

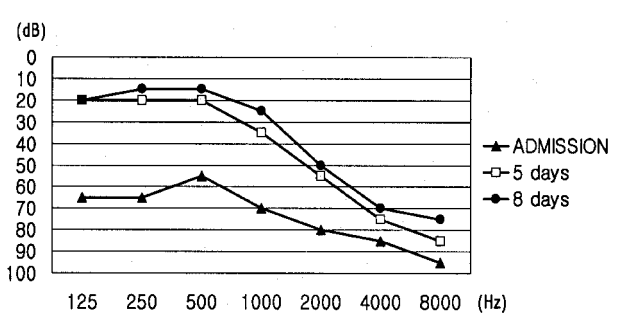

Fig. 1

Audiogram obtained at the first examination, five days and eight days after treatment

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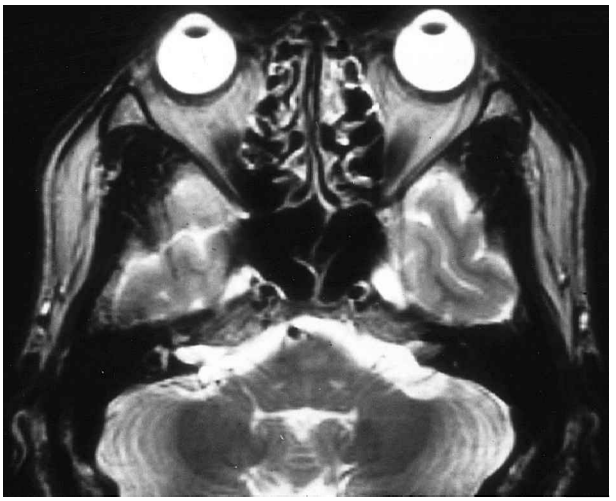

FIG. 2
T2-weighted MRI scan showing no detectable abnormality of the inner ear.

tin time (APTT) 41.5 seconds, prothrombin time (PT) 13.5 seconds, and bleeding time (BT) two minutes. Lactate dehydrogenase (LDH) was increased at 1:521 IU/L, suggesting myeloid proliferation. The patient was transferred to the department of haemato-oncology with suspected leukaemia. Chronic myelogenous leukaemia was diagnosed by a bone marrow biopsy. Chromosomal analysis revealed the translocation t(9;22), confirming the presence of the Philadelphia chromosome and the diagnosis of chronic myelogenous leukaemia.

Leukapheresis was instituted for three days. The patient was additionally treated with chemotherapy consisting of interferon α (three million–six million IU/day) and hydroxyurea (0.3–4 g/day) for 23 days. Steroids were not included in the chemotherapy regimen. After five days of therapy, the WBC count fell from 4.851×10^{11}/L to 2.942×10^{11}/L. On pure tone audiometry, the air conduction was 36 dB, and bone conduction 28 dB. Pure tone thresholds in the low and medium frequency range in particular were markedly improved (Figure 1). Ear fullness and tinnitis resolved with improvement in the hearing level. On the eighth day of chemotherapy, the WBC count fell further to 4.08×10^{10}/L, but there was no further improvement in the hearing level (Figure 1). The patient was discharged after 23 days of chemotherapy. A follow-up complete blood count four months after discharge showed the WBC count to be within the normal range at 5.8×10^{9}/L, but the hearing threshold level of the right ear was similar to that performed shortly before discharge at 36 dB air conduction, and 28 dB bone conduction.

Comment

Although it has been reported that between 16 per cent[5] and 40 per cent[6] of leukaemic patients have otolaryngological signs and symptoms such as sudden sensorineural hearing loss, vertigo, tinnitus, facial weakness and infection,[5,6] sudden sensorineural hearing loss as the first sign of leukaemia is extremely rare. These sequelae were initially believed to be associated more frequently with patients with acute lymphocytic leukaemia.[6] However, recent studies have implicated both acute and chronic leukaemia in the development of sudden sensorineural hearing loss.[7,8] Sudden sensorineural hearing loss, ear fullness, and tinnitus were the first symptoms of leukaemia in our patient.

Numerous studies have demonstrated histopathological changes in the temporal bones of patients with leukaemia. These include leukaemic infiltration,[5,6] inner ear haemorrhage,[5,6,9] infection,[5,6] and the hyperviscosity syndrome.[3,4] Sensorineural hearing loss attributable to the hyperviscosity syndrome may be seen in other blood dyscrasias with increased blood viscosity such as multiple myeloma and Waldenstrom's macroglobulinaemia.[2] Hyperviscosity syndrome is a constellation of signs and symptoms attributable to elevated blood viscosity. These include headache, dizziness, vertigo, hearing loss, visual disturbances, nystagmus, retinal haemorrhages, and congestive heart failure.[4] The hyperviscosity syndrome is often dependent on leukocyte counts greater than 5.0×10^{11}/L. Our patient's initial leukocrit of 4.851×10^{11}/L approximates the leukocyte count reported in the literature that causes the hyperviscosity syndrome.[11–13] Whole blood viscosity is a function of serum viscosity, haematocrit and leukocrit. The viscosity of leukocyte suspensions is greater than the viscosity of erythrocyte suspensions of an identical cytocrit because of the greater rigidity of leukocytes.[11–13] Occlusion of the very small-calibre labyrinthine artery due to hyperviscosity syndrome is thought to be responsible for the sudden sensorineural hearing loss and tinnitus.

Management of hyperleukotic chronic myelogenous leukaemia with hyperviscosity syndrome consists of lowering the leukocyte count to reverse the symptoms and signs of hyperviscosity.[4] Since response to corticosteroids and/or alkylating agents may occur only after six to 12 weeks of treatment, leukapheresis may be employed to lower leukocyte counts quickly with a rapid response in the neurological signs and symptoms.[4] We believe that reversal of the hearing loss in our patient was possible because it was possible to lower the leukocyte counts quickly through leukapheresis instituted within 48 hours after the onset of the hearing loss. Reasons implicated for non-reversible hearing loss may include leukaemic infiltration into the middle ear and inner ear infarction.[10] If sudden sensorineural hearing loss and tinnitus were a prelude to inner ear infarction due to the hyperviscosity syndrome, early diagnosis and treatment with leukapheresis and chemotherapy are imperative to lower the risk of deafness due to leukaemia.[10] We believe that in our case, decreased blood viscosity as a result of leukapheresis and chemotherapy restored cochlear vessel circulation, thus relieving the ischaemic state of the cochlea. Improvement in sudden deafness due to steroids could be ruled out because no steroid was included in the chemotherapy regimen.

In conclusion, we were able to improve the hearing of a chronic myelogenous leukaemia patient who presented with sudden sensorineural hearing loss through leukapheresis. Early onset of sudden deafness in a chronic myelogenous leukaemia patient may be due to the hyperviscosity syndrome, and it may be possible to reverse the hearing loss in these patients through early leukapheresis.

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