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Case Report Open Access

A Case of a Child with Chronic Myeloid Leukemia Presenting with Vision and Hearing Loss

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Abstract

Chronic myeloid leukemia (CML) is a relatively rare malignancy of cells of the hematopoietic system. Loss of vision and hearing is a very rare presenting manifestation of CML especially at a young age. We are reporting a rare case of CML in 16 years old male presenting with loss of hearing and vision in Muhimbili National Hospital in Tanzania. This case shows how the non-communicable disease can cause morbidity in children in a developing world and the importance of early diagnosis and management of CML in prognosis. In addition the case shows how adequate investigations and documentation is important in follow up of the patient clinical conditions and treatment outcomes. The patient was put on Tabs Alluprinol 300mg once daily for one month and Tabs hydroxyl urea (HU) 3g once daily for one month. 2 weeks later the child was reported to have confusion, being overactive and over talkative for 3 days prior to attending the scheduled hematology clinic. Complete Blood Count showed a marked decrease in White blood cells. With the rapid patient's clinical deterioration after initiation of HU, indicates that detailed medical investigations is crucial in disease management and the use of Tyrosine Kinase inhibitors should be encouraged as it has shown to be more tolerated and better survival outcomes in treatment of CML.

Keywords: Chronic myeloid leukemia; Vision loss; Hearing loss; Childhood leukemia; Tanzania

Introduction

Chronic myeloid leukemia (CML) is a relatively rare malignancy of cells of the hematopoietic system. The malignancy is characterized by an increase in myeloid cells, erythropoietin cells and platelets in peripheral blood. The diagnosis is done by analysis of the bone marrow which usually reveals marked myeloid hyperplasia. The basis of CML diagnosis is presence of translocation of chromosome known as Philadelphia (Ph) chromosome detected by PCR. The common symptoms and signs in patients with this disease are fatigue, weight loss, bleeding, splenomegally, abdominal fullness, anemia, thrombocytosis and leucocytosis. Loss of vision and hearing is a very rare presenting manifestation/complication of CML especially at a young age. Hearing and vision loss can be caused by inner ear hemorrhage or leucostasis in the auditory canal and the retina.With regard to treatment, drug therapy with Tyrosine Kinase inhibitors and hydroxyurea (HU) still remains to be a central of interest. HU has been reported to be well tolerated and commonly used in our settings for patients with Sickle cell Disease and CML. HU like any other drug has been reported to have a number of side effects including disorientation, hallucination and seizures[1-6].

The risk factors reported to be associated with CML includes include ionizing radiation and exposure to benzene. In children there is limited information reported regarding risk factors. Smoking and alcohol consumption during pregnancy, parental age, birth weight and infection have been found to be associated with childhood CML. With regard to gender and sex, literature shows that CML affects all age groups and sex. However in most patients the disease presents around the age of 45 to 55 years. In few cases the disease present during childhood accounting about 2-3% of all leukemia cases in the population and presenting with visual and hearing loss is also uncommon [7-9].

We are reporting a rare case of CML in 16 years old male presenting with loss of hearing and vision in Muhimbili National Hospital in Tanzania. This case shows how the non communicable disease can cause morbidity in children in developing world and the importance of early diagnosis and management of CML in prognosis. In addition the case shows how adequate investigations and documentation is important in follow up of the patient clinical conditions and treatment outcomes.

Case

A 16 years old male attended Lindi Regional Hospital in Southern Tanzania with complains of progressive tiredness, general body weakness, loss of weight and lower limb pain and swelling for six month. He also reported to have loss of vision in both eyes and hearing loss in both ears for one month and yellowish coloration of the eyes for three weeks. Laboratory results from the regional hospital showed all levels of maturation of myeloblasts. The patient was then referred to Muhimbili National Hospital (MNH) in Tanzania a week later with the provisional diagnosis of acute lymphocytic leukemia and was not put on any medication. At the time of admission at emergency department the patient reported similar symptoms. More information regarding patient illness was obtained from his father. The patient reported to have no history of paraplegia and no loss of consciousness; no other abnormality was detected from his present illness apart from loss of appetite and on and off headache.

Information obtained from his father shows that he was born through Spontaneous Vaginal Delivery and received all the vaccination as per extended program on immunization (IPI) of Tanzania. Father reported no history of alcohol intake neither smoking, same applies to both parents. He has been admitted 3 times since he was born, with the recent admission being the 3rd. The first admission was due to vomiting and abdominal pain with the diagnosis of Malaria. He was treated and improved; the second was due to the recent illness which he was then referred to MNH. The patient reported no history of blood transfusion nor trauma in the past.

General examination on the day of admission showed a weak young boy, wasted with mild jaundice, moderately pale and slightly distended abdomen. Blood pressure was 120/70, respiratory rate was 20 breaths per minute, body temperature was 36°C, pulse rate of 124 beats/minute and oxygen saturation (SPO_2) of 94. Clinical examination of the systems revealed uniformly distended abdomen, palpable non tender spleen about 18cm below the lower left coastal margin. Liver was not palpable and no ascites was noted. On cardiovascular system examination hyperactive apical area was noted with normal heart sounds. Central nervous system and mental status evaluation yielded no significant abnormality.

On the day of admission the following investigations were ordered; Complete Blood count with peripheral smear analysis, Comprehensive Chemistry Pannel, Chest X-ray, Electrocardiogram, ECHO, Reticulocyte count and bone marrow aspiration. It was also advised that the patient has to be reviewed by ophthalmologist and ear, nose and throat (ENT) specialist.

His blood count showed white cell count 651 K/uL, hemoglobin 7.26 g/dl, neutrophils 195, lymphocytes 11.9, monocytes 5.93, basophiles 0.329, MCV 77.9, MCHC 37.4, MCH 29.1, platelets 246 K/uL and RDW 18.7%. His peripheral blood smear showed normocytic normochromic red blood cells, white blood cells shows leucocytosis and all levels of maturation myeloblast, promyelocyte band and mature neutrophils with normal platelets features suggestive of myeloproliferative disorder most likely CML.

Bone marrow aspiration showed myeloid/erythroid ratio of 11:1, Granulopoiesis increased with left shift, Megakaryotes seen with normal maturation, plasma cells not increased, lymphocytes not increased, monocytes increased by 2%, abnormal cells/blasts not increased (<5%) and there was no abnormal non hematopoietic cells infiltrates noted.

Due to abdominal pain and weight loss, Hepatitis B and C antibody screening was done and results were non reactive. Moreover due to endemicity of HIV in Tanzania, HIV screening was also done and result was negative. Chest radiography showed features of mitral heart disease with impending heart failure. ECG was essentially normal. CCP showed urea 3.8 mmol/l which was within the normal range, high Uric acid level (0.54 mmol/l). Ophthalmology and Ear Nose and Throat review was not done.

The final diagnosis was hearing loss and visual loss secondary to chronic myeloid leukemia. The patient was put on Tabs Alluprinol 300 mg once daily for one month and Tabs hydroxyl urea 3 g once daily for one month. The patient was discharged 4 days later after starting alluprinol and hydroxyurea. He was supposed to continue with treatment and return to the hematology clinic after 2 weeks.

2 weeks later on attending outpatient hematology clinic visit, the father reported his child having confusion, being overactive and over talkative for 3 days prior to attending the scheduled hematology clinic. At hematology a quick mental status evaluation was done and no significant finding was observed apart from disorientation with the score of 6 out of 8; and rapid test for malaria parasite was positive. He was diagnosed with severe malaria; he was admitted and put on I.V quinine 350 mg every 8 hrs for 48 hrs then tabs 600 mg 8 hourly for 7 days. He was also given diazepam 5 gm start. CBC was taken again on this visit and results showed pancytopenia with marked decrease in White blood cells from 651 to 1.77 K/uL in a month time. Platelets were 372 K/uL, RDW of 44.4%, hemoglobin of 6.84g/dl, neutrophils= 0.724, lymphocytes=0.778, monocytes=0.157, basophiles=0.088, MCV=87.3 MCHC= 32.4 and MCH=28.3.

After a clinical discussion with hematologists the decision came to a conclusion that the patient is presenting with HU side effects and HU was stopped and the patient continued with Alluprinol and antimalarials.

Follow up and treatment outcome

After a week of inpatient monitoring, the patient was stable and discharged and advised to attend outpatient hematology clinic after 2 weeks. However the patient did not turn up in the consecutive visits as advised.

Discussion

CML presenting in children and associated with hearing and vision loss is a rare entity. In this 16 years old case, presence of the high white blood cells in complete blood count and normocytic normochromic red blood cells, leucocytosis and all levels of maturation myeloblast, promyelocyte band in peripheral blood smear the diagnosis of myeloproliferative disorder was suggestive and most likely CML [8]. The presence of hearing and vision loss with these hematological findings; CML was then entertained to be the primary cause of these symptoms. However, not being able to conduct Ophthalmology and ENT specialist evaluation on the patient to rule out other causes of vision and hearing loss limits our confirmation of CML being the primary cause of these symptoms.

The use of HU in treatment of CML in Tanzania has been the first choice due to unavailability of other treatment options including bone marrow transplant. With the reported rapid patient's clinical deterioration after initiation of HU, indicates that detailed medical investigations is crucial in disease management and the use of Tyrosine Kinase inhibitors should be encouraged as it has shown to be more tolerated and better survival outcomes in treatment of CML [4,6].

Adequate follow up and documentation of patients is crucial in understanding treatment outcomes of the patient. For this case we were not able to track the patient after the second visit which made it challenging to report on the treatment outcomes after stopping HU and discharge from the hospital.

Conclusion

CML with loss of hearing and vision in children is rare, and its treatment with HU has proven to work on decreasing the production of white blood cells from the hemopoietic stem system, however it is might have not be tolerated in this particular case due to rapid patient's clinical deterioration. Due to the fact that CML is rare, identifying the alternative cost effective and tolerable regimen is crucial and further studies need to be conducted to identify these newer therapies which

may fit our setting. A good patient follow up mechanism and documentation of patient information needs to be put in place in health facilities so that to be able to get treatment outcomes of the medical management.

Conflict of Interest

Authors do not have any conflict of interest to disclose.

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