International Journal of Contemporary Pediatrics
Sonalika C et al. Int J Contemp Pediatr. 2022 Sep;9(9):845-848
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20222124

Case Report

Isolated unilateral orbital myeloid sarcoma: a case report

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Received: 18 July 2022
Accepted: 08 August 2022

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ABSTRACT

Myeloid sarcoma (MS) is a rare solid tumor consisting of immature myeloid cells occurring at an extramedullary site. It can present before, with, or after the manifestation of acute myeloid leukemia or other myeloproliferative diseases, and a few patients never develop bone marrow infiltration. Only a few isolated cases of pediatric orbital MS have been reported, and they are often misdiagnosed. We report a rare case of pediatric isolated orbital MS in a 5-year-old boy who presented with unilateral proptosis. The patient was diagnosed with MS based on MRI and Immunohistochemistry results. Subsequently the patient underwent chemotherapy supported with radiotherapy and showed significant response. Isolated orbital MS presents with clinical and radiological features which are often misleading, making the diagnosis difficult. Therefore, MS should be considered in the differential diagnosis of orbital masses and proptosis even in the absence of acute myeloid leukemia (AML).

Keywords: MS, Orbit, Acute myeloid leukemia, Proptosis

INTRODUCTION

MS is the presence of tumor masses of primitive immature granulocytic precursors in extra-medullary sites such as orbit, central nervous system, bone, subcutaneous tissues, lymph nodes and gastrointestinal tract. Skin and orbit are the most common sites of MS in pediatric population.[1] In pediatric population, orbit is the favored site for MS.[2] Orbital MS can cause diagnostic dilemmas due to its mimickers by clinical as well as radiological findings. Diagnosing orbital MS in the absence of systemic leukemic manifestations is challenging. We present a case of 5-year-old boy who presented with unilateral proptosis with no hematological abnormalities and biopsy of the orbital lesion was confirmative of MS.

CASE REPORT

A 5-year-old boy presented with progressively increasing proptosis of left eye of 9 months duration. There was no preceding history of trauma. Proptosis not associated with fever, mucocutaneous bleeds or visual disturbances. On examination had protrusion of left eyeball with inferior displacement and conjunctival chemosis (Figure 1).

There was no lymphadenopathy or organomegaly. His complete hemogram and peripheral smear were normal. Clinical differential diagnosis included lymphoma, rhabdomyosarcoma, orbital Ewing's sarcoma, or metastatic neuroblastoma. CT orbit showed extraconal lesion in the upper inner quadrant of left orbit suggesting rhabdomyosarcoma (Figure 2).

Subsequently he underwent incision biopsy which showed a diffuse infiltration of round to oval cells of small medium-size and cells had a large oval and indented nuclei, prominent nucleoli and small amount of eosinophilic cytoplasm (Figure 3). Occasional scattered eosinophils were seen.

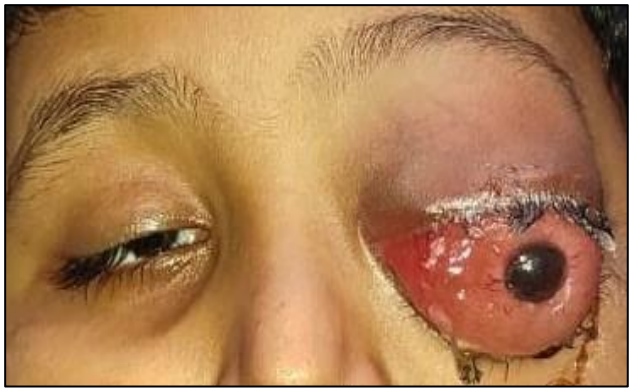

Figure 1: Severe protrusion of left eyeball with conjunctival chemosis and inferior displacement of left eye.

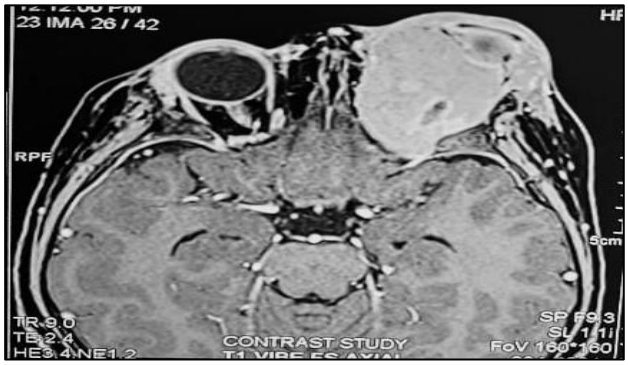

Figure 2: Axial CT orbit showing solid mass lesion in the upper inner quadrant of the left orbit encasing the superior and medial rectus muscles and extending to the intraconal space of left orbit.

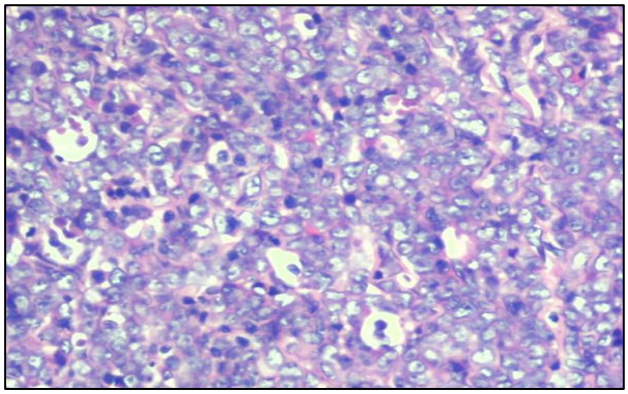

Figure 3: High power histology image from tissue showing small blue round cells with high nuclear: cytoplasmic ratio and prominent nucleoli (eosinophilic myeloblasts).

Immunohistochemistry was positive for myeloid markers (CD45, CD34, CD117 and MPO) and negative for TDT and FLY1, confirming the diagnosis of MS (Figure 4).

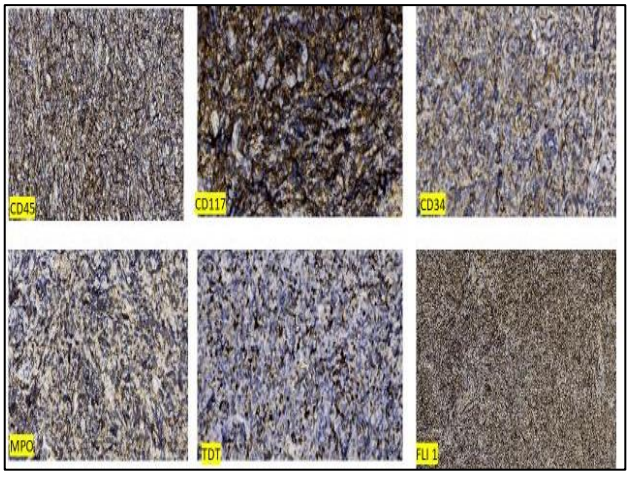

Figure 4: Immunohistochemistry positive for CD 45, CD 34, CD 117, MPO and negative for TDT and FLY1.

Bone marrow aspiration morphology and biopsy, and flow cytometry were negative for malignancy and whole body PETCT showed no significant lesion elsewhere. Child was treated with 2 cycles of induction and 2 cycles of consolidation AML chemotherapy. After first cycle of chemotherapy, there was significant clinical response (Figure 5) and imaging done after 4 cycles of chemotherapy showed a residual lesion for which he received 23.8 Gy radiotherapy. The child is on follow up and is currently disease free for more than 1 year.

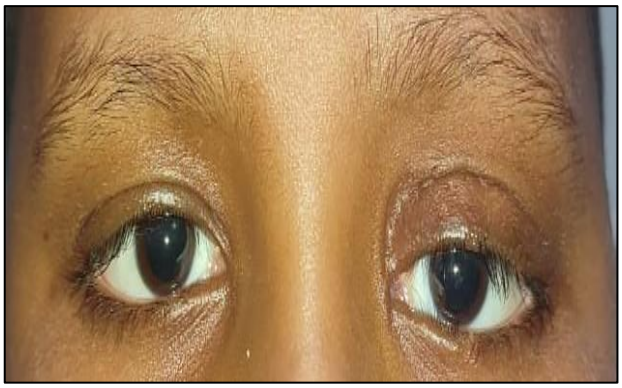

Figure 5: Significant resolution of proptosis after 2 cycles of induction chemotherapy.

DISCUSSION

MS is a rare condition occurring in 2.5-9% of AML cases and 15-35% are concomitant, 25% precede AML and around 50% occur after the diagnosis of AML.[3]

MS originates in bone marrow and the cells spread through Haversian canals, accumulate in sub-periosteum

and form soft tissue masses.[4] The direct infiltration of the orbits in leukemia can result in proptosis, chemosis, diplopia, lid edema, blurring of vision, palsies of the extra-ocular muscle, intra-retinal macular or sub-hyaloid hemorrhages, cotton wool spots, or papilledema due to the increased intracranial pressure.[5] Proptosis is the most common presentation of orbital MS.[3] Isolated unilateral proptosis as initial presentation of AML has been reported, wherein the bone marrow was positive for leukemic cells.[3,6] AlSemari et al has reported 2 cases of orbital MS and bone marrow was negative for leukemia in both cases. About 10% present with isolated lesion without evidence of AML.[7]

Few case reports have reported bilateral orbital involvement as an initial manifestation of AML.[8,9] In orbital MS, the proptosis is due to leukemic infiltrates in the orbital tissues, orbital muscle infiltration, retrobulbar hemorrhage or venous blockage.[10] In most cases, MS progresses to AML in few months but in a few, they did not develop hematologic malignancy for as long as 30 months.[11]

In the absence of systemic leukemic manifestations, the diagnosis of orbital MS can be challenging, and the other differentials are malignant lymphoma, rhabdomyosarcoma, neuroblastoma, retinoblastoma, small round cell tumors, Ewings sarcoma and metastasis.[12] MS is the second commonest orbital tumor after Burkitt's lymphoma that causes proptosis.[13]

On CT, orbital MS appears as a well-defined mass, which can occur intraconally or extraconally. The mass is usually homogeneous, isodense, or hyperdense to brain tissue, with homogeneous enhancement after contrast media injection. The non-enhancing areas corresponds to necrotic regions, which are signs of rapid growth.

In cases of isolated MS, biopsy is the only method for diagnosis. Immunohistochemical staining will be positive for myeloid markers, such as CD45RO/LCA CD68, and myeloperoxidase. Cell surface markers including CD4, CD30, CD34, TdT, and glycophorin A are also useful for diagnosis of MS.t (8:21) is associated with higher incidence of development of systemic leukemia and MS with chromosome 8 abnormalities have worse prognosis.

MS has high propensity to evolve into AML, and so if left untreated, they develop into AML within a year. The treatment strategy is to induce a remission to prevent evolution to AML. The treatment modalities include induction and consolidation therapy with regiments containing cytarabine like those with AML. Low dose radiation is recommended for patients with residual orbital disease after chemotherapy to improve the local disease and quality of life.[14] Allogeneic hematopoietic stem cell transplantation has proven survival benefit for poor risk cases of AML. For isolated orbital MS, combining systemic and local treatment promises a complete remission when compared to any other treatment alone.

In our case, the orbital mass did not present with any hematological manifestations. In such cases a high index of suspicion and proper diagnosis is helpful in prompt and early detection.

CONCLUSION

MS in children may present with unilateral, progressive, non-axial proptosis. Orbital imaging with planned incisional biopsy supported by histopathology may provide the final diagnosis. Pediatric hematology workup (for AML and other myeloproliferative syndromes) and timely management may provide satisfactory long-term outcomes.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Sonalika C, Sneha LM, Jayaraman D, Backiavathy V, Subramanian K, Gayathri SS, John C. Isolated unilateral orbital myeloid sarcoma: a case report. Int J Contemp Pediatr 2022;9:845-8.