Case Report
Open Access
CrossMark

Chloroma: an uncommon presentation of the acute lymphopblastic leukaemia (ALL) in a child

Abstract

Leukemia is the most common malignancy of childhood. Chloromas are a rare presentation of an underlying leukemia. Here, we discuss a rare case of acute lymphoblastic leukemia (ALL) presenting with swelling over right eye. The patient was evaluated radiologically for the extension of disease and treatment planning. ALL was diagnosed with complete blood count and bone marrow examination. The orbital swelling was the initial presentation which led to further diagnosis of acute lymphoblastic leukemia (ALL) in our case. Chloroma is usually a common clinical presentation of acute myeloid leukemia (AML), however, it may rarely be found in a case of acute lymphoblastic leukemia (ALL) such as ours.

Keywords: chloroma, acute lymphoblastic leukemia, ALL, orbital swelling

Volume 11 Issue 1 - 2021

Dr. Amar Taksande,[1] Dr. Rewat Meshram,[2] Dr. Amol Lohakare[3]

[1]Professor, Department of Pediatrics, Jawaharlal Nehru Medical College, India
[2]Associate Professor, Department of Pediatrics, Jawaharlal Nehru Medical College, India
[3]Senior Resident, Department of Pediatrics, Jawaharlal Nehru Medical College, India

Correspondence: Dr. Amar M Taksande, Department of Pediatrics, Jawaharlal Nehru Medical College, Sawangi Meghe, Wardha, Maharashtra 442004, India,
Email amar.taksande@gmail.com

Received: December 13, 2020 | **Published:** February 26, 2021

Introduction

Acute lymphoblastic leukemia (ALL) belongs to a family of hematopoetic neoplasms derived from the B- and T- cell progenitors.[1] In ALL, the malignant B-/T- progenitors multiply uncontrolled, producing an excess of progenitor lymphoblasts in the peripheral blood as well as the bone marrow.[2] Lack of differentiation and excessive proliferation of the progenitor lymphoblasts lead to abnormal hematopoiesis, ultimately resulting in bone marrow failure.[1,2] The extra-medullary form of this disease has been infrequently described in literature. Chloroma, also known as granulocytic sarcoma (GS), is a rare extra-medullary malignant form derived from the myeloid precursor cells. In 1811, it was first described by Burns and, later, in 1853 called Chloroma by King on account of its green color.[3,4] The gross morphology of these neoplasms varies with upto 30% of them not displaying the characteristic green color. These tumors were later renamed as granulocytic sarcomas (GS) due to the varying appearance.[5,6] When these sarcomas precede the development of systemic manifestation of leukemic process, the clinical diagnosis can be extremely difficult. Although, classically described in acute myeloblastic leukemia (AML), these sarcomas have been reported in chronic myeloid leukemia (CML), myelodisplastic syndromes (MDS), hypereosinophilic syndromes, and polycythemias (PT). Herein, we describe a child who presented with swelling over his right eye as the initial manifestation of ALL.

Case presentation

A 6 year old male child presented with history of swelling over right upper eyelid since last one month. Initially, the swelling started in the right upper eyelid and progressively increased involving the orbit resulting in proptosis of the right eye. There was also history of low grade fever since 1 month, which was not associated with chills and rigor, no diurnal variation and relived after taking medications. There was no history of gum bleeds, loss of vision, diplopia, skin bleeds, bone pains, or joint pains. He was fully immunized for his age. At admission, he was afebrile (98 °F), pulse was 86 beats/minute, blood pressure (BP) was 92/60 mmHg, and respiratory rate (RR) was 18 breaths/minute. Palpation of the right eyelid showed firm, immobile mass, located at upper medial orbital region (Figure 1). There were no other significant ophthalmic finding. Child had severe pallor with lymphadenopathy involving submental, supraclavicular, axillay and inguinal lymph nodes. On further examination, the lymph nodes were firm in consistency, non-tender, matted and immobile. Organomegaly was present. While the liver was just palpable, spleen could be palpated approximately 6 cm below the left costal margin. Testis was normal. Central nervous system (CNS) examination did not reveal any abnormality. Fundus examination was normal.

Figure 1 Swelling over the right eye at presentation.

On further investigating, patient had anaemia (Hb-5.8 gm%), total leukocyte count was 1,13,000 cells/cu. mm and platelet count of 25,000/cu. mm. Peripheral smear showed atypical lymphocytes (50%), and lymphoblasts (30%). Patient had normal prothrombin time and activated partial thromboplastin time. Liver function tests and renal function tests were within normal limits. Suspecting a hematological malignancy due to such high counts and atypical lymphocytes on smear, bone marrow aspiration was performed. It showed 35% lymphoblast and 40% lymphocytes 40%. Bone marrow aspiration was suggestive of ALL. Flow cytometry showed positivity for CD19, CD34, CD10, CD45, CD38, CD79a. Flow cytometry confirmed this

case as B-cell precursor ALL. CT orbit revealed an ill-defined soft tissue lesion measuring 2.8 x 0.9 cm seen arising from the antero-superior extra-conal compartment of right orbit and compressing the eyeball posterior-inferiorly, abutting the superior rectus and superior oblique muscle, suggestive of a granulocytic sarcoma. The child was treated as per the children's oncology group (COG) protocol for B- cell precursor ALL. The chemotherapy regimen consisted of L-asparginase, daunomycin, vincristine and dexamethasone. Intrathecal methotrexate was administered as central nervous system prophylaxis. The patient has tolerated the chemotherapy fairly well. The periorbital swelling resolved by day eight following a 4 week course of prednisolone.

Discussion

Leukemias are the most common malignancies of childhood. ALL accounts for almost 77% of these leukemias.[7] Initial presenting clinical features are non-specific and include fatigue, anorexia, malaise, and intermittent low-grade fever. As the disease advances, symptoms of bone marrow failure become more obvious with the onset of pallor, exercise intolerance, bruising, mucosal bleeding and infections.[7] In comparison, children with myelogenous leukemia are may present similarly. However, children with AML are known to develop subcutaneous nodules ("blueberry muffins") and discrete masses (granulocytic sarcoma). These masses are rare solid tumors derived from the primitive precursors of granulocytic series.[2] GS are commonly seen in children aged 7-8 years with a slightly male predominanace. In addition to the usual presentation with proptosis, child may have symptoms of periorbital cellulitis, swelling or mass of the lacrimal gland or eyelids.[9,10] Rarely, when GS is present intracranially, it presents with signs of raised of intracranial pressure or mass effect. Histopathological examination of these lesions have demonstrated these lesions to be composed of primitive granulocyte precursors (myeloblasts, promyelocytes and myelocytes) along with supporting connective tissue and vascular stroma.[2–4] The greenish color imparted to chloroma is due to the presence of myeloperoxidase enzyme in the immature granulocytic cells.[8–10] GS have been reported in association with chronic myelogenous leukemia (CML), and other myeloproliferative diseases, such as myelofibrosis, hypereosinophilic syndrome, or polycythemia vera (PV).[11] Granulocytic sarcoms have been classified by World Health Organization (WHO) into three main subtypes: blastic (composed mainly of myeloblasts), immature (composed of myeloblasts and promyelocytes) and differentiated (composed of promyelocytes and more mature myeloid cells).[12–14] While evaluating an orbital mass in a child, various benign as well as malignant conditions must be considered.

Chloromas are often multifocal and commonly originate in the bones (skull, orbit). Chloromas of the orbit arises from the subperiosteal region of the superior orbital wall with a tendency to invade into the surrounding sclera and the underlying bone.[2] On computed tomography (CT) imaging, chloromas are homogenously iso-attenuating to slightly hyperattenuating relative to brain or muscle. Infiltration into adjacent structures such as globe, extraocular muscles, retro-orbital fat and overlying skin are commonly seen. Calcification is usually absent.[13,14] The management of ALL involves intensive chemotherapy with vincristine, dexamethasone or prednisone, L-asparaginase, and/or an anthracyclin, high-dose methotrexate, cytarabine, cyclophosphamide. This is followed up with an intensification or consolidation therapy to eradicate residual leukemia, ensure continuation of remission and prevent or eliminate CNS leukemia. Our patient had received standard COG chemotherapy regimen consisting of dexamethasone, vincristine, daunorubicin, and intrathecal methotrexate. Newer therapeutic modalities includes drugs targeted at oncgenic kinases such as the janus kinase (JAK) inhibition, protein tyrosine kinase inhibition, Fms-like tyrosine (FLT3) receptor tyrosine kinase inhibition.[8–10,15,16]

Conclusion

In conclusion, for any child presenting with periorbital swelling, malignant etiologies must be thought of in addition to the usual infective causes. Although, granulocytic sarcoma (chloroma) is an infrequently encountered entity, it frequently missed early on resulting in delayed chemotherapy and further complications. Chloromas can be successfully treated with standard chemotherapy regimen which usually results in complete resolution of the lesion as observed in our case.

Acknowledgments

None.

Conflicts of interest

None.

References

1. Onciu M. Acute lymphoblastic leukemia. *Hematol Oncol Clin North Am.* 2009;23(4):655–674.
2. Guermazi A, Feger C, Rousselot P, et al. Granulocytic sarcoma (chloroma): imaging findings in adults and children. *AJR Am J Roentgenol.* 2002;178(2):319–325.
3. Balyen L, Deniz Balyen LS, Pasa Semir. A case of orbital granulocytic sarcoma (chloroma) well responded to the treatment. *Adv Ophthalmol Vis Syst.* 2016;5(1).
4. Dusenbery KE, Howells WB, Arthur DC, et al. Extramedullary leukemia in children with newly diagnosed acute myeloid leukemia: a report from the Children's Cancer Group. *J Pediatr Hematol Oncol.* 2003;25(10):760–768.
5. Stein-Wexler R, Wootton-Gorges SL, West DC. Orbital granulocytic sarcoma: an unusual presentation of acute myelocytic leukemia. *Pediatr Radiol.* 2003;33(2):136–139.
6. Murthy R, Vemuganti GK, Honavar SG, et al. Extramedullary leukemia in children presenting with proptosis. *J Hematol Oncol.* 2009;2:4.
7. Tubergen DG, Bleyer A, Kim Ritchey A, et al. The Leukemias. In: Nelson textbook of pediatrics. 21st ed. Philadelphia, PA: Elsevier; 2020. p. 2649–2656.
8. Pui MH, Fletcher BD, Langston JW. Granulocytic sarcoma in childhood leukemia: imaging features. *Radiology.* 1994;190(3):698–702.
9. Cavdar A, Arcasoy A, Babacan E, et al. Ocular granulocytic sarcoma (chloroma) with acute myelomonocytic leukemia in Turkish children. *Cancer.* 978;41(4):1606–1609.
10. Ab L-S, Rm L, L G, et al. Targeting paediatric acute lymphoblastic leukaemia: novel therapies currently in development. *Br J Haematol.* 2010;151(4):295–311.
11. Thakur B, Varma K, Misra V, et al. Granulocytic sarcoma presenting as an orbital mass: report of two cases. *J Clin Diagn Res JCDR.* 2013;7(8):1704–1706.
12. Jaffe E, Harris N, Stein H. World Health Organisation Classification of Tumours. *Tumours of Haematopoietic and Lymphoid Tissues.* 2001.
13. Kundu S, Chatterjee S, Mondol D, et al. Extramedullary granulocytic sarcoma. *Indian J Med Paediatr Oncol.* 2008;29(3):28.

Citation: Taksande DA, Meshram DR, Lohakare DA. Chloroma: an uncommon presentation of the acute lymphopblastic leukaemia (ALL) in a child. *J Pediatr Neonatal Care.* 2021;11(1):15–17. DOI: 10.15406/jpnc.2021.11.00435

14. Shields JA, Bakewell B, Augsburger JJ, et al. Space-occupying orbital masses in children. A review of 250 consecutive biopsies. *Ophthalmology.* 1986;93(3):379–384.

15. Ott CJ, Kopp N, Bird L, et al. BET bromodomain inhibition targets both c-Myc and IL7R in high-risk acute lymphoblastic leukemia. *Blood.* 2012;120(14):2843–2852.

16. Hossain IT, Moosajee M, Abou-Rayyah Y, et al. Orbital mass secondary to infantile acute lymphoblastic leukaemia. *BMJ Case Rep.* 2016;2016.

Citation: Taksande DA, Meshram DR, Lohakare DA. Chloroma: an uncommon presentation of the acute lymphopblastic leukaemia (ALL) in a child. *J Pediatr Neonatal Care.* 2021;11(1):15–17. DOI: 10.15406/jpnc.2021.11.00435