

Chloroma of Orbit: A Rare Initial Presentation in A Case of Acute Myeloid Leukemia

RASHMI M NAGARAJU, BHIMARAO

ABSTRACT

Leukemia is the most common malignancy of childhood. Chloromas are rare presentations of underlying leukemic disease. It is important to diagnose these cases early in the course as they respond more favourably to focal irradiation. We present a rare case of chloroma of orbit in a young male

patient who presented with severe proptosis and a large mass involving the left orbit. The orbital lesion was the initial presentation which led to further diagnosis of acute myeloid leukemia in our case. The patient was evaluated radiologically for the extension of disease and treatment planning.

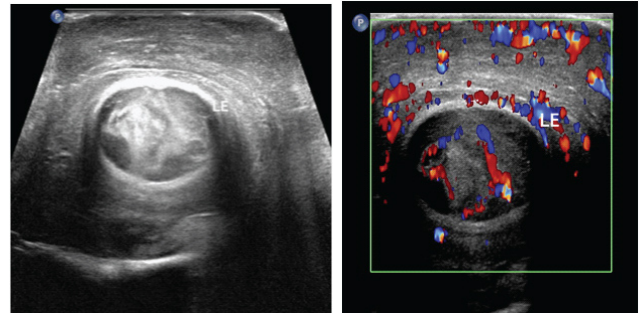
Keywords: Computed tomography, Proptosis, Ultrasound

CASE REPORT

A 16-year-old male presented with progressively increasing painful swelling involving left orbit since three months. The swelling started initially in the left lower lid and progressively increased involving the entire orbit causing axial proptosis which was non pulsatile and irreducible. Movements of the globe were grossly limited. There was complete loss of vision in left eye. Initially it was suspected to be the case of rhabdomyosarcoma. Routine blood investigations and peripheral smear revealed acute myeloid leukemia. Bone marrow examination also confirmed the same. Other investigations including chest X-ray and ultrasound abdomen were normal. He was referred for imaging evaluation to Radio Diagnosis Department, Victoria Hospital, Bangalore. Patient was explained about the radiological procedures and informed consent was taken from the patient and guardians for performing ultrasound and computed tomography.

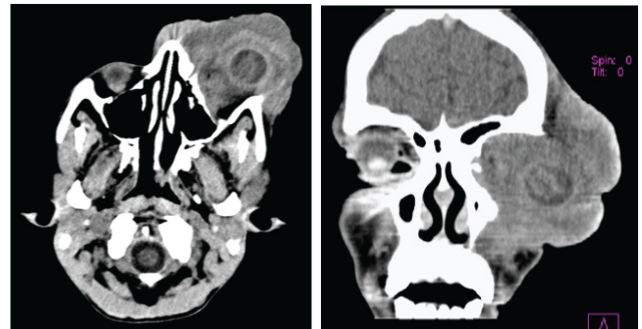
High resolution ultrasonography of left orbit [Table/Fig-1] revealed a large homogenous hyperechoic mass lesion involving both intraconal and extraconal spaces of orbit. There was axial proptosis of left eye with inferolateral displacement. The globe was completely encased with intraocular extension causing distortion. The lesion demonstrated significant internal vascularity on Doppler interrogation [Table/Fig-2].

Plain and contrast enhanced computed tomography of orbits [Table/Fig-3-5] revealed large fairly defined homogeneously enhancing soft tissue attenuating mass lesion involving both intra and extraconal spaces of left orbit. The lesion extended into preseptal and periorbital soft tissues, left superficial temporal fossa, abutting the left lacrimal bone medially, and



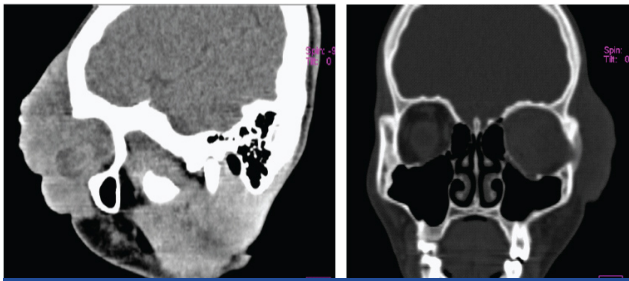
[Table/Fig-1]: High resolution ultrasonography showing homogenous hyperechoic lesion involving the entire left orbit, encasing and infiltrating the globe (left)

[Table/Fig-2]: Ultrasound with colour Doppler showing significant internal vascularity (right)



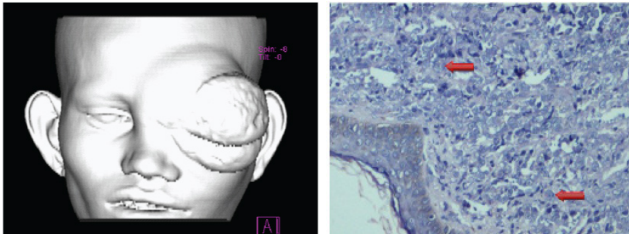
[Table/Fig-3]: Axial contrast CT image showing fairly defined homogeneously enhancing soft tissue attenuating lesion involving left orbit with intra and extraconal involvement, encasing and infiltrating the globe, extending to preseptal, periorbital and superficial temporal fossa regions (left)

[Table/Fig-4]: Coronal reformatted CT image showing extension of lesion (right)



[Table/Fig-5]: Sagittal reformat CT image showing extension of lesion (left)

[Table/Fig-6]: Coronal CT bone window depicting widening of left orbit with remodeling of lateral orbital wall (right)



[Table/Fig-7]: CT with shaded surface display image showing the gross swelling involving left orbit (left)

[Table/Fig-8]: Histopathology showing cells with round to ovoid indented nuclei with eosinophilic cytoplasmic granules (right)

causing remodelling of lateral orbital wall [Table/Fig-6]. Lesion was also seen encasing left globe with infiltration, displacing it antero-inferiorly and laterally. Retrobulbar structures were not delineable. There was no evidence of calcification within the lesion. No obvious intracranial extension was noted. Shaded surface display image from CT showed the gross appearance of the lesion [Table/Fig-7].

Based on laboratory and imaging findings, a diagnosis of chloroma of orbit was made. The differentials considered were rhabdomyosarcoma, metastatic neuroblastoma and Langerhans cell histiocytosis. Patient underwent biopsy of the lesion which showed cells with round to ovoid indented nuclei with eosinophilic cytoplasmic granules, characteristic of myeloid differentiation and was consistent with histopathological diagnosis of granulocytic sarcoma / chloroma [Table/Fig-8]. The patient was referred to regional cancer centre for further management. He underwent systemic chemotherapy and focal irradiation. He returned for follow up after three months, when he had partial marrow response and his orbital lesion had reduced in size with perception of light. He was instructed to continue the chemotherapy course; however the patient was lost to follow up.

DISCUSSION

Chloroma or granulocytic sarcoma is an uncommon solid neoplasm. It is composed of primitive granulocyte precursors namely myeloblasts, promyelocytes, and myelocytes along with supporting connective tissue and vascular stroma [1].

These tumours have been observed in patients with acute myelogenous leukemia, chronic myelogenous leukemia,

and other myeloproliferative diseases, such as myelofibrosis with myeloid metaplasia, hypereosinophilic syndrome, or polycythemia vera [2]. The commonest presentation of chloromas is progression of disease in a case of acute myeloid leukemia (73%) [3].

Leukemias are the commonest paediatric malignancies with 15%–20% of cases arising from myeloid origin [4]. Patients with myelogenous leukemia are prone to develop rare solid tumours of primitive precursors of granulocytic series of leucocytes termed chloromas or granulocytic sarcomas. These uncommon neoplasms can occur in a diagnosed case of myelogenous leukemia or may herald the future development of leukemic disease by months to years. In a patient with known diagnosis, development of chloromas may represent relapse of an already treated systemic disease or may be the presenting sign of associated systemic disease [1,5]. Skin, orbit, CNS and genitourinary system are commonly involved. In the head and neck region, the orbit is the most preferred site followed by skull and epidural spaces. Chloromas of the orbit usually arise from subperiosteal region of superior orbital wall, with a tendency to encase rather than invade sclera and underlying bone [1].

They are common in children than in adults, peak prevalence is in patients aged 7-8 years with slight male predominance. The lesions are often multifocal; and most frequently arise in bones, especially the skull and orbit [3]. Clinical presentation with orbital involvement most commonly is with proptosis. Other symptoms like periorbital cellulitis or swelling or mass in the lacrimal gland or eyelid may be seen. It may be bilateral [6-8]. At radiography, granulocytic sarcoma usually appears as a soft-tissue mass. On ultrasound, a non specific, homogeneous, hypoechoic or echogenic solid mass is seen. The borders may appear infiltrative. On unenhanced CT scans, granulocytic sarcomas are generally homogeneously isoattenuating to slightly hyperattenuating relative to muscle or brain. Invasion of adjacent structures including globe, extraocular muscles, retro orbital fat and overlying skin are commonly encountered. Calcifications are usually absent. On post contrast studies they exhibit uniform homogenous enhancement [1,9].

MR imaging helps in better characterisation of the soft tissue components of the orbit. Chloromas appear isointense to hypointense relative to gray matter or muscle on T1-weighted images, replacing the high signal intensity of the bone marrow. On T2-weighted images they are heterogeneously isointense to slightly hyperintense. The lesion is usually hyperintense in comparison to sclera in all sequences secondary to its low fibrous component, which helps to delineate the extension of actual tumour. On administration of intravenous gadolinium, these lesions demonstrate homogeneous enhancement [1, 9].

Differentials to consider in patients without history of leukemia include rhabdomyosarcoma, orbital cellulitis, metastatic neuroblastoma and Langerhans cell histiocytosis [10]. It is clinically challenging to differentiate granulocytic sarcoma

from other malignancy. In patients with retrobulbar orbital mass causing rapidly progressive proptosis possibilities of malignant and inflammatory processes need to be considered [11].

Rhabdomyosarcoma is the commonest extraocular orbital malignancy in children and much commoner compared to chloromas. It is more likely to cause bone erosions and commonly involves lateral orbit, in contrast to chloromas which have less propensity to erode the bone and which usually involve superior orbit. Orbital cellulitis usually presents with orbital swelling associated with inflammatory signs. Metastatic neuroblastoma usually head and neck, predominantly orbit. There may be multiple lytic lesions containing calcifications and causing aggressive periosteal reaction. Similar lesions in other bones and abdominal mass usually directs to a diagnosis of neuroblastoma. If lesions are bilateral, Langerhans' cell histiocytosis is to be considered in differentials. LCH typically presents with multiple well defined lytic lesions with beveled edges [11]. Possible differentials in a diagnosed case of leukemia include various complications of the disease, namely abscess, hematoma or development of secondary malignancy. Orbital abscess show peripheral wall enhancement and hematomas do not enhance compared to homogenous enhancement of chloromas [11].

As chloromas are manifestations of underlying systemic disease, they are treated accordingly with systemic chemotherapy. Focal irradiation or surgery is useful in intractable cases or in cases causing mass effect on adjacent structures. However, local treatment does not affect overall survival of the patient [12].

CONCLUSION

Chloromas are relatively rare tumours. It is important to diagnose these lesions early in their course as they respond favourably to focal irradiation. CT and MR imaging are helpful in localising and characterising these lesions. They provide valuable information regarding the extension of involvement, which is helpful for treatment planning including biopsy, radiotherapy and post treatment follow up. Although the imaging findings are non-specific, it should be included in

the differential diagnosis of an orbital mass in paediatric age group, especially when multicentric and recurrent.

REFERENCES

- [1] Guermazi A, Feger C, Rousselot P, Merad M, Benchaib N, Bourrier P, et al. Granulocytic sarcoma (chloroma): imaging findings in adults and children. *AJR Am J Roentgenol.* 2002 Feb; 178(2):319–25.
- [2] Liu PI, Ishimaru T, McGregor DH, Okada H, Steer A. Autopsy study of granulocytic sarcoma (chloroma) in patients with myelogenous leukemia, Hiroshima-Nagasaki 1949-1969. *Cancer.* 1973;31(4):948–55.
- [3] Ooi GC, Chim CS, Khong PL, Au WY, Lie AK, Tsang KW, et al. Radiologic manifestations of granulocytic sarcoma in adult leukemia. *AJR Am J Roentgenol.* 2001; 176(6):1427-31.
- [4] Byrd JC, Weiss RB. Recurrent granulocytic sarcoma: an unusual variation of acute myeloid leukemia associated with 8; 21 chromosomal translocation and blast expression of the neural cell adhesion molecule. *Cancer.* 1994 15; 73(8):2107-12.
- [5] Lee SG, Park TS, Cheong JW, Yang WI, Song J, Lee KA, et al. Preceding orbital granulocytic sarcoma in an adult patient with acute myelogenous leukemia with t(8;21): a case study and review of the literature. *Cancer Genet Cytogenet.* 2008;185(1):51-54
- [6] Murthy R, Vemuganti GK, Honavar SG, Naik M, Reddy V. Extramedullary leukemia in children presenting with proptosis. *J Hematol Oncol.* 2009; 24; 2:4.
- [7] Alkatan H, Chaudhry IA. Myeloid sarcoma of the orbit. *Ann Saudi Med.* 2008; 28:461–65.
- [8] Cavdar AO, Arcasoy A, Babacan E, Gözdaolu S, Topuz U, Fraumeni JF Jr. Ocular granulocytic sarcoma (chloroma) with acute myelomonocytic leukemia in Turkish children. *Cancer.* 1978; 41(4):1606–09.
- [9] Noh BW, Park SW, Chun JE, Kim JH, Kim HJ, Lim MK. Granulocytic sarcoma in the head and neck: CT and MR imaging findings. *Clin Exp Otorhinolaryngol.* 2009; 2(2):66–71.
- [10] Sethi A, Ghose S, Gujral S, Jain P, Kumar R. Childhood proptosis: the invaluable, though often overlooked peripheral blood smear. *Indian J Ophthalmol.* 2001; 49:121-23.
- [11] Chung EM, Murphey MD, Specht CS, Cube R, Smirniotopoulos JG. From the Archives of the AFIP. Pediatric orbit tumors and tumor like lesions: osseous lesions of the orbit. *Radiographics.* 2008;28(4):1193-214.
- [12] Bagri PK, Beniwal V, Beniwal SK, Jakhar SL, Sharma N, Kumar HS, Sharma A. Orbital granulocytic sarcoma (Chloroma) as Initial manifestation of Acute Myeloid Leukaemia. *Delhi J Ophthalmol* 2014; 24 (3): 174-77.

AUTHOR(S):

1. Dr. Rashmi M Nagaraju
2. Dr. Bhimarao

PARTICULARS OF CONTRIBUTORS:

1. Assistant Professor, Department of Radiodiagnosis
P K Das Institute of Medical Sciences Palakkad,
Kerala, India.
2. Senior Resident, Department of Radiodiagnosis
P K Das Institute Of Medical Sciences, Palakkad,
Kerala, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Rashmi M Nagaraju,
Nagambika Nilaya, No 86, Shivapura, Srirampura Post,
Manandavadi Road, Mysore- 570008, Karnataka, India.
E-mail: rashmi83nagaraj@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Publishing: Jul 01, 2015