

CASE REPORT

Bilateral Sudden Sensorineural Hearing Loss as a First Sign of Chronic Myeloid Leukemia

Fatma Tulin Kayhan, Hakan Kaya, Zahide Mine Yazici, Hasan Goksoy, Sultan Biskin, Omer Erdur

*Bakırköy Education and Training Hospital, Clinic of Otorhinolaryngology, Head and Neck Surgery, Istanbul, Turkey (FJK, HK, ZMY, SB, OE)
Istanbul University School of Medicine, Department of Internal Medicine, Division of Hematology, Istanbul, Turkey, (HG)*

Sudden sensorineural hearing loss as the initial sign of a hematological disease is very rare condition. A 34-year-old male presented to our clinic with bilateral sudden sensorineural hearing loss. The patient was found to have chronic myelogenous leukaemia during a work-up for his hearing loss. This paper reports an unusual case in which presented bilateral sudden sensorineural hearing loss as the first manifestation of the chronic lymphocytic leukemia. Pure tone audiometry revealed profound sensorineural hearing loss of the bilateral ear at all frequencies. The patient was diagnosed as having idiopathic sudden sensorineural hearing loss and admitted for treatment, but his laboratory data indicated that he had an undiagnosed kind of leukemia. The mechanism producing bilateral sudden sensorineural hearing loss in leukemic patient was reviewed and discussed, and the importance of differentiating possible underlying diseases before we diagnose idiopathic sudden sensorineural hearing loss was highlighted.

Submitted : 13 May 2009

Accepted : 02 August 2009

Introduction

Chronic myeloid leukemia (CML) is a myeloproliferative disorder characterized by infiltration of the blood, bone marrow and other tissues by neoplastic cells of the hemopoietic system particularly of granulocytic lineage^[1]. The clinical onset of disease is generally insidious and less than % 50 percent of patients are symptomatic at the time of diagnosis. Less than 50% of the patients are symptomatic^[1]. The disease is characterized by anemia, extreme blood granulocytosis, often thrombocytosis on splenomegaly. Less common features are related to granulocyte or platelet dysfunction such as infections, thrombosis or bleeding^[2]. Occasionally, patients present with leukostatic manifestations such as mental status changes, headache, visual disturbance, stroke, cerebellar signs and priapism^[3].

Common symptoms are bone pain, weight loss, excess sweating, fatigue, and early satiety and abdominal discomfort related to splenomegaly. Uncommon presenting symptoms include those related to leukostasis^[3].

Deafness in association with vestibular symptoms are rarely occurs in CML as the first sign. Otologic symptoms can originate from bleeding, infiltration of tumor, infection or hyperleukocytosis^[4].

We report our experience of a 34-year-old man with bilateral sudden sensorineural hearing loss (SSHL) as the first sign of chronic myeloid leukemia. Clinical and radiological aspects for CML with deafness and vertigo were discussed reviewing the literature.

Case Report

A 34-year-old man presented to our clinic with bilateral sudden hearing loss, vertigo and nausea for a duration of 3 days. Hearing loss was described as sudden onset and also reported to impair social activity. It was not associated with otalgia and otorrhoea. There was no history or sign of physical trauma, drug intake or infectious diseases. Patient had no prior systemic infection or operation. Hearing loss had been started at right ear first and progressively became bilateral within 24 hour. Both external auditory canals and tympanic membranes were normal as well as their ENT examinations. Nystagmus was not observed, but blurred vision was added to his hearing complaints. Systemic physical examination revealed moderately enlarged spleen and mild hepatomegaly. There was no significant finding in neurologic examination other than findings associated with eighth cranial nerve. The audiometry showed bilateral profound sensorineural type hearing loss on both ears with pure tone average 87 dB on left ear and 93 dB on right ear (Figure 1).

Corresponding address:

Mine Yazici,
Bakırköy Eğitim ve Araştırma Hastanesi Kulak Burun Boğaz ABD Tevfik Sağlam cad., No: 13, 34147 Zuhuratbaba, Istanbul, Turkey
Telefax: + 90 212 5424491; Telephone: + 90 212 4147250; E-mail: minealmaz@yahoo.com

Copyright 2005 © The Mediterranean Society of Otolaryngology and Audiology

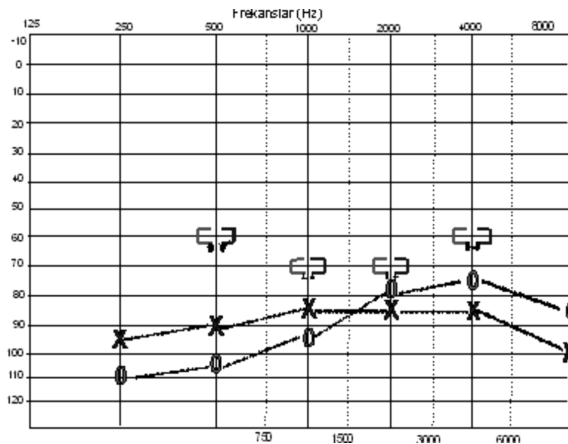


Figure 1. Audiometry showing sensorineural type of hearing loss on the bilateral ear

Laboratory investigations revealed profound leukocytosis of $390.800/\text{mm}^3$ with mild anemia (hemoglobin was 11.5 g/dl) and thrombocytosis (platelet count $683 \times 103/\text{microL}$). Coagulation tests were normal. Patient was referred to hematology department for investigation of leukocytosis. Abundance of granulocytic cells in all maturity levels was seen in microscopic examination of peripheral blood smear. Bone marrow aspiration and biopsy were done for pathological and cytogenetic examinations with suspicion of myeloproliferative disorder. Bone marrow biopsy was reported to be hypercellular bone



Figure 2. Cerebellopontine angle on axial T1 weighted MRI demonstrated no lesion

marrow with significant increase at granulocytic cell lineage with blasts less than % 5 suggesting CML in chronic phase. Cytogenetic analysis revealed hundred percent Ph chromosome positivity and definite diagnosis as CML. Chromosomal analysis at hematology department showed the translocation $t(9;22)$ designed as Philadelphia chromosome was positive.

With a diagnosis of CML concomitant with bilateral SSHL, the patient underwent further radiologic examination. Magnetic resonance imaging (MRI) of temporal bone with intravenous gadolinium was normal (Figure 2). Cranial MRI showed loss of density on third and lateral ventricle which was evaluated as benign intracranial hypertension (Figure 3) Ophthalmologic examination showed optic disc edema and central retinal vein occlusion. Orbita MR was found normal.

Our standard treatment of SSHL included intramuscular injection of 250 mg prednisolone only one time followed by tapering dosing schedule of oral methyl-prednisolone 1 mg/kg/day for a month. There was no change, clinically and audiological with the treatment of systemic steroid after 6 months of follow-up. Hydroxyurea therapy started by hematology department for reduction of leukocyte count and patients complaints associated with leukostasis disappeared gradually. After hydroxiurea therapy,

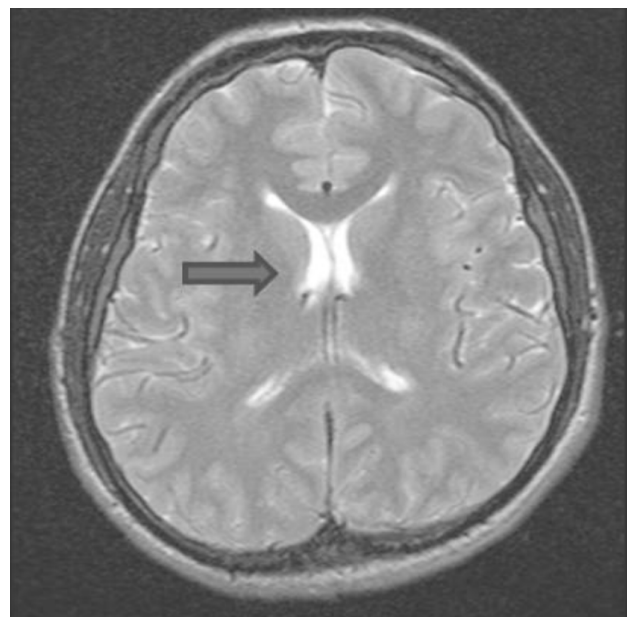


Figure 3. Cranial MRI showed loss of density on third and lateral ventricle

imatinib (0.4-0.8 g/day) was added to chemotherapy regimen for 29 days. Currently the patient is still continuing to take imatinib (0.4 g/day) under follow-up.

Discussion

Sudden sensorineural hearing loss is an idiopathic condition of acute hearing impairment with an incidence of 5 to 20 per 100,000 persons in a year^[5]. The definition of SSHL is a hearing loss greater than 30 dB at more than three contiguous frequencies developing within less than 3 days. Various etiologies of sudden hearing loss should be considered and evaluated.

Chronic myeloid leukemia is a clonal disease, arising from a transformed hemopoietic pluripotent stem cell and resulting abundance of all cell lineages, especially granulocytic one. The disease is characterized by anemia, extreme blood granulocytosis, often thrombocytosis and splenomegaly. The clinical onset of disease is generally insidious and only less than 50 percent of patients are symptomatic at the time of diagnosis.

Common neurological signs and symptoms in CML were documented to be ataxia, nystagmus, papilledema, blindness, retinal alteration, tinnitus, vestibular syndrome, facial palsy, headache, dysarthria, coma and deafness^[6]. Deafness in leukemia was first described by Donne in 1844 as cited by Hsu et al.^[7]. It is described as being sensorineural, unilateral, bilateral or starting as unilateral and progressing to become bilateral^[1].

Pathogenesis of hearing complaint symptom in leukemia is very complex, may include multiple mechanisms such as hyperleukocytosis, causing leukostasis and abnormal microvascular perfusion, leukemic infiltration and infection^[1]. In the 1970's, Paparella et al. investigated the largest series of temporal bones from patients with various forms of leukemia^[4]. Autopsy cases from patients with acute myelogenous leukemia, chronic myelogenous leukemia, acute lymphocytic leukemia and chronic lymphocytic leukemia demonstrated hemorrhage, leukemic infiltration, infection and reduced hair cells^[4]. He observed that auditory complications develop earlier and more frequently in the acute than the chronic leukemias^[4].

Our patient presented with SSHL and visual alterations as the clinical manifestations of hyperleukocytosis of CML like some published cases^[8-10]. Most symptoms associated with leukostasis are reversible with a rapid reduction of the leukocyte count. Deafness is likely to be due leukostasis with occlusion of the labyrinthine artery

resulting in irreversible deafness despite a rapid reduction of leukocyte count^[8]. Some cases of sudden hearing loss in CML may be improved by sitoreduction and chemotherapy. But unfortunately we did not observe improvement in our patient. The absence of improvement in hearing most probably occurred as a result of hyperleukocytosis with leukostasis and occlusion of labyrinthine and other small arteries of the vertebro-basilar area. This case illustrates the importance of follow-up.

References

1. Naithani R, Chandra J, Mathur NN et al. Hearing loss in chronic myeloid leukemia. *Pediatr Blood Cancer* 2005; 45:54-56.
2. Wetzler M, Byrd JC, Bloomfield CD. Acute and chronic myeloid leukemia. In: Fauci AS, Braunwald E, Kasper DL, Hauser SL, Longo DL, Jameson JL, Loscalzo J, eds. 17th edition *Harrison's principles of internal medicine*. New York: McGraw Hill Publishers, 2008: 683-685.
3. Kelly KE, Lange B. Oncologic emergencies. *Pediatr Clin N Am* 1997; 44: 809-830.
4. Paparella MM, Berlinger NT, Oda M et al. Otolological manifestations of leukemia. *Laryngoscope* 1973; 83:1510-1526.
5. Byl FM Jr. Sudden hearing loss: eight years' experience and suggested prognostic table. *Laryngoscope* 1984; 94: 647-61.
6. Chae SW, Cho JH, Lee HJ et al. Sudden hearing loss in chronic myelogenous leukaemia implicating the hyperviscosity syndrome. *J. Laryngol Otol* 2002; 116:291-293.
7. Hsu YC, Su CY, Hsu RF. Unilateral sudden hearing loss as a presenting manifestation of chronic myeloid leukemia: case report. *Otolaryngol Head Neck Surg*. 2004;130:271-3.
8. Resende LS, Coradazzi AL, Rocha-Junior C et al. Sudden bilateral deafness from hyperleukocytosis in chronic myeloid leukemia. *Acta Haematol* 2000; 104:6-9.
9. Acar GO, Acioğlu E, Enver O, Ar C, Sahin S. Unilateral sudden hearing loss as the first sign of chronic myeloid leukemia. *Eur Arch Otorhinolaryngol* 2007; 264:1513-6.
10. Genden EM, Bahadori RS. Bilateral sensorineural hearing loss as a first symptom of chronic myelogenous leukemia. *Otolaryngol Head Neck Surg* 1995;113:499-501.