A Rare Case of Acute Myeloid Leukemia Presenting as Myeloid Sarcoma of Paranasal Sinuses with Intracranial Extension

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Abstract
Myeloid sarcoma (MS)1 is a rare localised extramedullary tumor of myeloid precursor cells and often represents a subtype of myeloid leukaemia. MS of paranasal sinus origin is very rare. Here we report a case of 43 year old male who presented with Myeloid Sarcoma of ethmoid sinus with intracranial extension.

Introduction
Acute myeloid leukemia (AML) is a malignant disease of the bone marrow in which hematopoietic precursors are arrested in an early stage of development. Most AML subtypes are distinguished from other related blood disorders by the presence of more than 20% blasts in the bone marrow. The underlying pathophysiology in AML consists of a maturational arrest of bone marrow cells in the earliest stages of development. Several factors have been implicated in the causation of AML, including antecedent hematologic disorders, familial syndromes, environmental exposures, and drug exposures.

Case Report
43 year old Male was admitted with left sided headache and left upper eyelid swelling2 for 1 month.

- On examination-there was no lymphadenopathy or hepatosplenomegaly. Laboratory investigation-Hb-7.1%,Rbc-2.12million/cmm, TLC-3000cells, platelets-1.11lac/cmm.

- CT & MRI Brain3 identified a T1/T2 hypointense lesion involving bilateral ethmoidal sinuses with erosion of air cells and intracranial extension of lesion involving cribiform plate with adjacent patchy leptomeningeal enhancement, few areas of GRE susceptible within the lesion, with evidence of mucosal thickening involving pan sinuses suggestive of either invasive fungal sinusitis or lymphoproliferative deposit.

- Nasal endoscopy detected grey coloured homogeneous mass occupying the left nasal cavity was detected, biopsy showed immature blast like cells with MPO positivity.

- Peripheral smear-pancytopenia with Blast and atypical cells-35%, N-19%, L-45% E-1%, few cells showed Auer rods, with Sudan Black B positivity. Patient was diagnosed to have MS of ethmoid sinus with intracranial extension.
• Patient was advised chemotherapy and allogenic bone marrow transplant and referred for the same.

CT PNS-The lesion is causing bone erosion of ethmoid air cells with bone defect in anterior cranial fossa, superior to lesion involving the cribiform plate.

Ill defined infiltrative T1\T2 isointense lesion noted involving anterior and posterior ethmoidair cells.

Evidence of GRE blooming noted within ethmoidal lesion

Vasogenic edema is noted in bilateral basifrontal regions predominantly in white matter on right side

The lesion shows enhancement with adjacent patchy meningeal and leptomeningeal enhancement and thickening
Discussion
Myeloid sarcoma (MS) is a rare disease entity that can present as an isolated extramedullary tumor (EM) of immature granulocytic cells. It was first described in 1812 and later named chloroma by King, due to its green color attributed to the presence of myeloperoxidase enzymes. MS has been reported in 2.5–8.0% of patients with acute myeloid leukemia (AML) and occurs concurrently with or at relapse of bone marrow leukemia. MS of the paranasal sinuses is very rare, and the clinical and pathological characteristics are therefore not fully understood. In conclusion, granulocytic sarcoma is a rare complication of AML which may develop during the course of the disease. Intracranial extension may occur through perineural spread.

Conclusion
Any extramedullary tumor showing myeloid precursor cell, should be investigated for MS. MS is easily misdiagnosed as solid tumor or invasive fungal sinusitis.

Reference