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CASE REPORT

# Unilateral sudden hearing loss as the first sign of chronic myeloid leukemia

**Gül Özbilen Acar · Engin Acıoğlu · Özgün Enver · Cem Ar · Serap Şahin**

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**Abstract** Chronic myeloid leukemia (CML) is one of the etiologic causes of sudden hearing loss and vertigo. However, deafness in association with vestibular symptoms rarely occurs in CML as the first sign. In this article, a 50-year-old male with CML whose first signs and symptoms were unilateral sudden hearing loss and tinnitus in the right ear, vertigo and nausea was presented. Aetiopathogenetic mechanisms, clinical and radiological aspects and therapeutic options for CML with deafness and vertigo were discussed reviewing the literature.

**Keywords** Chronic myeloid leukemia · Sudden hearing loss · Deafness · Vertigo · Intratympanic steroid therapy

## Introduction

Chronic myeloid leukemia is described as a myeloproliferative disease. The blood, bone marrow and other tissues and organs are invaded by neoplastic cells of the hematopoetic system, mainly of granulocytic descent [11]. Less than 50% of the patients are symptomatic in the chronic period of CML. Common findings in this disorder are weakness (due to anemia), dysphagia, abdominal distension and pain (related to hepatosplenomegaly), as well as fever, weight loss and night sweat (originating from hypermetabolism) [8].

Acute or chronic myeloid leukemia rarely affects ear [1]. Otological symptoms can originate from bleeding, infiltration of the tumor, infection, or hyperleukocytosis [6, 9, 10].

We report the clinical, laboratory and radiological findings in a 50-year-old male with CML who presented with deafness, tinnitus, vertigo and nausea and we focus on possible aetiopathogenetic factors and therapeutic approach for this disorder.

## Case report

A 50-year-old man present to a public hospital with unilateral (right) sudden hearing loss and tinnitus, vertigo, and nausea for a duration of 2 days in October 2005. Complaints of the patient had not improved by medical treatment. Because laboratory results indicated leukocytosis, the patient was transfered to our hospital after 6 days. At this time, blurred vision was added to his other complaints. There was no otalgia or otorrhoea. No previous history of otologic trauma, drug intake, noise exposure or upper airway infection was noted.

The clinical examination showed that the patient had massive hepatosplenomegaly, and bilateral multiple cervical, submandibular, axillar and inguinal lymphadenopathies. Signs of stasis (crepitations) were found on the inferior part of the right chest and on the median and

G. Ö. Acar · E. Acıoğlu · Ö. Enver · C. Ar · S. Şahin
Cerrahpaşa Medical School,
Department of Otorhinolaryngology,
Istanbul University, Istanbul, Turkey

G. Ö. Acar · E. Acıoğlu · Ö. Enver · C. Ar · S. Şahin
Cerrahpaşa Medical School, Division of Hematology,
Department of Internal Medicine,
Istanbul University, Istanbul, Turkey

G. Ö. Acar (✉)
Ata :3 / 4, D: 249, Sedef Caddesi Ataşehir,
Kadıköy, Istanbul, Turkey
e-mail: gulozbilenacar@gmail.com

inferior part of the left chest. Cardiophrenic sinus was closed on the left chest. Cardiovascular system was normal. Mental functions, motor system, sensory system and the examinations of all cranial nerves except the eighth were normal. Nystagmus was not detected. Hemorrhage and exudative lesions were found in the fundus examination.

Both external auditory canal and tympanic membranes were assessed normal. The response of tuning fork test (weber) was lateralized to the left side. The audiometric examination showed total sensorineural type of hearing loss on the right ear with a pure tone average of 110 dB. Audiogram was normal on the left ear (7 dB) (Fig. 1). BAEP, using Chiappa's methodology, revealed unilateral (right) abnormal I–V interpeak latencies.

Outcomes of laboratory showed total leukocyte count 567,000 $mm^3$, haemoglobin 12 g/dl, haematocrit 37%, and platelet count 270,000 $mm^3$. There was myelodysplastic aspect and 3% atypical mononuclear cell in the peripheral blood. Coagulation tests revealed activated partial thromboplastine time (APTT) 60%, and protrombin time (PT) 15.8 s. Lactate dehydrogenase (LDH) had risen to 1,364 (U/L) indicating myeloid proliferation. Bone marrow aspiration findings suggested the chronic phase of CML with myeloid hyperplasia, increased promyelocytes, and 3% blast cell. In addition, chromosomal analysis showed that the translocation *t* (9.22) was positive.

Cranial computed tomography (CT) and cranial magnetic resonance imaging (MRI) were found normal. However, thorax CT showed some findings related to bilateral leukemic infiltration of the chest. Temporal bone MRI with intravenous gadolinium was evaluated normal (Figs. 2, 3).

Therefore, leukapheresis was urgently applied on the patient for 3 days. Additionally, alkalization of the urine with sodium-bicarbonate 8.4% (3 × 1 flacon/day) and 0.9% sodium chloride (3 × 1,000 ml/day) was included in the therapy regimen for first 5 days. Chemotherapy was administered with alloprunol (0.6 g/day) and hydroxiurea (1–4 g/day) for first 10 days. After 1 month, imatinib (0.4–0.8 g/day) was added to chemotherapy regiment for 29 days. Currently, the patient is still continuing to take imatinib (0.4 g/day) under follow-up.

An application of intratympanic steroid [dexamethasone 8 mg (1–2cc)] was administered into the right ear under the local anaesthesia five times in 1 month for treatment of deafness. There was no change, clinically and audiologically, in his hearing on the right side with the treatment of intratympanic steroid after 1 month, 6 months and 1 year of follow-up. Although intensity of tinnitus decreased, it has been still continued. Vestibular symptoms disappeared during follow-up in 1 week.

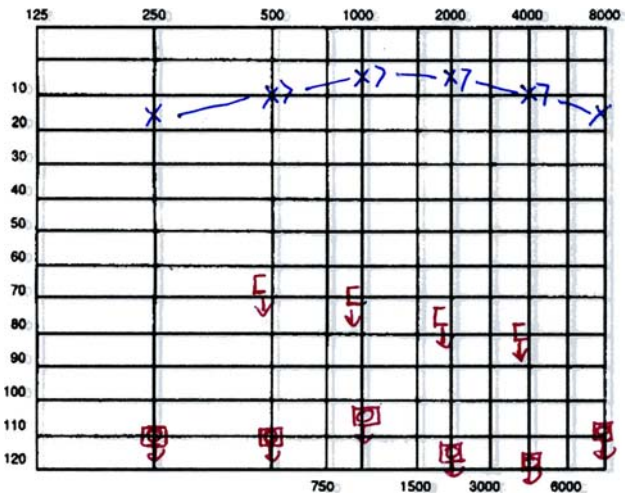


**Fig. 1** Audiometry showing total sensorineural type of hearing loss on the right ear and normal hearing on the left ear

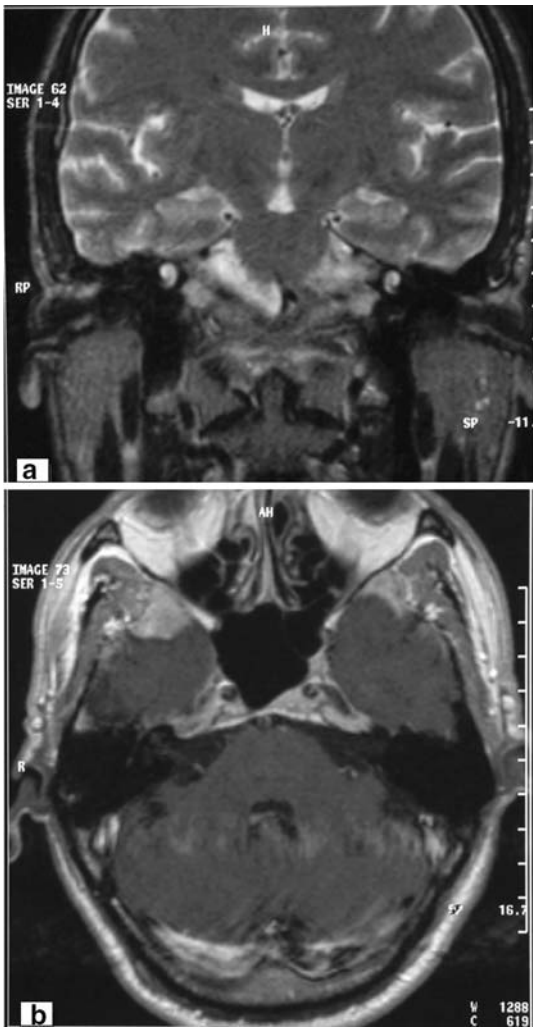


**Fig. 2** Coronal, T2 weighted MRI scan with gadolinium contrast and axial, T1 weighted MRI scan without contrast shows no cochlear pathology

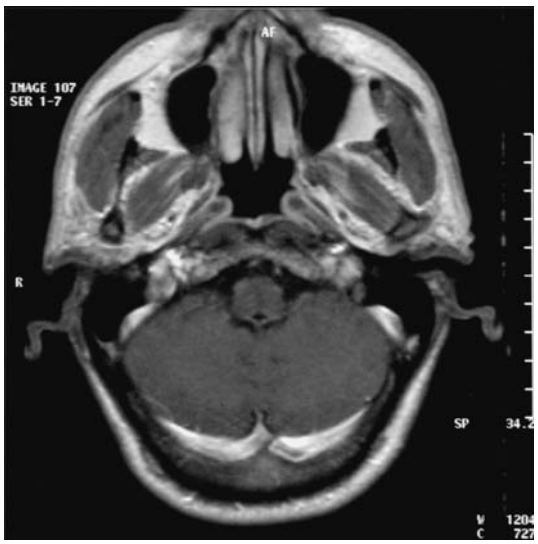


**Fig. 3** Any lesion is not demontrated in the cerebellopontine angle on axial, T1 weighted MRI scan with gadolinium contrast

Upon 2 months of therapy, the hepatosplenomegaly disappeared. Total leukocyte count decreased to 5,400 $mm^3$. Bone marrow control biopsy revealed findings of remission. The total leukocyte count was found within the normal range at discharge after 2 months, as well as after 1 year of follow-up.

## Discussion

Till date, only 28 cases of CML with deafness have been reported. In these papers, the total numbers of male/female were 13/15. Common neurological signs and symptoms in this disease were documented to be ataxia, nystagmus, papilledema, blindness, retinal alterations, tinnitus, vestibular sendrome, facial palsy, headache, dysarthria, coma and deafness [7, 11].

Otological findings, for instance, sudden hearing loss, vertigo, tinnitus, facial weakness, and infection, were found in 16–40% of leukemic patients. However, deafness as an initial sign of this disease is very rare [3]. Sudden sensorineural hearing loss and tinnitus in the right ear, vertigo, and nausea were the first findings of CML in our patient.

CML associated deafness presents itself as sensorineural, unilateral, bilateral or starting unilaterally and developing into bilateral [8]. The pathogenesis of this clinical symptom in leukemia is very complex, and may include multiple mechanisms such as leukemic infiltration, hyperviscosity syndrom, inner ear hemorrhage, and infection [2, 5, 9, 10, 12]. A relationship between leukemic infiltration of the cochlea and impairment of hearing in CML was determined [9]. This neoplasm was documented to have predilection for the labyrinth by some authors [14]. In some literature, the inner ear hemorrhage has been shown as a cause of sudden hearing loss in leukemia [13]. Hyperleukocytosis is considered to be the foremost pathogenesis of CML with hearing loss, as suggested by recovery of hearing upon leukapheresis in some cases [4]. Small aggregates and/or leukocyte thrombi develop from hyperleukocytosis syndrome, creating the possibility of tissue infarction. Irreversible deafness in spite of a rapid decreased leukocyte probably arises from leukostasis with occlusion of the labyrinthine artery which supplies the cochlea and vestibule [5].

Some cases of sudden hearing loss in CML may be improved by leukapheresis and chemotherapy. This has been achieved in some patients with hyperviscosity syndrome, suggesting that this type of deafness is reversible [8]. Besides, in one case with leukemic infiltration of cochlea, hearing was improved significantly after administration of intrathecal methotrexate [7].

In conclusion, we performed intratympanic steroid treatment for CML with deafness as a first time. Unfortunately, we did not observe any improvement in hearing. However, application of local steroid therapy did not cause any adverse influence on the overall progress of CML, either.

The absence of improvement in hearing led us to assume that in our patient, deafness was due to hyperviscosity with occlusion of the labyrinthine and other small arteries of the vertebro-basiler area.

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