Case Report

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Granulocytic Sarcoma Presenting with Unilateral Proptosis: A Rare Disease Entity and Presentation

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ABSTRACT

Granulocytic Sarcoma is an extramedullary tumor composed of immature granulocytic cells. It is also called myelosarcoma or myeloblastoma or chloroma. Granulocytic Sarcomas may develop during the course of the disease, or as a presenting sign of myelogenous leukemia. Herein, we report a case of unilateral GS of the orbit in a 12 year boy, who presented to the eye outpatient department with proptosis of the right eye, and was subsequently diagnosed as acute myeloid leukemia.

Keywords

Granulocytic Sarcoma, Proptosis, AML.

Introduction

Acute Myeloid Leukemia (AML) constitute a malignant clonal proliferation of myeloid series cells with predominance of blasts, replacing the bone marrow, and circulating in the blood with invasion into various organs.

AML may rarely involve the orbit as a solid tumor termed Granulocytic Sarcoma (GS). Granulocytic sarcoma is an extramedullary tumor composed of immature granulocytic cells. It is also called myelosarcoma or myeloblastoma or chloroma. Granulocytic sarcomas may develop during the course of the disease, or as a presenting sign of myelogenous leukemia. Less frequently (in ~ 35% of cases), granulocytic sarcomas may precede AML by months or years [1].

These tumor masses are most frequently located in close proximity to the bone and are often present in perineural and epidural structures, but they may occur anywhere in the body [2,3]. It usually involves the orbits and subcutaneous tissues, but may also occur in the paranasal sinuses, lymph nodes, bones, the spine, the brain, pleural and peritoneal cavities, breasts, thyroid, salivary glands, small bowel, lungs, pelvic organs, and the skin [3-5]. Orbital GS has to be differentiated from rhabdomyosarcoma, metastatic neuroblastoma, African Burkitt's lymphoma, and idiopathic inflammatory pseudo-tumor. It is more common in the pediatric age group than adults and 60% of patients are younger than 15 years [1,6,7]. Herein, we report a case of unilateral GS of the orbit in a 12 year boy, who presented to the eye outpatient department with proptosis of the right eye, and was subsequently diagnosed as acute myeloid leukemia.

Case Summary

A 12 year old male child presented to eye outpatient department with complaints of progressive unilateral proptosis (right side) with mild pain since 1 month. There was no history of trauma or facial sepsis. On examination, the patient was of average built, afebrile, with normal respiratory and cardiovascular system with unpalpable spleen and liver. Multiple firm cervical lymph nodes were palpable, largest of 1.5x1.5 cm in size. (Figure 1) The patient also had a scar mark with a previous history of swelling in neck which had ruptured after some medication from a quack.

On ocular examination, the right eye showed axial proptosis with no limitation of ocular motility (Figure 2). No conjunctival chemosis was noted. Cornea was clear, pupil showed normal light reaction with normal visual acquity. Fundoscopy was normal. Left eye showed no abnormality.

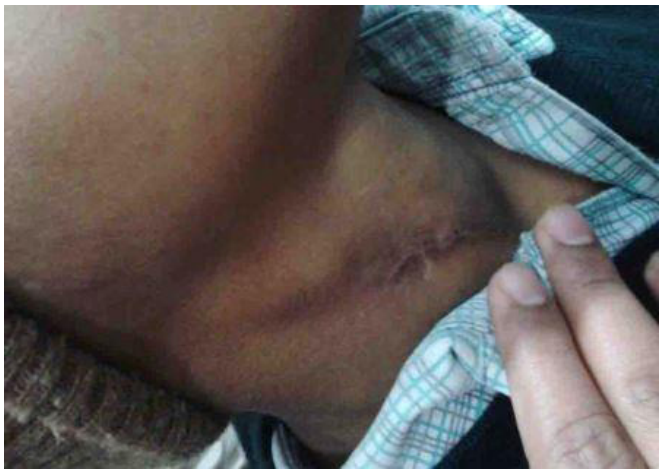

Figure 1: Multiple firm cervical lymph nodes were palpable, largest of 1.5x1.5 cm in size.

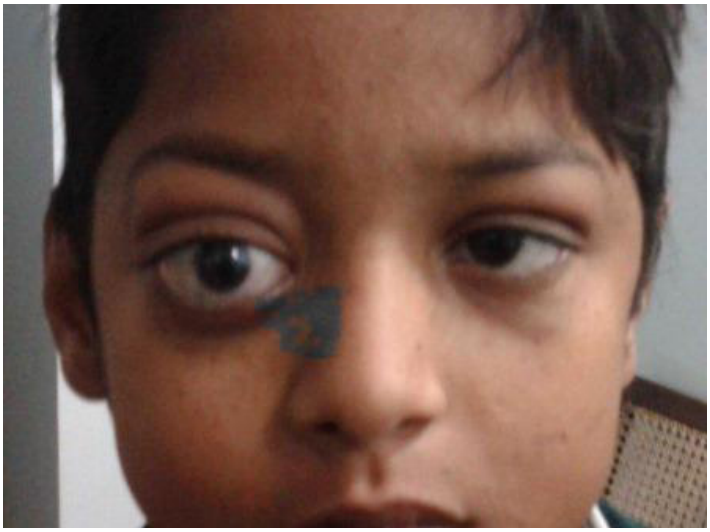

Figure 2: On ocular examination, the right eye showed axial proptosis with no limitation of ocular motility.

A clinical diagnosis of Rhabdomyosarcoma was made and the patient was advised routine blood investigations with CT scan of the head. Patient's hemoglobin was10gm/dl, with a total leucocyte count of 10,000 cells/mm^3 and platelet count of 50000 cells/mm^3. On smear examination, the red cells showed normocytic and normochromic picture with a differential count of 60% myeloblast, 8% promyelocyte, 5% myelocyte, 2% metamyelocyte, 10% neutrophils and 15% lymphocytes. Morphologically, the blasts showed a compact chromatin network with 2-3 inconspicious nucleoli and granular dark blue scanty cytoplasm.

Bone marrow aspiration smears showed hypercellularity with normoblastic erythrocytes and marked proliferation of myeloid series cells with myeloblasts (70%) and promyelocytes (10%) with myelocytes and metamyelocytes and erythroid precursors. MPO was positive in more than 50% of the cells whereas PAS stain was negative (Figure 3). Megakaryocytes were of normal morphology. On flow cytometry, CD 34, CD33, HLA- DR was positive whereas lymphoid and monocytic markers were negative. On the basis of these investigations, a diagnosis of acute myeloid leukemia (FAB -M2) was made. Other biochemical investigations were within normal range for the age. Fine Needle Aspiration Cytology of the cervical lymphnodes showed reactive changes only, with no infiltration of leukemic cells.

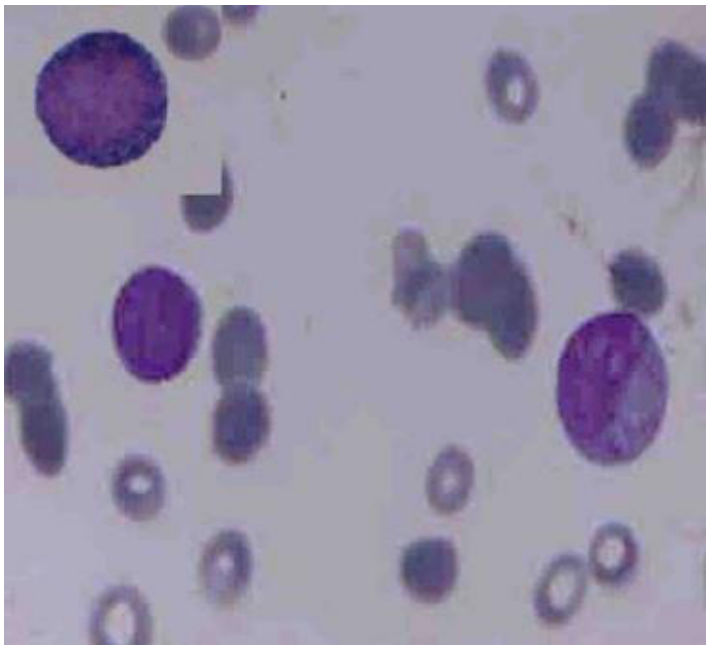

Figure 3: Bone marrow aspiration smears showed hypercellularity with normoblastic erythrocytes and marked proliferation of myeloid series cells with predominance of myeloblasts. Inset shows Myeloperoxidase positive Myeloblast. Leishman x 40.

Computed tomography scan showed a large, homogenously contrast enhancing soft tissue lesion of size 3.5x2.6x2.3 cm in the extraconal space at the supero-lateral aspect of the right orbit. (Figure 4) The lesion was seen to displace the lateral rectus medially and globe anteriorly, causing proptosis. Based on the CT scan findings and clinical examination, a final diagnosis of Granulocytic Sarcoma was made.

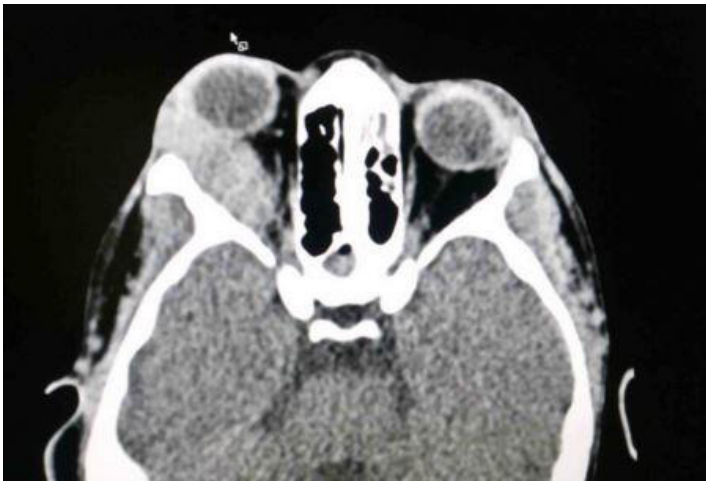

Figure 4: Contrast-enhanced axial CT image of the orbit showed a large homogeneous mass with uniform contrast enhancement located in the right orbit at the supero-lateral aspect, displacing lateral rectus medially and globe anteriorly.

Discussion

Granulocytic Sarcoma is more common in children, with a median age of 7 years and is present in 2.5-9.1% of patients with AML. GS has a slight male predominance [6,7].

Proptosis is seen in 86% of cases of GS with over sixty percent presenting bilaterally [8]. It is often slowly progressive and may occur at any point of time in the disease process. Zimmerman, 1975 reported in review of 33 cases of myeloid sarcoma, that usually clinical presentation preceded systemic leukemia [9]. The proptosis in cases of leukemia tends to be bilateral with other features like conjuctival chemosis, corneal ulceration, blurring of vision and restriction of ocular movement [10-12].

In a prospective study of 120 newly diagnosed cases of leukemia, 3% had primary ocular involvement, 39% showed secondary manifestations, and 5% had visual loss [5]. Ophthalmic findings may precede the finding of leukemia or occur concurrently with the systemic disease during blast crisis, or present as the first sign of relapse, with or without bone marrow involvement. GS is a diagnostic challenge in patients presenting without prior history of leukemia and illustrates the importance of detailed ophthalmic evaluation.

Clinical and hematological evidences may raise suspicion of orbital granulocytic sarcoma but the diagnosis is proven by biopsy. An elevated white blood cell count and peripheral blasts and medullary blasts percentage of more than 5% and 30% respectively, with the presence of auer rod, is pathognomonic of AML [13]. In the present case, the peripheral blood was diagnostic of AML. Since the visual acquity was good with no other ocular problem, and CT scan was highly suggestive of granulocytic sarcoma, so biopsy or FNAC was not advised in the present case. We should always suspect GS in a child presenting with proptosis after excluding all the other causes.

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