CASE REPORT

BILATERAL SUDDEN PROFOUND SENSORINEURAL HEARING LOSS IN A PATIENT WITH CHRONIC MYELOID LEUKEMIA: CASE REPORT

Nchifor K, Irungu C

Department of Surgery, ENT Head and Neck Surgery, School of Medicine University of Nairobi, Kenya

Address for correspondence: Dr Keafon Nchifor. Email: keafonnchifor.kn@gmail.com

ABSTRACT

Sudden hearing loss is a rare complication of Chronic Myeloid Leukaemia (CML). In our report, we describe the case of a 41-year-old male who presented as an emergency, was diagnosed with CML, Diabetes mellitus and hypertension and admitted for management. His initial white blood cell count was 812000cells/mm^3 and he was severely anaemic. The patient developed sudden bilateral hearing loss in the course of admission with a pure tone audiometric assessment revealing bilateral profound sensorineural hearing loss. The patient was started on imatinib with a plan for 2 weekly audiology reviews, but was lost to follow up. This report suggests the hyperviscosity syndrome as a cause of focal neurological manifestations, as well as a synergistic effect of cardiovascular risk factors in hearing loss.

Key words: Chronic myeloid leukemia, Sensorineural hearing loss, Hyperviscosity syndrome

INTRODUCTION

Sudden sensorineural hearing loss in chronic myeloid leukemia is very rare, with a few case reports recorded in literature. The hyperviscosity syndrome demonstrated in myeloproliferative disorders has been implicated as a cause of sensorineural hearing loss. Hyperleukocytosis is responsible for an increase in blood viscosity, and is defined as white cell counts greater than 100000cells/mm^3 [1]. The result is an increase in resistance to blood flow, vascular stasis and therefore hypoperfusion of several organs. Patients tend to present with visual disturbances, bleeding disorders and focal neurological signs[1]. In our case report, we discuss a patient with CML who developed sudden sensorineural hearing loss in the course of admission.

CASE PRESENTATION

Case of a 41-year-old male who presented to our health facility with complaints of left upper quadrant abdominal pains and dizziness of one-month duration. Patient was admitted and was found to be severely anaemic (Hb 7.4g/dl) during the course of hospitalization. His full blood count revealed a white blood cell count of 812000cells/mm^3, with a differential count showing 90% neutrophils, 3.5% lymphocytes, 4.1% monocytes, 1.8% eosinophils and 0.6% basophils. Genetic studies revealed the Philadelphia chromosome and BCR-ABL gene, and a diagnosis of Chronic Myeloid Leukemia (CML) was made. The patient was also newly diagnosed of type 2 Diabetes mellitus (glycated haemoglobin at 13%) and hypertension during this admission. Following diagnosis of CML, treatment was immediately started with hydroxyurea 500mg thrice daily, and transfusion of 4 units of blood. He also received mixtard 16 IU morning and 8IU evening, metformin 500mg twice daily, lorsatan 100mg once daily. Allopurinol 300mg twice daily was added to his treatment two days later.

Eight days following admission, the patient complained of sudden bilateral hearing loss. His hearing had been normal on admission with no previous history of ear disease. A physical examination showed normal external auditory canals bilaterally, with both tympanic membranes intact. Cranial nerve examination was normal. There was no response to tuning fork tests. A pure tone audiometric assessment revealed bilateral profound sensorineural hearing loss with no response at 120decibels across all frequencies (Figure 1). At the time of ENT examination on day 8 of admission, the WBC had dropped to 700000cells/mm^3. A plan to carry out CT scans of the brain and temporal bones was made, but due to the patient's financial constraints, these could not be done.

Still in the course of his admission, the patient developed priapism, but declined corpora carvenosa shunting. He was transferred to another facility to be initiated on imatinib, and was given a review at the ENT clinic for two-weekly audiometric assessments. The patient however did not show up for the reviews. Phone calls were made regularly to the patient's spouse and the report was that the patient was still deaf a year after the onset of his illness.

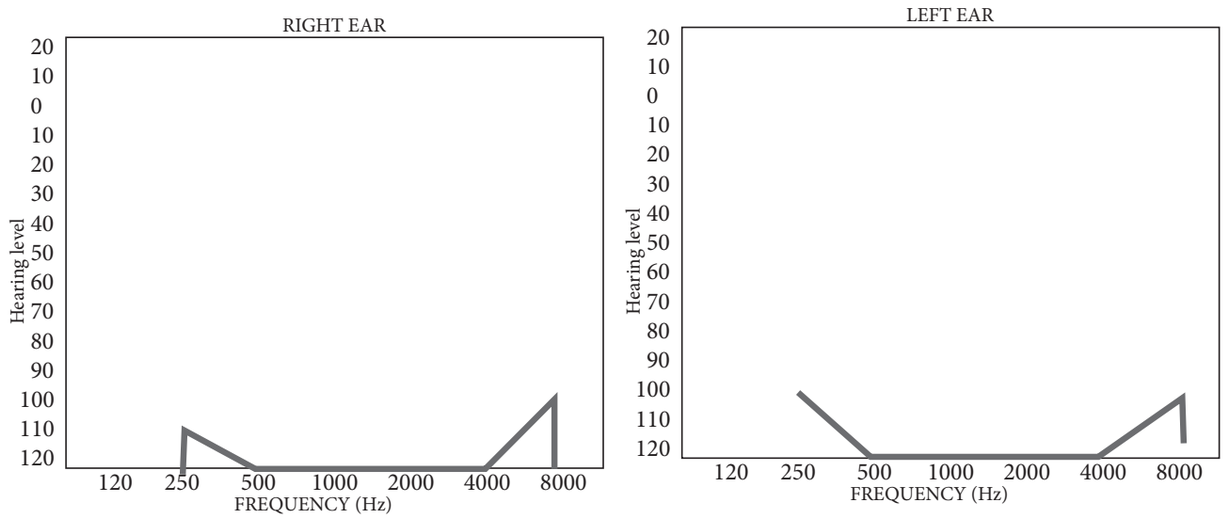


Figure 1: Pure tone audiometric assessment of the patient

DISCUSSION

Bilateral sudden profound sensorineural hearing loss in CML is rare, with several case reports postulating different aetiologies. This case was reported at the Kenyatta National Hospital in Kenya, and is the first of its kind to be reported. Critical levels of WBC are said to be greater than 300000cells/mm^3 and can be associated with hyperviscosity syndrome. In our report, our patient had initial WBC of 812000cells/mm^3, more than twice the critical levels. A case was described by Amancio *et al*[1] associated hyperviscosity syndrome and acute hearing loss in a patient initially presenting with hearing loss and eventually diagnosed with CML on admission. The patient had leukocyte counts of 645000cells/mm^3 and anaemia of 7g/dl. Hydration therapy, hydroxyurea, allopurinol, dexamethasone and leukapheresis were some of the treatment options applied. Hearing had not improved by the time he was discharged home, about 1 month following admission.

Resende *et al*[2] reported a case similar to ours with significant hyperleukocytosis and bilateral hearing loss as well as other neurological symptoms. Their patient had bilateral hypoacusis confirmed by Brainstem Auditory Evoked Potentials (BAEP), and a brain MRI showing areas of subcortical ischaemic infarction. Their patient did well on 5 sessions of leukapheresis and hydroxyurea 4g/day, with significant resolution of all clinical and laboratory parameters. Our institution does not offer leukapheresis as a treatment option. Considering his financial status, our patient was unable to get any imaging tests or more detailed audiological assessments. Being lost to follow up, we were unable to describe the resolution of his symptoms on imitinib as this treatment was carried out in another health facility.

Leukapheresis is a procedure in which blood of the patient is passed through a leukapheresis machine, which separates and collects the WBCs cells, while returning the rest of the patient's blood with or without addition of replacement fluid such as colloid, human albumin, or a crystalloid solution. This procedure has been shown to reduce the WBC count by 10-70% in just one session[1]. Prospective studies on the efficacy of leukapheresis on CML are lacking.

Several other case reports cite hearing loss as the initial manifestation of CML[4,5]. Chih-Cheng Hsiao *et al*[4] described the case of a 12 year old who presented with a 2 day history of sudden right sided hearing loss, with the left side later involved in the course of her treatment. CML was diagnosed in the course of her medical assessment. In her case, despite several interventions including hydration therapy, allopurinol, and hydroxyurea (30 mg/kg/day, per os), interferon-alpha, leukapheresis and cytarabine, she deteriorated and died on day 9 of admission.

Our case report makes mention of comorbidities such as Diabetes mellitus and hypertension which were diagnosed in our patient on admission. Type 2 diabetes has still not been proven to be a sure cause of sudden sensorineural deafness. However, several studies try to bring out a relationship between these comorbidities and hearing loss[6,7]. Aimoni *et al*[8] in a case controlled study revealed even greater risk of idiopathic sudden sensorineural hearing loss in patients with more than one cardiovascular risk factor. The patient in this report had high glycated haemoglobin levels of 13% which have also been shown to increase the risk of hearing loss[6,7].

CONCLUSION

There is a possibility that the patient presented in our case report developed bilateral sudden profound sensorineural hearing loss due to hyperleukocytosis and resultant hyperviscosity of blood. This may have

been worsened by the presence of cardiovascular diseases at the time of presentation. Diagnosis and intervention should be timely in order to improve on disease outcomes and prevent disease progression and complications.

Conflict of interest: The authors declare that there is no conflict of interest regarding the publication of this case report.

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