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Case Report

Sudden Sensorineural Hearing Loss in patient with Chronic Myelogenous Leukemia – A case study

Abstract

Rapid progressive sensorineural hearing damage as the early sign of a hematological illness is often found in patients with Chronic Myeloid Leukemia. A 100-year-old man presented to our outpatient department with 2 hours history of bilateral sudden sensorineural hearing loss (SSNHL). After the hematological, bone marrow and radiological workup, the patient was found to have chronic myelogenous leukemia.

The patient was identified as having unexplained SSNHL and was admitted for treatment, but his hematological investigations indicated that he had an undiagnosed form of leukemia. The possible pathogenesis of developing SSNHL from CML is discussed here within the report with highlights of differential diagnosis of SSNHL.

Introduction

Chronic Myeloid Leukemia (CML) is a myeloproliferative disorder which results in excessive production of granulocytic elements at every period of differentiation [1]. This results in infiltration of blood, bone marrow and other tissue by neoplastic cells of granulocyte lineage [2]. Subjects are often diagnosed parenthetically when raised White Blood Cell count (WBC) is found on routine Complete Blood Count (CBC) or when splenomegaly is discovered during an abdominal exam or ultrasonography. Non-specific symptoms of giddiness, fatigability, and weight loss over a period of time may be encountered after the beginning of the ailment.

Ear manifestations have been discovered in 15–40% of leukemic patients [3–6]. These include moderate to profound sensorineural hearing loss (SNHL), tinnitus, giddiness, and facial nerve paresis. Less common features are related to granulocyte or platelet dysfunction such as infections, thrombosis or bleeding [7]. Rarely, patients present with leukostatic presentations such as irritability, headache, visual disturbance, ischemic stroke, cerebellar signs and priapism [8].

Hearing loss in conjunction with vestibular signs symptoms is rarely seen in CML as the first manifestation. These symptoms can originate from hemorrhage, infiltration by tumor cells, infection or leukostasis reactions [4].

Case Report

A 100yr old urology patient who was being managed as the

case of Benign prostatic hypertrophy (BPH), present in ENT clinic with 3 month history of generalized body weakness, dizziness and blurring of vision, with 2 hours history of bilateral Sudden hearing loss and tinnitus. No ear pain, exposure to loud noise, trauma to the ear, history of foreign body to the ear, facial pain and no history suggestive of nasal nor throat symptom.

No bleeding from any of the craniofacial orifices and no prolonged bleeding from the punctured site. No contact with ionizing irradiations, and history suggestive of the respiratory symptom.

Not a known hypertensive, diabetic, peptic ulcer disease nor asthmatic patient. Nil history of surgery nor blood transfusion in the past.

The patient had been on Tablet Tamsulosin for 1 month prior to the onset of the hearing loss.

Examination revealed an elderly man who was conscious and alert, pale, anicteric, acyanosed and not dehydrated. Vital signs were within normal limits. The Liver was about 4–5cm down below the costal margin, while the Spleen was about 4–5cm down below the costal margin. Direct digital examination revealed mild prostate enlargement.

Otoscope examination was essentially normal. However Pure Tone Audiometry (PTA) revealed bilateral mild-moderate sensorineural hearing loss (Figure 1).

Full blood count results were as follows: PCV=24%, WBC= $294.4 \times 10^9/L$, Blast cell=13%, Promyelocyte=9%, Myelocyte= 10%, Metamyelocyte=3%, Neutrophil=51%, Lymphocyte: 8%, Monocyte=2%, Basophil: 4%.

Peripheral blood film showed a Chronic Myeloid Leukemic (CML) patient in accelerated phase (Figure 2). Marked leucocytosis showing sheets of myeloid cell series.

The myeloblast and promyelocyte make up to 18% of the total white blood cell. There was thrombocytosis.

Bone marrow aspiration indicated a Chronic Myeloid Leukemia (CML) patient in accelerated phase (Figure 3). There was Hypercellular marrow with Myeloid: Erythroid ratio 25:2.

Erythropoiesis was severely depressed. Myelopoiesis was markedly increased and showed sequential maturation along the myeloid series. There was increased Basophil and Eosinophil precursors, constituting about 20% of the nucleated marrow element. Lymphopoiesis was increased and Megakaryocytes were actively budding off platelets.

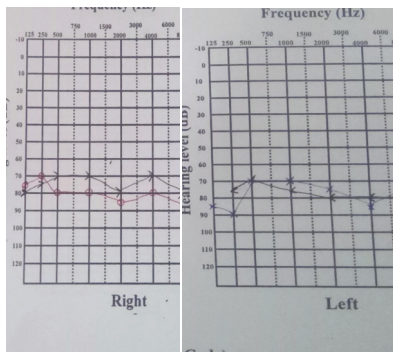


Figure 1: Pure Tone Audiometry showing a bilateral mild-moderate hearing loss.

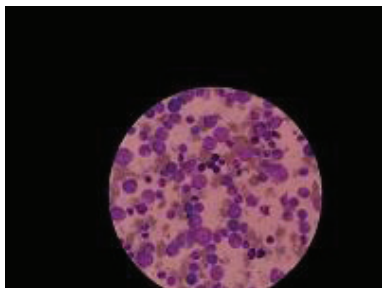


Figure 2: Peripheral blood film of a Chronic Myeloid Leukemic (CML) patient in accelerated phase. Marked leucocytosis with sheets of myeloid cell series.

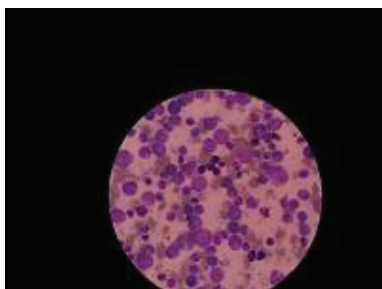


Figure 3: Bone marrow aspiration showing hypercellular marrow with Myeloid: Erythroid ratio 25:2. Erythropoiesis is severely depressed.

Magnetic Resonance imaging (MRI) of his brain showed hypodense third and lateral ventricle. He was commenced on routine therapy for CML: Allopurinol, cyclophosphamide, and hydroxyurea. Also added to the medication were neuron stimulants: Nicotinic acid and neurobion. The patient did well after 3 months and the hearing remarkably improved.

Discussion

Hearing loss as a primary finding of CML is rare. Very few cases have been reported in works [9-12]. The pathogenesis of this entity appears to be multifactorial and includes various mechanisms such as leukostasis, leukemic infiltration of the cochlea, hyperviscosity syndrome, thrombohemorrhagic complications, and infections.

Sudden sensorineural hearing loss (SSNHL) is an idiopathic condition of acute hearing injury with an incidence of 5 to 20 per 100,000 persons in a year [13]. SSNHL is defined as hearing damage more than 30 dB at more than three consecutive frequencies developing within less than 3 days. Various reasons of SSNHL should be taken into account and assessed.

CML is a clonal disease, raised from an altered hematopoietic pluripotent stem cell causing an abundance of all cell lineages, particularly granulocytic one. This ailment is characterized by anemia, extremely elevated blood granulocytes, along with thrombocytosis and splenomegaly. The clinical commencement of illness is generally gradual and only <50% of subjects are symptomatic at the time of diagnosis.

Mechanism of deafness complaint symptom in leukemia is very complex, may comprise several mechanisms such as hyper-leucocytosis, affecting leukostasis and abnormal microvascular perfusion, leukemic infiltration and infection [2]. In the 1970's, Paparella et al. inspected the major series of temporal bones from subjects with different types 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].

This subject presented with SSNHL and visual blurring as the clinical symptom of a leukostatic reaction of CML like some published studies [14-16]. Most problems in conjunction with leukostatic reaction are recoverable with a rapid drop in the leukocyte count by plasmapheresis. Hearing loss is likely due to the leukostatic reaction resulting in occlusion of the labyrinthine artery resulting in irreversible SSNHL despite a rapid drop of leukocyte count [14]. Some cases of SSNHL in CML may be improved by cytoreduction and chemotherapy. But regrettably, we did not detect enhancement in our subject. The absence of improving hearing symptom in our patient most probably happened as a result of hyper-leucocytosis with resultant leukostasis reaction and blocking of labyrinthine and other small arteries of the vertebro-basilar region. This case demonstrates the utmost importance of follow-up.

Conclusion

SSNHL as an initial symptom of CML is very rare. Our patient presented with sudden deafness when the white blood cell count had increased to $294.4 \times 10^9/L$, making hyperleucocytosis as a likely cause. Imaging of his brain showed hypodense third and lateral ventricle which was assessed as benign intracranial hypertension. Therefore, it is likely that as a result of leukostasis reaction labyrinthine and other small arteries were blocked resulting in SSNHL.

Consent Form

Written informed Consent was obtained from the patient for the publication of this case report including the images.

Ethical Approval

This study was approved by the Research and Ethical committee of Federal Teaching Hospital Ido-Ekiti, Ekiti State, Nigeria.

Author's Contribution

Shuaib Kayode Aremu: Designed and supervised the study, performed clinical examination and gave final approval for the manuscript to be published. Ibijola AA: Collected demographic data and proof read the manuscript, and Dosunmu Adepeju Oluwatona helped in writing manuscript.

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