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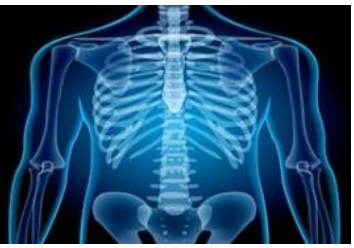

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Rare sites of extramedullary hematopoiesis involving the bilateral orbits, paranasal sinuses and dura in a patient with acute myeloid leukemia

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Abstract
Hematopoiesis usually occurs in bone marrow in adults and when it occurs at sites except for bone marrow, it is termed as extramedullary hematopoiesis (EMH). It is usually found in organs, which are vigorously involved in fetal hematopoiesis, including the liver, spleen, and lymph nodes. Extramedullary hematopoiesis is a complication of a number of myeloproliferative disorders. MR imaging of the brain was performed on a patient with right orbital swelling with a known myeloproliferative disorder. The study revealed symmetric, bilateral enhancing masses in the extra-conal spaces and paranasal sinuses and multiple well-defined lobulated and ovoid meningeal deposits over bilateral cerebral convexities with diffuse pachymeningeal thickening.

Keywords: Extramedullary hematopoiesis, myeloproliferative disorders, diffuse pachymeningeal thickening, extra-conal spaces and proptosis

Introduction
Extramedullary hematopoiesis (EMH) refers to the ectopic development of hematopoietic tissue outside the bone marrow [1]. It is a compensatory mechanism by which the body attempts to maintain a level of erythrogenesis sufficient for its demands [2]. This response occurs in a variety of disorders characterized by a chronic hematopoietic deficit. EMH arises from multi-potential cells in any tissue, such as the liver, spleen, lymph nodes, paraspinal regions, kidney, pleura and intestine, but intracranial hematopoiesis is rare [3]. EMH patients can be divided into two groups. The first shows para-osseous foci that herniate from the medullary tissue of underlying bone. This response is seen in hemolytic disorders such as thalassemia where the marrow is hyperactive. The other group shows extraosseous soft-tissue foci arising from multipotential stem cells where the marrow activity is ineffective as in myelofibrosis [2].
EMH, as a tumor-like mass, is not uncommon and usually originates from the costovertebral angles of the upper thoracic regions, the anterior end of the ribs and the presacral region. However, these masses have been reported in the liver, spleen, lymph nodes, pleura, thyroid, breast, kidneys, maxillary antra and the falx cerebri [3–6]. The most frequently reported causes of intracranial involvement by EMH are thalassemia (50%) and myelofibrosis (31%) [1, 7].
A case report of A 20-year male presented to the emergency room with a history of progressively increasing right eye swelling (proptosis) in the last 8-10 days with headache, generalized abdominal and joint pain and weakness. An contrast enhanced - MRI Brain and CT head scan was performed, and the patient was found to have enhancing homogenous masses in extra-axial cerebral convexities, scalp and bilateral orbital extra conal spaces with paranasal sinuses.

Case Report
A 20-year-old male child was admitted to the emergency room at regional hospital with history of progressively increasing right eye swelling (proptosis) with diminution of vision in the last 8-10 days with generalized abdominal and joint pain and weakness. Admission examination: temperature 37.6 ℃, pulse rate 107 bpm, blood pressure 106/62 mmHg.

Laboratory tests showed severe anemia and thrombocytopenia with a hemoglobin level of 4.3 gm%, platelet count of 20.000/cu mm, and extremely high total leucocyte count of 85,500 cells/cu mm with predominant immature cells and polymorphs. The liver function test and kidney function test were non-significant. The patient was apparently well 2 months ago after which he has 2 febrile episodes with pain in bilateral hip joints and was diagnosed with synovitis on USG. Then he started developing diffuse abdominal pain with vomiting and loss of appetite and was suspected to have abdominal Koch's after which he presented to emergency with the above-mentioned complaints.

An contrast enhanced MRI brain with orbit was performed at the radiology department by two experienced radiologists, and MR Brain with orbit showed - Multiple well-defined lobulated and ovoid, homogenously T2 hypointense extra-axial dural-based lesions largest measuring ~ (3) cm over bilateral cerebral convexities, left temporal region and just outside skull base in the scalp region showing moderate to intense homogenous enhancement with mass effect in the form of sulcal effacement, mild vasogenic edema over frontoparietal regions with diffuse pachymeningeal thickening. Right eye proptosis with focal T_2 hypointense lesions with homogenous enhancement in the bilateral superolateral extra conal space with compression over the lacrimal gland (right >left) with mass effect over the extra-ocular muscles displacing them infero-medially. T_2 hypointense mass, isointense T_1 signal noted within bilateral maxillary, frontal and sphenoid sinus with extension into posterior ethmoid sinuses. The diagnosis of Extra medullary hematopoietic tissue was made. The patient underwent bone marrow biopsy from iliac bone and was found to have acute myeloid leukemia.

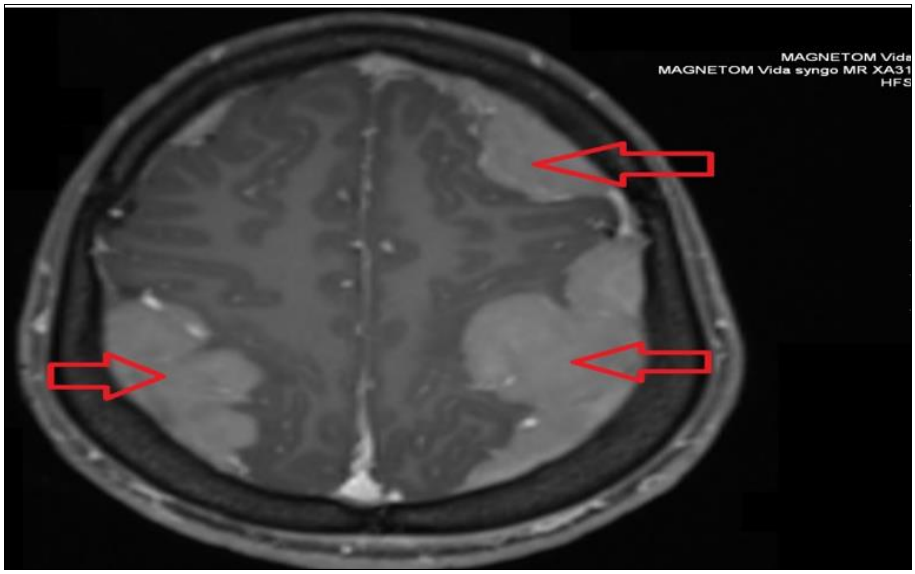

Fig 1: Contrast-enhanced MRI Brain shows multiple well-defined lobulated and ovoid extra-axial dural-based lesions over bilateral cerebral convexities showing moderate to intense homogenous enhancement (Red arrows)

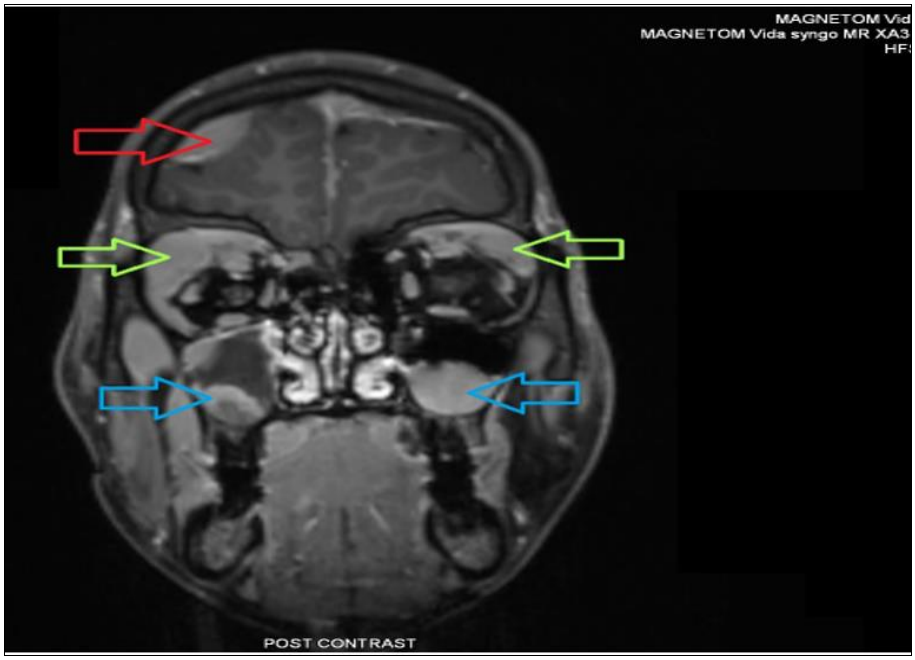

Fig 2: Contrast-enhanced MRI Brain showing focal T2 hypointense lesions with homogenous enhancement in the bilateral supero-lateral extra conal space (green arrows) andT2 hypointense mass with heterogenous enhancement in bilateral maxillary sinuses with dural masses (red arrow)

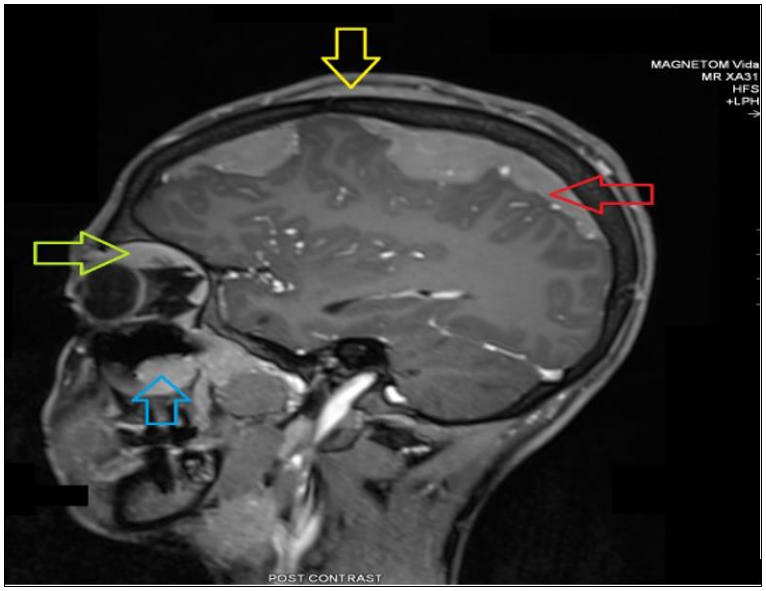

Fig 3: CE-MRI Brain showing rare sites of extra medullary hematopoiesis 1. Dural masses (red arrow) 2. Extraconal orbital masses (green arrows) 3. Maxillary nasal masses (blue arrows) 4. Scalp region (yellow arrow).

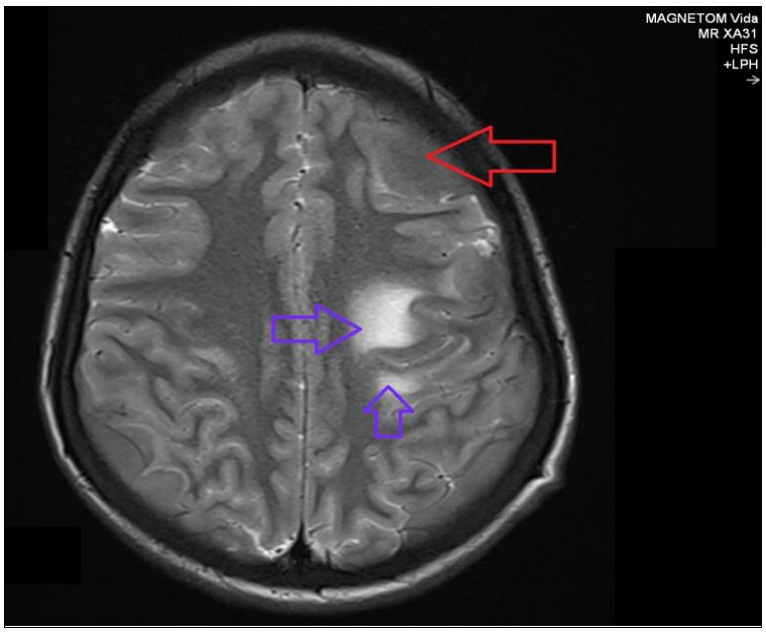

Fig 4: MRI Brain showing T2 hyperintense vasogenic edema (purple arrows) along with Hypointense ovoid dural mass (red arrow).

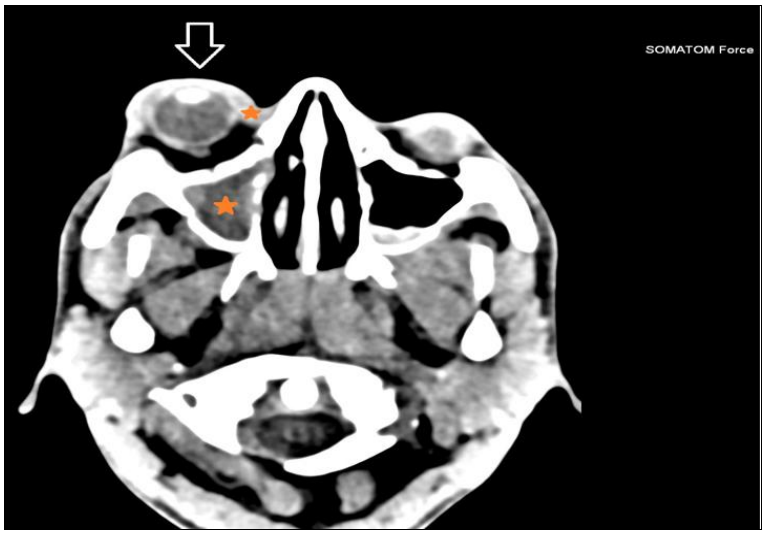

Fig 5: CT Head and Orbit reveals right eye proptosis (white arrow) with soft tissue density masses in bilateral maxillary sinuses and extra conal space of right orbit (orange star).

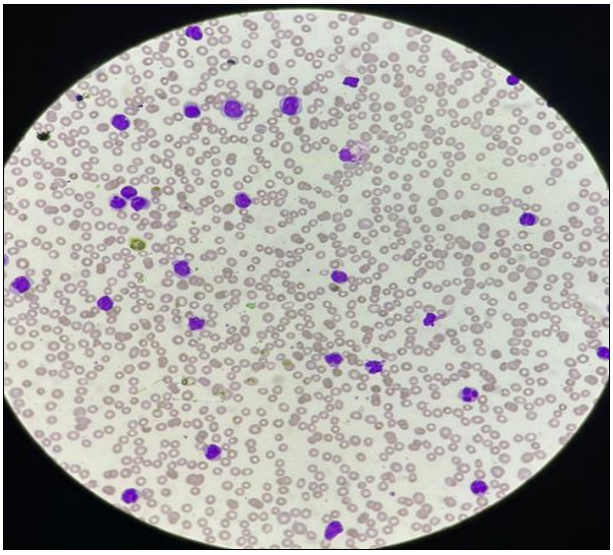

Fig 6: Bone marrow biopsy showing acute myeloid leukemia, arrows indicate myeloblasts

Discussion

Intracranial involvement in extramedullary hematopoiesis (EMH) is rare, but it should be suspected in patients with acute myeloid leukemia presenting with chronic severe headache and anemia. We present a 20-year-old boy with known myeloproliferative disorder whose headaches and anemia were unresponsive to routine treatment presenting with acute progressive right orbital swelling. CT and MRI studies of the brain showed diffuse pachymeningeal thickening. It is generally believed that EMH is a compensatory process associated with anemia and other bone marrow diseases which leads to chronic hematopoietic deficiency. EMH sites in adults are most commonly liver, spleen, and lymph nodes. However, EMH can occur in any site including the central nervous system. Intracranial EMH is a rare event and is most frequently associated with thalassemia and myelofibrosis [8]. The cranial dura, especially the falx, is the most commonly involved site. Any sinus can theoretically be affected, but the most commonly affected location was the maxillary sinus. There were 9 cases with maxillary involvement, 6 with sphenoid involvement, and 4 with ethmoid involvement [9].

Conclusion

In summary, in this study, we described the clinical features of a pediatric case of myeloid leukemia with blast crisis with extramedullary hematopoiesis and multi-organ involvement from Lucknow. Given the acute onset of symptoms and its rare presentation involving the dura, orbits and paranasal sinuses in a patient with chronic anemia, a biopsy is required to diagnose EMH and rule out an underlying malignancy. We recommend that EMH be included in the differential diagnosis of a soft tissue, expansile sinus and orbital mass presenting in patients with known hematologic conditions It should be differentiated from Langerhans cell histiocytosis, lymphoma and Erdheim-Chester and Rosai-Dorfman disease.

Conflict of Interest

Not available

Financial Support

Not available

References

1. Rizzo L, Greco Crasto S, Tola E, *et al*. Extramedullary hematopoiesis: unusual meningeal and paranasal sinuses presentation in paget. Case report. La Radiol Med. 2003;105(4):376–381.
2. Lund R, Aldridge N. Computed tomography of intracranial extramedullary hematopoiesis. J Comput Assist Tomogr. 1984;8(4):788–790.
3. Ohtsubo M, Hayashi K, Fukushima T, *et al*. Case report: intracranial extramedullary haematopoiesis in post polycythemic myelofibrosis. Br J Radiol. 1994;67(795):299–302.
4. Boyacigil S, Ali A, Ardic S, *et al*. Epidural extramedullary hematopoiesis in thalassemia. Aust Radiol. 2002;46(2):180-182
5. Urman M, O'Sullivan R, Nugent R, *et al*. Intracranial extramedullary hematopoiesis: CT and bone marrow scan findings. Clin Nucl Med. 1991;16(6):431-434
6. Chourmouzi D, Pistevou K, Plataniotis G, *et al*. MRI findings of extramedullary haemopoiesis. Eur Radiol. 2001;11:1803–1806.
7. Bizhan A, Mansur H, Vahid R. Visual failure caused by suprasellar extramedullary hematopoiesis in beta thalassemia: case report. Neurosurgery. 1998;42:922–925
8. Haidar S, Ortiz-Neira C, Shroff M, Gilday D, Blaser S. Intracranial involvement in extramedullary hematopoiesis: Case report and review of the literature. Pediatric radiology. 2005 Jun;35:630-4.
9. Clark CA, Worden CP, Thorp BD, Ebert Jr CS, Zanation AM, Senior BA, *et al*. Extramedullary hematopoiesis in the sinonasal cavity: a case report and review of the literature. Allergy & Rhinology. 2020 Apr;11:2152656720918874.

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