Extramedullary Hematopoiesis: Imaging and Clinical Implications

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INTRODUCTION

Extramedullary Hematopoiesis is the production of blood cells outside of the normal location of the bone marrow, occurring secondary to inadequate production of blood cells. EMH may be due to inefficient blood cell production or a compromise in the quality of blood cells. In the fetus, the primary sites of hematopoiesis are the yolk sac, liver, spleen, and bone marrow. After birth, hematopoiesis should occur only in the bone marrow and any extramedullary location is considered abnormal. Causes of EMH are congenital or acquired hemolytic diseases, ineffective erythropoiesis, loss of control of stem cell differentiation, or nonmyeloid neoplastic diseases. Conditions causing EMH include thalassemia, sickle cell anemia, myelofibrosis, leukemia, and lymphoma.

METHODS

• Four cases from Tulane University Hospital were reviewed. All patients were found to have EMH findings projected on the head and neck, spine, pelvis, and musculoskeletal system.
• These cases were evaluated using Magnetic Resonance Imaging (MRI) and Computed Tomography (CT).
• Recent literature and guidelines were reviewed, and a search for EMH was performed in PubMed. From the literature, we were able to identify cases of EMH and compare them to our case series. Localized anatomic landmarks were identified and pertinent findings are presented with each case.

PATHOPHYSIOLOGY

EMH occurs when bone marrow is no longer able to support normal hematopoiesis. EMH can occur under conditions of local production of hematopoietic factors that maintain and induce differentiation of the stem and progenitor cells, when there are supporting cells, and when there is accommodation of hematopoietic progenitors. The cascade begins with displacement and mobilization of stem and progenitor cells. Consequently, hematopoietic stem and progenitor cells occupy other locations as alternative sites of hematopoiesis.

DISCUSSION

EMH occurs in the liver and spleen for 95% of cases, with the remaining 5% of cases occurring in almost every tissue in the human body, including the adrenal gland, thymus, kidney, pleura, pulmonary interstitium, breast, skin, and gastrointestinal tract. Very rarely, EMH can develop in the CNS, head, and neck, and spine. After hepatosplenomegaly, bilateral heterogeneous paravertebral masses are the next most common manifestation. Symptoms for the majority of patients with non-hepatosplenic EMH (63%) will be site-specific. Patients also often present with generalized symptoms such as fatigue (15%) or are asymptomatic (22%). EMH has high risk for hemorrhage complications and paraspinal lesions may affect the spinal cord and peripheral nerves, causing symptoms such as weakness and radiculopathy.

TREATMENT

The treatment approach for EMH depends on a number of factors, including the size of the mass, severity of symptoms, the clinical condition of the patient, and previous treatment methods. Excisional biopsy, radiation therapy, and frequent blood transfusions to limit hematopoietic stimuli are some treatment options. Therapy may be required in cases such as EMH manifestations in the spinal canal causing spinal cord compression, and asymptomatic cases may require no therapy.

CONCLUSION

EMH is a non-neoplastic formation of blood or blood cells outside the bone marrow found in patients suffering from various hematologic disorders. EMH usually presents as hematopoietic masses that may occur in almost all body sites, including rare manifestations in the CNS, head and neck, and spine. When imaging features suggestive of EMH are identified, ECM should be strongly considered. Knowledge of these atypical locations correlated with clinical history is needed to reach a diagnosis of EMH and to exclude other pathology. Treatment may be required if the case is symptomatic, such as when manifestation occurs within the spinal canal causing cord compression. EMH in these locations should be closely monitored with follow-up imaging.

REFERENCE


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