International Journal of Scientific Research in Dental and Medical Sciences

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Orbital Myeloid Sarcoma in Acute Myeloid Leukemia: A Case Report

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ARTICLE INFO

Article history:
Received 16 October 2022
Received in revised form 02 December 2022
Accepted 10 December 2022
Available online 13 December 2022

Keywords:
Consolidation Chemotherapy
Induction Chemotherapy
Leukemia
Sarcoma

ABSTRACT

Orbital myeloid sarcoma is a common presentation in acute myeloid leukemia. A 6 years old girl child presented with progressive proptosis of the left eye for two weeks with associated redness, watering, and reduced vision in the left eye. Imaging showed the left extraconal lobulated avidly enhancing smoothly marginated lesion in the superolateral aspect. Histopathological examination showed leukemic infiltration. Bone marrow analysis confirmed the diagnosis of acute myeloid leukemia with maturation (AML, M2). The patient was treated with induction chemotherapy followed by prophylactic cranial irradiation and subsequent consolidation chemotherapy. The patient expired ten months after diagnosis. Orbital involvement by AML leads to poor survival, even after combination therapy. Patients with proptosis need to be evaluated for malignant conditions, specifically the pediatric age.

1. Introduction

Orbital myeloid sarcoma, composed of immature granulocytes, is an uncommon presentation in acute myeloid leukemia (AML) in the pediatric age group.[1] Accumulating the leukemic cells in the soft tissues or bone, termed myeloid sarcoma, may occur in 2-8% of AML patients.[2] Myeloid sarcomas are most common in certain subtypes of AML, particularly in M5a (monoblastic), M5b (monocytic), M4 (myelomonocytic), and M2 (myeloblastic with maturation).[3] Orbital involvement by AML leads to poor survival, even after combining radiotherapy and chemotherapy.[4] We report a case of 6 years-old girls with proptosis on the left eye as an initial presentation of AML.

2. Case Presentation

A 6 years old girl child presented in the Radiation Oncology Out Patient Department (OPD) with progressive proptosis of the left eye for the last two weeks with associated redness and watering from the left eye with reduced vision. On general examination, she was found to have left eye swelling (Fig. 1).

Fig. 1. Proptosis of the left eye.

She lost appetite, and there were no petechial patches and no pallor. No symptoms of increased intracranial tension were present. There was no peripheral lymphadenopathy or palpable abdominal organomegaly. A non-reducible axial proptosis of the left eye without subconjunctival hemorrhage was discovered during an ophthalmological examination. She had dystocia and trichiasis and was digitally tense on palpation of the left eyeball. Visual acuity was 6/6 in the right eye and 6/36 in the left eye. Ocular movements were not restricted in all directions, and Pupillary reflexes were positive in both eyes. Both contrast-enhanced CT and MRI of orbits showed left eye proptosis due to the left extraconal lobulated avidly enhancing smoothly marginated lesion in the superolateral aspect measuring (3.6 x 2.3 x 2.8) cm (Figs. 2 and 3).

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https://doi.org/10.30485/IJSRDMS.2022.369208.1397

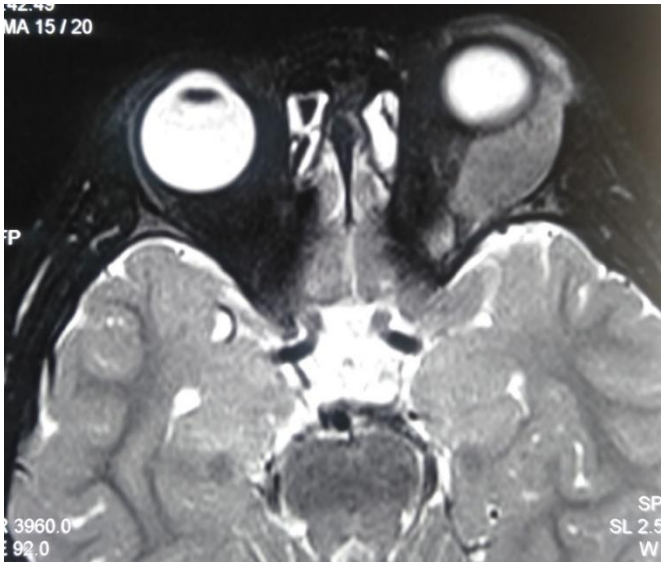

Fig. 2. Axial MR image showing extension of the lesion in Left eye.

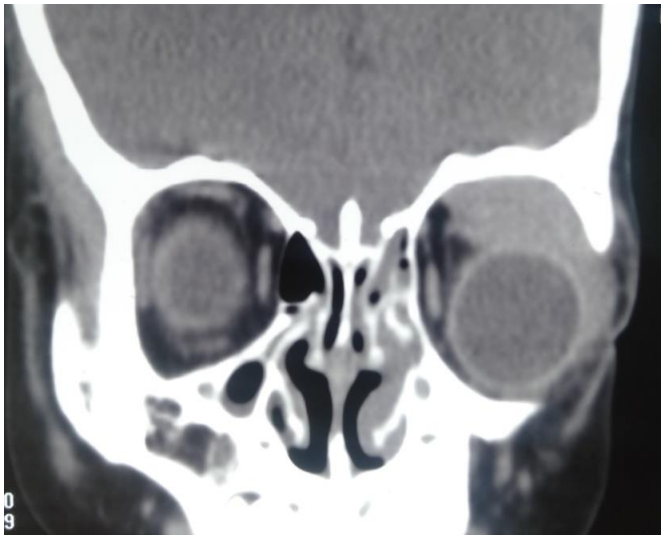

Fig. 3. Coronal CT image showing extension of the tumor in the left eye.

The left lateral rectus appeared bulky. The lesion was seen displacing the superior rectus muscle, lateral rectus muscle, and globe inferiorly. The lesion was also abutting the left lacrimal gland anteroinferior; laterally, the lacrimal gland was not distinctly separate from the mass. The patient initial complete blood count (CBC) was normal, with only 2% immature cells. The patient had undergone excision of the extraconal left eye mass with debulking of the anterior part of the mass. Intraoperatively tumor was infiltrating the levator palpebrae superioris (LPS), lacrimal gland, and surrounding tissues extending from the LPS and lacrimal gland anteriorly to the roof and lateral wall of the orbit; posterior extension could not be obtained. Histopathological examination was suggestive of leukemic infiltration. Immunohistochemistry markers CD3 and CD20 were negative. In her subsequent CBC, hemoglobin was 10.2 gm%, and the total leukocyte count was 19060/mm3; differentiated leukocyte count showed 23% neutrophils, 38% lymphocytes, 1% eosinophils and 32% blast, and platelet count was 150,000/mm3. Peripheral smear showed leukocytosis with myeloblast. The bone marrow aspiration showed hypercellular marrow smears for age; 38% blast cells, 2% promyelocytes, 10% myelocytes, 5% metamyelocytes, 23% band cells with increased myeloid erythroid (M: E) ratio of 3.6:1 and > 3% MPO positive blast cells conforming to the diagnosis of acute myeloblastic leukemia with maturation (AML, M2) (Fig. 4).

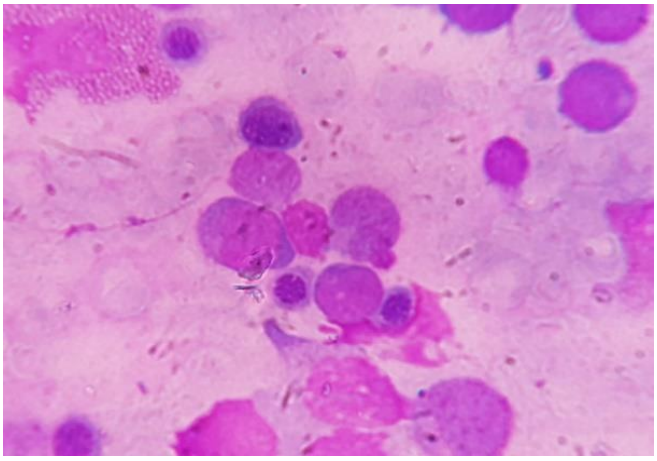

Fig. 4. Bone marrow analysis showing myeloblastic cells (100X magnification, H&E stain).

Genetic study and immunophenotyping were advised, but the family could not afford them. Due to the low platelet count and imaging findings that indicated leukemic proptosis, an incisional biopsy of the orbital swelling was avoided. Both the CSF tests and the brain MRI analysis were normal. As a result, the patient was diagnosed with extramedullary disease affecting the left orbit and AML, M2. Treatment started with induction chemotherapy with an injection of Daunorubicin 45g/m^2 and Cytarabine 200mg/m2 (7 + 3 regimen). After the second cycle of induction chemotherapy, there was partial remission of proptosis. Bone marrow aspiration suggested AML on remission with blast cell 2%. Prophylactic cranial irradiation was done with a Cobalt-60 teletherapy machine for a total dose of 24 Gray in 12 fractions. The family was advised to take the patient to a higher center for bone marrow transplantation, but due to financial conditions, they could not go. The patient came with recurrence after three months. Subsequently, consolidation chemotherapy was given with cytarabine 3 mg/m^2 q12 hourly on days 1, 3 and 5 for 2 cycles, and the patient was in remission for 2-3 months. Unfortunately, the patient expired 10 months after diagnosis at her home.

3. Discussion

The incidence of AML is around 15% of all leukemias in children. About 3% of AML patients will develop granulocytic sarcoma, a rare disease form.[5] Leukemia ocular manifestations are more frequently shown in acute than in chronic leukemias. With involvement in up to 90% of cases, the retina and choroid are the two most common ocular sites of leukemic infiltration.[6] The infiltration of leukemic cells in the extramedullary sites causes granulocytic sarcomas. Allen Burns first referred to it as a "green tumor" in 1811. When leukemic cells are exposed to ultraviolet light, the myeloperoxidase enzyme causes them to appear green; however, the phenomenon may not be present in around one-third of tumors.[7] The ophthalmologist must consider several benign and malignant conditions while evaluating an orbital mass in a child. About 90% of orbital masses of childhood are benign lesions such as inflammation, dermoid cyst, capillary hemangioma, and lymphangioma. Though less common, malignant tumors such as retinoblastoma and rhabdomyosarcoma can be life- and vision-threatening.[8]

Granulocytic sarcoma typically presents on radiographs as a soft-tissue mass. A non-specific, homogenous, hypoechoic, or solid echogenic mass is visible on ultrasonography, and the borders could seem infiltrative.

Granulocytic sarcomas typically appear on CT scans as homogeneously attenuating to slightly hyperattenuating in relation to muscle or brain. The soft tissue components of the orbit are better to see with the help of MRI. On T1-weighted images, chloromas appear isointense to hypointense in relation to gray matter or muscle, while on T2-weighted images, they are heterogeneously isointense to slightly hyperintense. In all sequences, the lesion is typically more hyperintense than the sclera.[9] The role of peripheral blood smear in diagnosing AML in patients with proptosis is pivotal, and subsequent incisional biopsy may be done for further immunohistochemical and genetic marker studies.[10] Chemotherapy is the mainstay of treatment in cases of AML, including both intensive and consolidation phases. Anthracyclins and cytarabine are the most commonly used drugs. Once remission has been achieved, the best option for long-term remission-free survival is an allogenic bone marrow transplant from a matched family donor. Even though the overall survival rate for leukemia patients has increased over the past several decades, patients who acquire orbital leukemic tumors remain to have a high mortality rate.[2,11]

4. Conclusion

Orbital myeloid sarcoma is a rare initial presentation of AML. Patients with proptosis need to be evaluated for any malignant conditions. Peripheral blood smear and image work-up for acute proptosis is necessary for the pediatric age group. CT and MR imaging help localize and characterize these lesions. Overall survival is poor in orbital leukemia patients.

Conflict of Interest

The authors declared that there is no conflict of interest.

Acknowledgements

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

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How to Cite this Article: Baidya K, Dinita Devi N, Nilima Devi N, Singh YI. Orbital Myeloid Sarcoma in Acute Myeloid Leukemia: A Case Report. International Journal of Scientific Research in Dental and Medical Sciences. 2022;4(4):201-203. https://doi.org/10.30485/IJSRDMS.2022.369208.1397.