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Case Report

Magnetic resonance imaging findings in intracranial extramedullary hematopoiesis in myelofibrosis with myeloid metaplasia: a case report

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ABSTRACT

The formation of blood cells in tissues other than bone marrow is known as extramedullary hematopoiesis. The most common sites of hematopoiesis are liver, spleen and lymph nodes. Extramedullary hematopoiesis can occur in various locations within the body and is characterized by soft tissue masses detected on imaging. Extramedullary hematopoiesis is a known complication of myeloproliferative disorders, which include chronic myelogenous leukemia, polycythemiavera, essential thrombocytosis and mylofibrosis with myeloid metaplasia. An intracranial extramedullary hematopoiesis is an extremely rare occurrence that frequently involves the cranial dura, falx, cerebral parenchyma, optic nerve sheath, and diploic space of skull. MRI remains the modality of choice in investigations for the same. This case report describes the intracranial MR imaging features seen in myelofibrosis with myeloid metaplasia.

Keywords: Extramedullary hematopoiesis, Myeloid metaplasia, Myelofibrosis

INTRODUCTION

Intracranial deposits of extramedullary hematopoiesis are extremely rare, and limited experience with the treatment of these lesions has been reported. Extramedullary hematopoiesis is a rare compensatory process associated with many hematologic disorders and bone marrow dysfunction.

Extramedullary hematopoiesis is described as an ectopic production of erythroid, myeloid and megakaryocytic elements. This ectopic production is believed to be a compensatory mechanism that is subsequent to bone marrow stress and inability to compensate for body's hematologic demands.

Here, we present the MRI findings in a rare case of intracranial extramedullary hematopoiesis in a patientwith myeloid metaplasia.

CASE REPORT

A 38 year old male suffering from myelofibrosis with myeloid metaplasia for 2 years, with liver and spleen involvement. He underwent splenectomy 8 months prior. Now presented with complaints of headache and diplopia since 2 weeks. Due to the severity, he was referred for MRI brain. Cranial MRI showed T1 hypointense (Figure 1), T2 hypointenseparafalcine and paratentorial lobulated soft tissue lesions (Figure 2) which showed gradient blooming on T2* GRE images (Figure 3) and homogenous enhancement after IV injection of gadopentetatedimeglumine (Figure 4). These soft tissue masses showed no perilesional edema. Calvarium was thickened with diploic space widening and showed multiple subcentimetric well circumscribed enhancing lesions (Figure 5). These calvarial lesions were noted to be lytic on CT. These findings were consistent with features of extramedullary hematopoiesis.

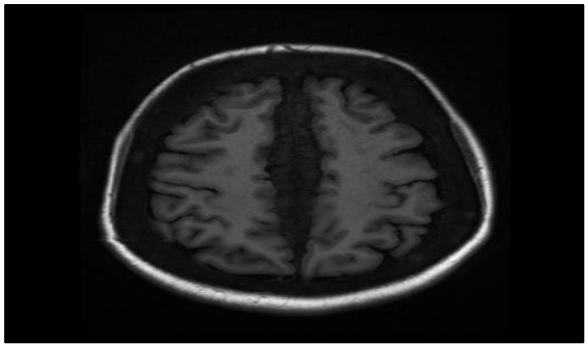

Figure 1: (Axial t1 flair image) parafalcine lobulated hypointense soft tissue masses.

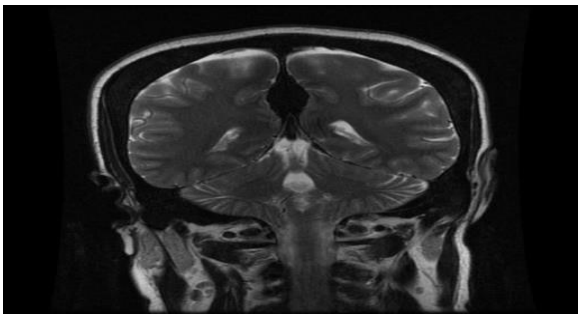

Figure 2: (Coronal T2W image) parafalcine and paratentorialhypointense lesions.

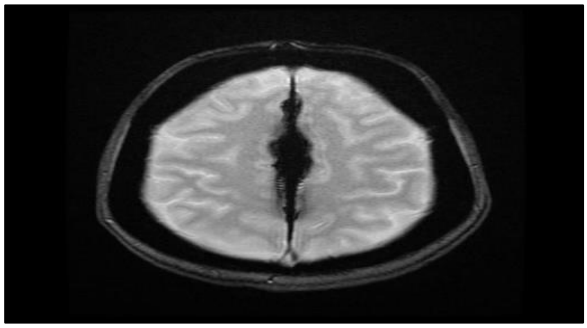

Figure 3: Axial T2* Gradient Recalled echo image showing parafalcine blooming.

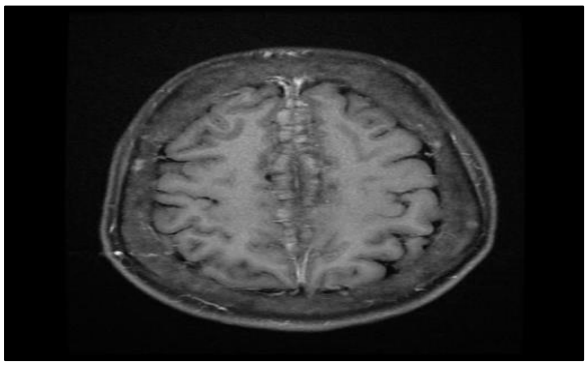

Figure 4: (Axial T1 fatsat contrast image) homogenous contrast enhancement of the lesion with IV gadopentetatedimeglumine administration.

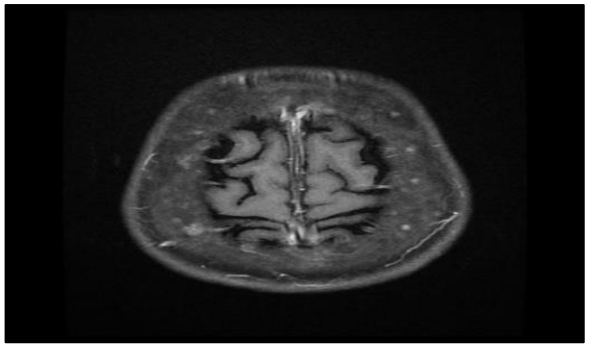

Figure 5: (Axial T1 contrast image) thickened calvarium with widened diploic space and multiple subcentimetric circumscribed enhancing lesions.

DISCUSSION

In extramedullary hematopoiesis, a compensatory response by proliferation of the pleuripotent mesenchymal stem cells outside the bone marrow is attempted by the body to meet the body's demand for anemia.[1] Extramedullary hematopoiesis is commonly seen in hemoglobinopathies such as thalassemia and sickle cell disease and also in marrow replacing disorders such as myelofibrosis. Commonest sites of involvement are liver, spleen and lymph nodes.[2] Reported CNS sites of involvement include the choroid plexus and duramater (over the cerebral convexities, along the falxcerebri, and within the epidural space of the spinal canal.[3] It has also been reported in association with an intracranial hemangioblastoma and meningioma.

The clinical presentation is variable from being asymptomatic to having intracranial hypertension. Meningeal extramedullary haematopoiesis may present as headache, drowsiness, disorientation, obtundation, diplopia, epilepsy and hemiparesis.[4] It has also been described as an incidental finding. Diagnosis is with cross-sectional imaging with or without biopsy.

In this case, the peripheral smear showed classic findings of leukoerythroblastosis and giant platelets. As the spleen is the prime site of hematopoiesis in myeloid metaplasia, its removal might have facilitated extramedullary hematopoiesis at various sites including the intracranial locations to compensate for myelophthisis.Our differential diagnoses included extramedullary hematopoiesis, subdural hematoma, meningioma and meningeal metastasis. The MRI features could be extrapolated to the clinical picture and laboratory findings thus reinforcing the diagnosis of extramedullary hematopoiesis of the falxcerebri and tentorium. MRI remains the modality of choice for showing intermediate to low signal intensity on T1-weighted images and characteristic low signal intensity on T2-weighted images. This hypointensity is contributed to by a magnetic susceptibility effect caused by hemosiderin.Both imaging features and clinical findings

lead to this diagnosis. In this case further examinations were not asked by the oncologist.

CONCLUSION

Extramedullary hematopoiesis should be considered as a close differential diagnosis of intracranial masses in patients with known conditions that may predispose them to extramedullary erythropoiesis. Intracranial lesions with T2 shortening and intense homogenous enhancement may not necessarily prove malignant in origin.

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Ethical approval: Not required

REFERENCES

1. Lee CM, Salzman KL, Blumenthal DT, Gaffney DK. Intracranial extramedullary hematopoiesis : brief review of response to radiation therapy. 2005;152:151-2.
2. Bisset R. Report intracranial with extramedullary correlation hematopoiesis : MR findings pathologic. 1994;1419-20.
3. Palma J, Dominguez PD, Riverol M. Intracranial extramedullary hematopoiesis associated with multiple myeloma. Neurology. 2013;80(17):1620.
4. Karki B, Xu Y, Tamrakar K, Wu Y. Intracranial extramedullary hematopoiesis in. 2012;13(2):240-3.

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