LARGE ERYTHEMATOUS MASS OF THE AURICLE IN A 17-YEAR OLD MALE: AN UNCOMMON PRESENTATION OF ACUTE MYELOGENOUS LEUKEMIA

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ABSTRACT

OBJECTIVE: To present an enlarging mass in the right auricle of a 17 year-old male as the initial symptom of Acute Myelogenous Leukemia.

STUDY DESIGN: Case Report

SETTING: Tertiary Hospital

PARTICIPANT: One patient

CASE REPORT: A 17-year old male with on and off fever, ecchymosis on the elbow and knees complained of an enlarging erythematous mass of the right auricle. This was his initial presentation to the attending physician. He was referred to the Otolaryngology (ENT) service were incision and drainage of the right auricle was done and revealed the presence of nonclotting blood. The edematous erythemas of the right auricle progressively enlarged to auricular hematomas. When the laboratory examinations showed pancytopenia with mild splenomegaly, an impression of acute leukemia was made. The diagnosis was confirmed by the findings of myeloid hyperplasia and cytoplasmic myeloperoxidase in his bone marrow. The mass later increased in size to 10 x 7cm, fluctuant, non-tender, fixed and friable occupying the pre auricular and post auricular area. The external ear canal was so edematous that inspection of the tympanic membrane could not be substantiated.

CONCLUSION: Enlarging mass of the auricle is not always due to simple trauma, but may be a cause of other detrimental systemic disease entity like Acute Leukemia in our patient. Incision and drainage of seroma or hematoma of the ear should be done with caution and a good history-taking is a must to avoid complications like bleeding and infection.

KEYWORDS: Acute Myelogenous Leukemia, Auricular hematomas, incision and drainage

INTRODUCTION

Leukaemia is a cancer of cells in the bone marrow (the cells which develop into blood cells). There are many types of cancer which arise from different types of cell. What all cancers have in common is that the cancer cells are abnormal and do not respond to normal control mechanisms. Large numbers of cancer cells build up because they multiply out of control, or because they live much longer than normal cells, or both.

Acute myelogenous leukemia (AML) is a fast-growing cancer of the blood and bone marrow. It is also sometimes called acute myeloid leukemia. In AML, the bone marrow makes many cancerous cells called leukemic blasts and this does not develop properly and cannot fight infections. These leukemic blasts grow quickly and crowd out the bone marrow, preventing it from making the normal red blood cells, white blood cells, and platelets that the body needs.[1]

The signs and symptoms depend on how many normal blood cells you have. It also depends on the number of leukemia cells in your body and where they are[1]. People with leukemia are at significantly increased risk for developing infections, anemia, and bleeding. Other symptoms and signs include easy bruising, weight loss, night sweats, and unexplained fevers. The diagnosis of leukemia is supported by findings of the medical history and examination, and examining blood and bone marrow samples under a microscope.

Auricular hematoma or hematoma auris is a collection of blood between the auricular cartilage and perichondrium. It is more common in males (84%) and the mean age is around 30 years old.[2] It is one of the otological emergencies that occurs secondary to trauma.[2] Although occasionally the spontaneous rupture of a blood vessel may be the cause.[2] Acute auricular hematoma is seen after blunt trauma to the side of the head. A network of vessels provides a rich blood supply to the ear, and the ear cartilage receives its nutrients from the overlying perichondrium. The hematoma occurs almost exclusively on the anterior surface of the auricle where the skin is tightly adherent to the underlying perichondrium, so that sharing forces applied to the ear separate the perichondrium from the cartilage. On the posterior surface, intervening areolar tissue allows the skin to glide over the perichondrium.[3] Auricular hematomas are often encountered in a sports medicine practice, most commonly among wrestlers, but also in boxers, football and rugby players, and judo athletes.[4]

Hematoma is a localized collection of blood, usually clotted, in a tissue or organ. It can occur almost anywhere on the body. Minor injuries occur routinely and the body is usually able to repair the damaged vessel wall by activating the blood clotting cascade and forming fibrin patches. Sometimes the repair fails if the damage is extensive and the large defect allows for continued bleeding. Contusions (bruises) and black eyes are familiar forms of hematoma. Less serious types include hematoma auris (in the tissues of the outer ear, better known as cauliflower ear)[5].

When a blood vessel is damaged blood leaks into the surrounding tissue; this blood tends to coagulate or clot. The greater the amount of bleeding that occurs, the larger the amount of clot formation. Occasionally, diseases may occur that decrease the number of platelets in the bloodstream (thrombocytopenia) or their ability to function. The platelets are the cells in the bloodstream that help initiate blood clot and fibrin formation[5].

Acute myeloid leukemia is an aggressive type of haematological malignancy characterized by abnormal proliferation of white blood cells and their precursors. An extramedullary deposit of leukemic cells in different parts of the body is not unusual. Different forms of leukemia may affect the ear usually middle ear and rarely the inner ear as well[7]. Histologically, the middle ear showed leukemic infiltration and/or hemorrhage much more frequently than did the inner ear or external auditory canal. No sound relationship exists between the anatomical location of hemorrhage in the temporal bone and clinical otological symptomatology[6]. Otologic complications occur almost invariably in those patients with the acute forms, particularly acute lymphocytic leukemia. The changes seen in the temporal bone could be due to leukemic infiltration, haemorrhage or infection[7].

Acute myeloid leukemia (AML) is a cancer of the myeloid line of blood cells, characterized by the rapid growth of abnormal white blood cells that accumulate in the bone marrow and interfere with the production of normal blood cells often vague and nonspecific. The symptoms of AML are caused by replacement of normal bone marrow with leukemic cells, which causes a drop in red blood cells, platelets, and normal white blood cells. These symptoms include fatigue, shortness of breath, easy bruising and bleeding, and increased risk of infection[9].

Enlargement of the spleen may occur in AML, but it is typically mild and asymptomatic. Skin findings in leukemias are very diverse and conventionally divided into specific lesions (leukemia cutis) and nonspecific lesions (leukemids) which may be found in up to 80% of all patients with leukemias[10].

This case is presented to show the consequence of no definitive diagnosis prior to evacuation of hematoma of the auricle and its complications.

CASE REPORT

A case of 17 year old male with on and off fever, ecchymosis on the elbow and knees complained of an enlarging erythematous mass of the right auricle.

History dates back 3 months with erythematous mass of the right auricle. This was associated with ecchymosis at right knee and left elbow, hemoptysis, easy fatigability and night sweats. He consulted at the Davao Regional Medical Center where laboratory work ups revealed pancytopenia. The clinical impression was acute leukemia so 5 units of packed red blood cell was transfused and he was discharged.

The erythematous mass of the right auricle gradually increased in size in a month. He returned back to the hospital for generalized body weakness, fever and abdominal pain and loss of weight. Repeat Complete blood count test was done and showed low hemoglobin, hematocrit and platelet count. Two units of platelet and 3 units of packed RBC were transfused.

Because of the progression of symptoms, he was referred to South Cotabato Medical Center where 6 units packed RBC, 2 units platelet concentrate and 3 units fresh whole blood was transfused. A week before referral and admission to VMMC for Bone marrow aspiration, an incision and drainage was done by the ENT service. There was non-clotting blood removed and so he was given antibiotics. Despite the intervention, the auricle further increased in size.

The past medical, personal and social and family history was unremarkable.

At Veterans Memorial Medical Center, physical examination showed a pale, febrile patient with generalized body weakness and episode of hematemesis. Ear examination showed 2 masses at the auricle 2.5 x 2.5cm and 2 x 2cm non-tender with active bleeding at the right periauricular area. (Appendix I). This later increased in size to (Appendix II) 10 x 7cm, fluctuant, non-tender, fixed, friable mass almost occupying the periauricular area, external auditory canal not visualized and inspection of the tympanic membrane could not be substantiated. CT scan of Temporal Bone was requested which revealed normal finding.

He was seen by the Pediatric Hematologist and Oncologist.

Laboratory investigation revealed microcytic normochromic RBCS, decrease hemoglobin, hematocrit, platelets and increase WBCs, Lymphocytes, positive blasts.

Bone Marrow Aspiration revealed Acute Myelogenous Leukemia. After blood transfusion of 13 units of packed red blood cell and 21 units of platelet concentrate, he was started on cycles of Chemotherapy (Doxorubicin, Cytarabine and Etoposide). Transfusion was given. The patient condition improved and was then discharged.

DISCUSSION

Leukemias are a group of heterogeneous neoplastic disorders of white blood cells. Based on their origin, myeloid or lymphoid, they can be divided into 2 types. Leukemias traditionally have been designated as acute or chronic, based on their untreated course. Acute leukemias usually present with hemorrhage, anemia, infection, or infiltration of organs. All types of leukemia show various degrees of infiltration, depending upon the type of leukemia. The liver and the spleen are common sites of infiltration in myelocytic leukemia. Our patient were mild splenomegaly with pancytopenia, and pale, febrile, weight loss with generalized body weakness, ecchymosis at right knee and left elbow and

edematous erythemas on the right auricle.

Extramedullary leukemic tumors or Granulocytic sarcoma (GS) in AML are uncommon, and may become clinically apparent before or concurrent with clinical evidence of marrow involvement.[10,11,12,13].

The most common otolaryngologic manifestations of GS are oral and pharyngeal lesions[14]. Leukemic infiltration of the ear is uncommon, occurring as acute mastoiditis, conductive or sensorineural hearing loss, vertigo, acute hemorrhagic otitis media, retro-auricular mass or facial nerve paralysis[15,16,17,18]. Paparella et al.[19] reviewed the temporal bones of 25 patients with leukemia and found that 20% of these patients, all being children aged between 11 month-old and 16 years of age, (10% of AML, 50% of ALL) experienced otologic complications directly attributable to their leukemia at admission. When these patients were examined histologically, the middle ear showed leukemic infiltration and/or hemorrhage much more frequently than did the inner ear or external auditory canal. No inner ear involvement and no leukemic infiltration of the mastoid were noted in our patient.

Nonspecific skin findings in leukemias are frequent and very variable. Most of them are the result of impairment function of bone marrow and include purpura, hemorrhage or ecchymoses or they are a manifestation of impaired immunity and include various skin infections. In our case, the patient initially presented with multiple lesions on the right auricle which progressively enlarged to as big as 10 x 7 cm. The incision and drainage was not indicated in this case knowing that the patient was diagnosed as Acute Leukemia. Complications like continuous bleeding and infection could happen.

Skin eruptions are reported to occur in 36% of patients, but most of them have been reported as leukemids, i.e., with no skin infiltration of leukemia cells. Only biopsy could confirm diagnosis. And this was not done in the patient because of the risk of bleeding and infection.

Treatment needs to begin soon after AML is diagnosed, as it progresses very quickly. Chemotherapy is the main form of treatment for AML. It is usually divided into 2 phases: Remission induction (induction) and Consolidation (post-remission therapy).Initially, the aim of treatment is to destroy leukaemic cells and induce a remission.

In our case, patient started on cycles of Chemotherapy (Doxorubicin, Cytarabine and Etoposide). After the first cycle, the patient conditions improved. (Appendix III). Ear examination crusted, non-tender, non-friable mass in right periauricular area, still external auditory canal not visualized. Patient tolerated the chemotherapy treatment and was then discharged. One month after the 1st first cycle, the patient initiated the 2nd cycle of chemotherapy. (Appendix IV). There was noted a decreased size of the mass. Ear examination noted with dry, hyperpigmented, non-tender, wrinkled skin or presence of scarring tissue

on periauricular area. Hyperemic, narrowed ear canal and tympanic membrane not visualized. After one month, the 3rd cycle of chemotherapy has done (Appendix V). A regression of the mass of the right auricle noted with hyper pigmented, wrinkled skin on periauricular area, narrowed external auditory canal with clear fluid, non-foul smell discharge, tympanic membrane not visualized.

In our case, chemotherapy quickly led to resolution of otological manifestations and complete hematologic remission.

The significance of presenting this rare case of skin manifestation in the ear of Acute myelogenous leukemia (AML) is: a.) the awareness of systemic disease like leukemia that could present with skin involvement b.) importance of history taking like bleeding etc. c.) Not all auricle lesions are simply seroma or hematoma d.) Not all auricular lesions are infection that warrants incision and drainage.

It is appropriate for an otorhinolaryngologist to consider complete blood count, peripheral blood smear, and temporal bone imaging and exclude any other systemic disease like leukemia to avoid complications and infection.

CONCLUSION

Enlarging mass of the auricle is not always due to simple trauma, but may be a cause of other detrimental systemic disease entity like Acute Leukemia in our patient. Incision and drainage of seroma or hematoma of the ear should be done with caution and a good history taking is a must to avoid complications like bleeding and infection.

REFERENCES

1. Howlader N, Noone AM, Krapcho M, Garshell J, Neyman N, Altekruse SF, Kosary CL, Yu M, Ruhl J, Tatalovich Z, Cho H, Mariotto A, Lewis DR, Chen HS, Feuer EJ, Cronin KA (eds). SEER Cancer Statistics Review, 1975-2010, National Cancer Institute. Bethesda, MD, http://seer.cancer.gov/csr/1975_2010/, based on November 2012 SEER data submission, posted to the SEER website, April 2013

2. Al-juboori AN (2013) Treatment of Auricular Hematoma with Compression Using X-ray Films. General Med 1: 101. doi: 10.4172/2327-5146.1000101

3. Loock JW (2008) Haematoma auris. In: Gleeson M, Browning GG, Burton MJ, Clarke R, Hibbert J, et al. (eds.). Scott-Brown´s Otorhinolaryngology, head and Neck Surgery (7thedn) New York, Hodder Arnold 3: 3373-3374.

4. Lane SE, Rhame GL, Wroble RL (1998) A silicone splint for auricular hematoma. Phys Sportsmed 26: 77-78.

5. Dr. Ben Wedro. Hematoma. Nov 19, 2013.www.medicinenet.com › ... › skin center › skin az

6. Michael M. Paparella M.D.et al. Otological manifestations of leukemia. The Laryngoscope Volume 83, Issue 9, pages 1510–1526, September 1973

7. Chapman P. Johnson AN. Mastoid chloroma in acute myeloid leukemia. Journal of Laryngology and Otology 1980-94:1423-1427.

8. http://en.wikipedia.org/wiki/Acute_myeloid_leukemia

9. Büchner SA. Specific and nonspecific skin manifestations in leukemia. Schweiz Rundsch Med Prax 2002; 91(24): 1071–7.

10. Jeong Hee Cho-Vega, MD, PhD, et al. Leukemia Cutis. American Society for Clinical Pathology. Am J Clin Pathol 2008;129:130-142

11. Byrd JC, Edenfield WJ, Shields DJ, et al. Extramedullary myeloid cell tumors in cute nonlymphocytic leukemia: A clinical Review. Journal of Clinical Oncology 1995;13:1800-1816.

12. Billström R, Johansson B, Fioretos T, et al. Poor survival in t(8;21) (q22;22)-associated acute myeloid leukaemia with leukocytosis. Eur J Haematol 1997; 59: 47-52.

13. Hiçsönmez G, Çetin M, Tuncer AM, et al. Children with acute myeloblastic leukemia presenting with extramedullary infiltration: the effects of high-dose steroid treatment. Leuk Res 2004; 28:25-34.

14. Çavdar AO, Babacan E, Gözdaşoğlu S, et al. High risk sub-group of acute myelo-monocytic leukemia with orbitoocular granulocytic sarcoma in Turkish children. Acta Haematol 1989; 81:80-85.

15. Gökcan MK,Batı khan H,Çalgüner M, Tatarağası AI. Unilateral hearing loss as a presenting manifestation of granulocytic sarcoma (chloroma). Otology & Neurotology 2005; 27: 106-109.

16. Todd NW, Bowman CA. Acute myelogenous leukemia presenting as aty-pical mastoiditis with facial paralysis. Int J Ped Otorhinolarynogology 1984; 7:173-177.

17. Almadori G, Ninno MD, Cadoni G, et al. Facial nevre paralysis in acute otomas-toiditis as presenting symptom of FAB M2, T8; 21 leukemic relapse. Case report and review of the literature. Int J Ped Otorhinolarynogology 1996; 36:45-52.

18. Kurtz JE, Andres E, Veillon F, et al. The American Journal of Medicine 2000; 15:509 -510.

19. Hsu YC, Su CY, Hsu RF, et al. Unilateral sudden hearing loss as a presenting manifestation of chronic myeloid leukemia: Case report. Otolaryngol Head Neck Surg 2004; 130:271-273.

20. Hsu YC, Su CY, Hsu RF, et al. Unilateral sudden hearing loss as a presenting manifestation of chronic myeloid leukemia: Case report. Otolaryngol Head Neck Surg 2004; 130:271-273.

APPENDIX

I. 1 week PTA

II.

III. After the 1st Cycle of Chemotherapy

IV. 2nd Cycle of Chemotherapy

V. 3rd Cycle of Chemotherapy