Case Report

Extra Medullary Myeloid Cell Tumor - A Case Report

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Keywords: *Extra Medullary Myeloid Cell Tumor, Acute Myeloid Leukemia, Parotid Swelling*

ABSTRACT

Extra medullary myeloid cell tumors or granulocytic sarcomas or chloromas represents a localised tumor of myeloblasts or monoblasts infiltrating extramedullary sites. The term is used for any solid collection of leukemic cells in the extra haematopoietic sites. There have been reports of these tumors occurring concomitantly with, after, or rarely prior to the onset of leukemia. We present a case of a child presented with swelling in the right parotid region, which was diagnosed as chloroma on fine needle aspiration cytology. Further confirmation was done on peripheral blood and bone marrow evaluation.

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Introduction

Extra medullary myeloid cell tumor or granulocytic sarcoma is a localised tumor of myeloblasts or monoblasts infiltrating extramedullary sites [1,5]. These tumors are called chloromas, the term is derived from Greek word chloros (for green) because some appear green or turn green in dilute acid secondary to expression of myeloperoxidase [1,3,4]. These tumors are reported in 3.1- 9.1% of patients with myeloblastic leukemias and occur concomitantly with, after, or rarely prior to the onset of leukemia in 0.6% of cases or upon relapse.

We report a case of a child presented with swelling in the right parotid region. The diagnosis was suspected on fine needle aspiration cytology (FNAC) of parotid swelling which subsequently led to the diagnosis of the underlying acute leukemia.

Case Report

A 3 year male child presented with painful swelling in the right parotid region of 1 month duration. On examination, the swelling was diffuse, 5x4 cm, firm in consistency; tender [Fig-1]. Overlying skin was normal with no local rise of temperature. There was no hepatosplenomegaly or generalised lymphadenopathy. Previous complete blood counts done out side were within normal limits. Chest x-ray was normal. Clinical diagnosis was chronic sialadenitis.

FNAC was done from the parotid swelling. Both wet fixed and air dried smears were prepared for H&E and Leishman staining respectively. Leishman stained aspiration smears showed scant cellularity comprising a few atypical blastoid cells mixed with blood elements. The cells were round, discrete, large with round to oval nuclei, fine chromatin and 1-2 prominent nucleoli (Fig-2). Normal parotid tissue was not seen. CECT Brain was done, which revealed a homogenously enhancing mass lesion centered in the right intrazygomatic temporal region, encircling the mandible with intracranial extension causing erosion and thinning of right ramus of mandible [Fig-3]. Extramedullary myeloid cell tumor was suspected and hemogram was repeated in our hospital.

Peripheral smear examination showed normocytic, normochromic anemia. Total leucocyte count was 9000 cells/mm^3 with differential count showing 15% of blasts (Fig-4). Platelets were adequate. Bone marrow showed reduction in the erythroid precursors and megakaryocytes and 40% of blasts (Fig-5). There were no immature eosinophils. Based on peripheral smear and bone marrow morphology, the diagnosis of acute myeloid leukemia –M2 was made. Subsequently, flow cytometry was performed on bone marrow aspirate, which showed positvity for myeloid markers. So the final diagnosis was AML-M2 with extra medullary myeloid cell tumor in right parotid region.

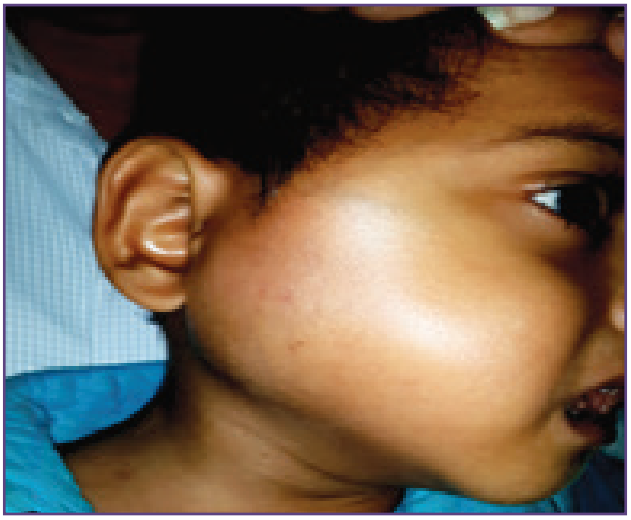

Fig. 1: *A 3 year old child with right parotid swelling*

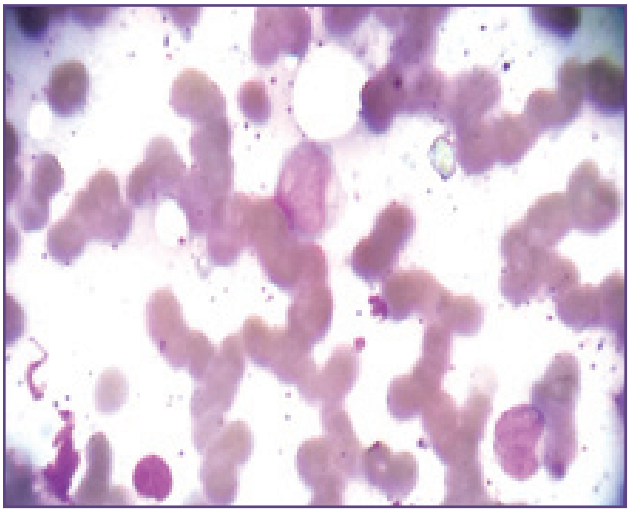

Fig. 2: *FNA of the swelling showing atypical blastoid cells against haemorrhagic background. Leishman stain, x100.*

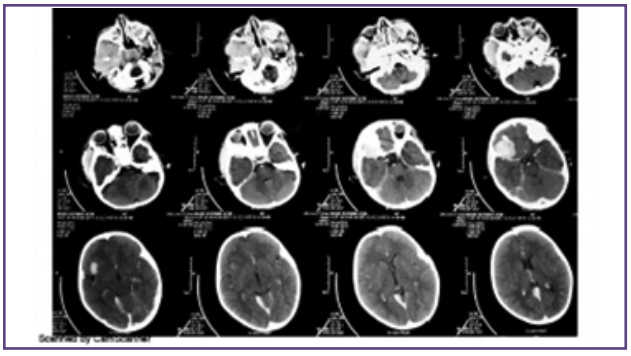

Fig. 3: *CECT Brain showing homogenously enhancing mass in the right infra temporal fossa with intracranial extension into right frontal region*

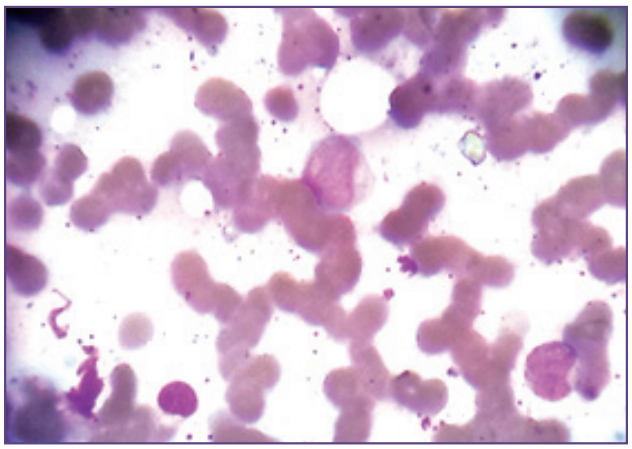

Fig. 4: Myeloblasts with auer rodson peripheral blood smear. Leishman, x100

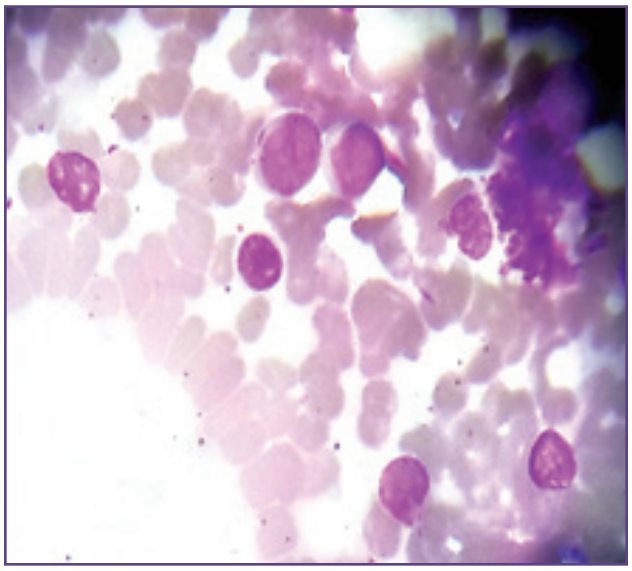

Fig. 5: Myeloblasts with auer rods on bone marrow aspiration smear. Leishman, x100.

Discussion

Myeloid (granulocytic) sarcoma or myeloblastoma or chloroma or extramedullary myeloid cell tumor is an extramedullary deposit that occurs in 2-14% of cases of acute myeloid leukemias.[1,2,3,4]. These tumors are called chloromas because some appear green or turn green in dilute acid secondary to expression of myeloperoxidase [1,3,4]. These tumors are usually localised. They often involve bone,periosteum,soft tissues, lymph nodes or skin. Common sites are the orbit and the paranasal sinuses, but other sites reported include the gastrointestinal tract, genitourinary tract, breast, cervix, salivary glands, mediastinum, pleura, peritoneum and bile duct.[1,3] Extra medullary myeloid cell tumors in the soft tissues may present months before the involvement of peripheral blood and bone marrow[10].

Granulocytic sarcomas may occur at the diagnosis of acute myeloid leukemia or may precede the diagnosis in 0.6% of cases or may occur upon relapse. They have also been seen in association with myelodysplastic syndrome or myeloproliferative disease and usually predict transformation to acute leukemia [2,3,4]. The WHO has classified EMCT or granulocytic sarcomas into 3 main types, depending upon the degree of maturation: Blastic- composed mainly of myeloblasts, Immature- myeloblasts and promyelocytes, Differentiated- promyelocytes and more mature myeloid cells. Rarer types consists of a monoblastic sarcoma, associated with monoblastic leukemia[7].

The diagnosis of EMCT can be difficult. As in this case, the disease is often not suspected on clinical grounds and a high index of suspicion is needed. Clinically and cytologically, the diagnosis needs to be distinguished from the other haematological malignancies like Hodgkin's lymphoma, Non Hodgkin's lymphoma and other small blue round cell tumors. Peripheral smear and bonemarrow aspirate showed presence of blasts, few of them showing characteristic auer rods in the cytoplasm.

The diagnosis can be made if auer rods are detected or if myeloid origin is confirmed by cytochemical and immunohistochemical methods. Neutropenia is common in patients with granulocytic sarcomas. The marrow may reveal increased number of blasts.

A helpful finding in the diagnosis of AML is the presence of Auer rods with romonowsky stains. Auer rods are linear or spindle shaped, red-purple cytoplasmic inclusions in myeloblasts or promyelocytes. Auer rods are derivatives of azurophilic granules and stain positively for SBB, MPO, CAE and acid phosphatases. Auer rods can be found in any of the subtypes of AML, but they are especially associated with M1 to M3.

The diagnosis on FNAC depends on recognising the nature of the primitive cells that can often be mistaken for solid tumors including Non Hodgkin's lymphomas, amelanotic melanomas or undifferentiated carcinoma. The diagnosis is facilitated by making touch imprint preparations of fresh cut sections of the tumors and staining them with romonowsky stains by cytochemical reactions and immunophenotyping.

In formalin fixed, embedded tissue the naphthol ASO chloroacetate esterase reaction and the use of immunocytochemical methods employing polyclonal and monoclonal antibodies against determinants found in myeloid and monocytic cells may be very helpful.

AntiMPO and CD68 are particularly useful for granulocytic or monocytic lineage in paraffin sections. In addition, ultrastructural examination of these tissues may be crucial. Immunophenotyping of bone marrow aspirate by flow cytometry shows a predominant myeloblast population that is positive for CD34, CD13, CD33, CD4 and HLA-DR. These blasts are negative for CD14, CD16, CD56, CD19, CD10, CD117.

Certain risk factors for developing extra medullary myeloid cell tumors associated with any underlying myeloid disorder have been recognised. FAB types M2 and M5 of the underlying leukemia, expression of the surface markers CD2,7 and 56, and cytogenetic abnormalities t (8;21) and inv (16)[8]. Our case also presented withAML-M2 with extra medullary myeloid cell tumor in right parotid region.

Fine needle aspirate from the tumor mass reveals presence of myeloblasts and can be confirmed by MPO stain/ CD13 or CD68 positivity.

Prognostic significance of EMMT is not very well known. It is regarded as a poor prognostic indicator by some, whereas others have not considered it as an independent prognostic factor [11]. The main stay of treatment of EMMCT is treating the underlying leukemia. Most tumors, whether detected prior to the therapy of leukemia, or concurrent with leukemia, respond well to standard chemotherapeutic agents. Evidence of residual tumor may be an indication of surgery or radiation therapy. Median survival is apporoximately 22 months, with worse outcomes more common in elderly patients [9].

Conclusions

Extra medullary myeloid cell tumors or granulocytic sarcomas are very rare tumors. High index of suspicion and awareness of the entity are needed in diagnosing on cytology, especially in cases presenting prior to the onset of leukemia. Correlation and close follow up with haematological parameters is mandatory to detect it early and to avoid unnecessary delay in the treatment.

Acknowledgements

We acknowledge Dr. P. ShanmugaRaju for assisting and guiding us.

Funding

None

Competing Interest

None

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