Case Report

Case report of the extramedullary hematopoiesis presented as a hypervascular intracranial mass

Nazila Tayari, Mohamad Hossein Ahrar, Mohamad Saleh Jafarpishe

Department of Radiology, Al-Zahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran

Abstract Thalassemia is a hematologic disorder that causes ineffective hematopoiesis and is related to severe anemia, iron overload, extramedullary hematopoiesis, and hepatomegaly. Hepatomegaly is related to significant extramedullary hematopoiesis. The other sites that are involved in extramedullary hematopoiesis are spleen, lymph nodes, paraspinal regions, kidney, pleura, and intestine, but intracranial involvement is a rare presentation. We discuss about a case with intracranial medullary hematopoiesis in a thalassemic patient.

Key Words: Cerebral ventricle, extra medullary hematopoiesis, paraventricle

Address for correspondence:
Dr. Nazila Tayari, Department of Radiology, Al-Zahra Hospital, Isfahan University of Medical Sciences, Isfahan, Iran. E-mail: nazila524@yahoo.com
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INTRODUCTION

Extramedullary hematopoiesis (EMH) is a rare compensatory process associated with many hematologic disorders and bone marrow dysfunction with anemia.[1] It has been reported to occur in approximately 15% of cases of thalassemia, and it also occurs in myelofibrosis and in other anemic conditions.[2] EMH arises from multipotential cells in any tissue, such as the liver, spleen, lymph nodes, paraspinal regions, kidney, pleura and intestine, but intracranial hematopoiesis is a rare condition.[3]

We present a case with intracranial medullary hematopoiesis.

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CASE REPORT

A 34-year-old man presented with progressive headache and epileptic condition for the last 10 months. He was known to have intermediate B-thalassemia. He experienced severe nausea, vomiting, and growing loss of vision. He was admitted in the neurosurgery ward of Alzahra Hospital, Isfahan, Iran.

On the day of admission, he was anemic and received transfusion therapy for the first time, after he became a candidate for surgery. On lateral skull X-ray showed thickening of diploic space [Figures 1]. Non-contrast and contrast-enhanced brain computed tomography scan (CT) in bony and soft tissue window revealed external and internal skull Tables and diploic space thickening and one hyperdense enhancible left paraventricular mass with peripheral edema [Figures 2 - 4]. On axial T1-W and T2-W brain magnetic resonance imaging (MRI), a hyper- and hypointense paraventricular mass with peripheral edema was identifi ed [Figures 5 and 6]. After injection of gadolinium, sagittal T1-W images showed intense and homogeneous enhancement [Figures 7-8]. A surgical biopsy showed infi ltration by megakaryocytes, erythroblasts, and myeloid cells corresponding to EMH..

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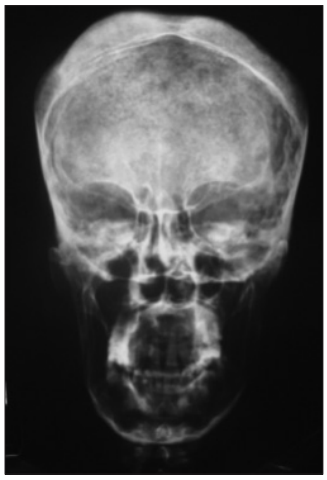

Figure 1: AP skull X-ray showing thickening of diploic space

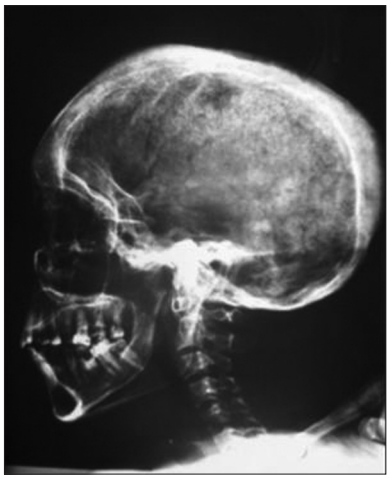

Figure 2: Lateral skull X-ray showing thickening of diploic space

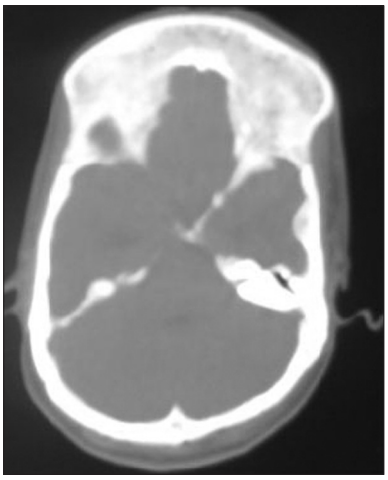

Figure 3: Brain CT scan (bony window) showing external and internal skull Tables and diploic space thickening

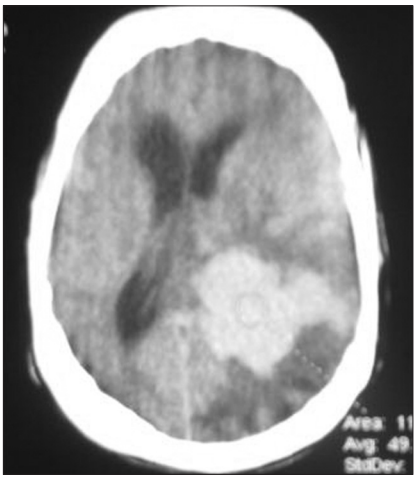

Figure 4: Brain CT scan (soft tissue window) showing a hyperdense paraventricular mass with peripheral edema

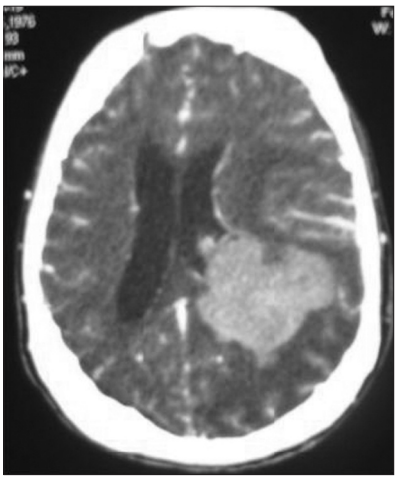

Figure 5: Contrast-enhanced brain CT scan showing an intense enhancible paraventricular mass

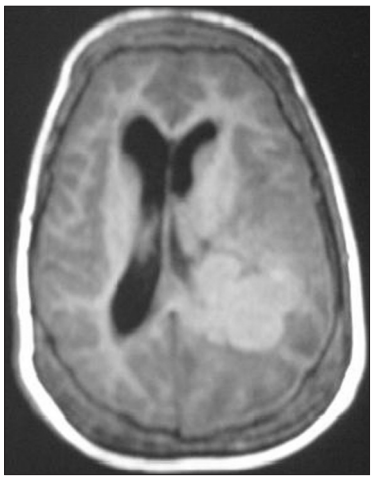

Figure 6: Axial T1-W brain MRI revealing a hyperintense paraventricular mass

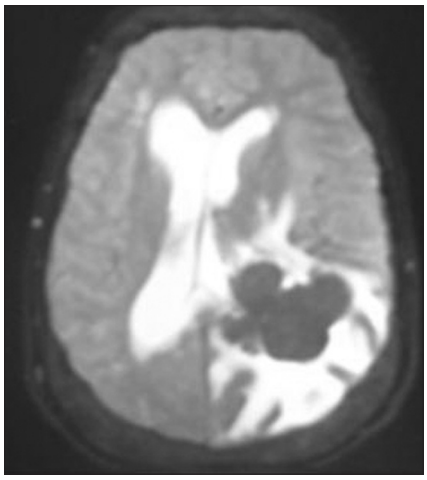

Figure 7: Axial T2-W brain MRI showing a hypointense paraventricular mass with hyperintense peripheral edema

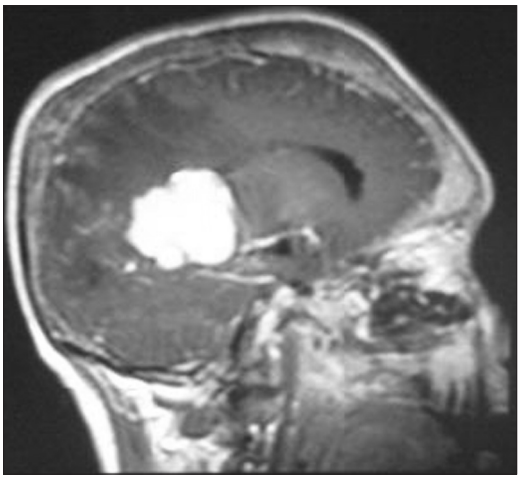

Figure 8: Sagittal contrast-enhanced T1-W brain MRI revealing an intense homogeneous enhancible paraventricular mass

DISCUSSION

Hematopoiesis is the formation and maturation of blood elements. When the primary sites of hematopoiesis in the adult fail, as in myelofibrosis and hemoglobinopathies (especially thalassemia and sickle cell disease), various extramedullary sites take on the role of blood formation.[4]

Several clinical problems can lead to EMH. These include, among others, hemolytic anemia, primary and secondary myelofibrosis, leukemia, lymphoma and bone metastasis, and myelodysplastic syndromes, characterized by abnormal morphology and inadequate production of blood cells,[1,5] while the uncommon intracranial EMH occurs without predisposing bone marrow disorders. Isolated intracranial EMH occurs most often in association with hemangioblastomas; it has also been observed in two intracranial lipomas, a meningioma, a few subdural hematomas, an encephalocele, a tumor-like mass of papillary endothelial hyperplasia, and a pilocytic astrocytoma.[6]

EMH most often involves the spleen, the liver, and paraspinal regions of the thorax. However, in addition to these common sites of EMH, the process can involve virtually any organ or tissue. Reported sites include abdominal viscera, pleura, lymph nodes, adrenal glands, breast, thymus, kidneys, gastrointestinal tract, and intracranial structures.[4,7-9] There are two main groups in EMH. The first group shows paraosseous foci that may result from herniation of medullary tissue from the underlying bone and is seen in hemolytic disorders such as thalassemia and sickle cell anemia, where the marrow has tremendous activity. The second group shows extraosseous soft tissue foci, which may arise from multipotential stem cells. This happens when the marrow activity is ineffective, as in idiopathic myelofibrosis or, rarely, with toxic or tumoral marrow destruction.[10]

Intracranial EMH is extremely rare.[11] Most patients do not have signs and symptoms related directly to the disorder. Most foci of EMH are noted as incidental findings on imaging studies or postmortem examination, although occasionally EMH in the intracranial or intraspinal epidural space can lead to serious neurogenic complications (increased intracranial pressure, hemiplegia, altered levels of consciousness, or visual disturbances, including subdural hemorrhage, delirium, increased intracranial pressure, papilledema, coma, motor and sensory impairment, and limb paralysis due to direct mass effect upon adjacent structures).[5,8,10,11]

The most frequently reported causes of intracranial involvement by EMH are thalassemia (50%) and myelofibrosis (31%).[1]

Reported CNS sites of involvement include the choroid plexus and dura mater (over the cerebral convexities, along the falx cerebri, and within the epidural space of the spinal canal), optic nerve sheath, and the diploic space of the skull.[8,11] Intraparenchmal mass is a rare presentation of EMH.

Diagnosis of EMH is based on the clinical circumstances, laboratory data, and the use of different diagnostic imaging modalities.[1] In cases with intracranial EMH, the CT appearance is characterized by the heterogeneous lobulated soft tissue density mass.[7] Lund and Aldridge described EMH as multiple extra-axial masses over the brain surface with attenuation values equal to or higher than those of gray matter.[1]

MRI is the diagnostic investigation of choice. Intracranial EMH appears as unique or multiple iso- or hyperintense extra-axial masses appended to the meninges, with homogeneous enhancement after contrast administration. These masses are usually lobular, well-circumscribed masses of intermediate signal intensity on T1-weighted images and low signal intensity on T2-weighted images. These masses may show significant enhancement after gadolinium administration.[1,11,12] MRI demonstrates a high signal intensity rim representing fatty tissue on both T1- and T2-weighted images.[5]

The multiple intracranial extra-axial EMH can mimic multiple meningiomas, lymphoma, myeloma, leukemia and neuroblastoma, and other metastatic malignant diseases. In addition, the differential diagnoses include epidural hematoma, abscess, chloroma, granulomatous diseases like tuberculosis and sarcoidosis, and pachymeningeal thickening related to rheumatoid disease.[1,12]

This case suggests that EH must be taken into consideration when masses with characteristic radiologic appearance are identified somewhere in patients with various blood dyscrasias.[7]

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