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Case Report

Multifocal extramedullary hematopoiesis in a 15-year-old girl with beta thalassemia: A case report ^{☆,☆☆}

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ABSTRACT

Extramedullary hematopoiesis is a rare disorder in which hematopoietic cells proliferate in tissues other than the bone marrow as a result of a range of hematologic illnesses. Our case is unique in that it covers a number of extramedullary hematopoiesis sites in a 15-year-old girl, some of which are uncommon.

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Introduction

When the normal process of red blood cell generation in the bone marrow is disturbed, extramedullary hematopoiesis is a physiological phenomenon in which the body seeks to maintain functional erythropoiesis [1]. These so-called relay organs participate in erythropoiesis during fetal life and then stop after delivery, while keeping this potential in the event of inefficient erythropoiesis. The most common causes include myelofibrosis, disseminated malignancy, leukemia, and hemoglobinopathies [2].

Case report

A 15-year-old girl with a medical history of B thalassaemia, which she has been followed for since childhood. Since she was five years old, she has been undergoing PRBC (pure red blood cell) transfusions every 6 months to keep her pre-transfusion hemoglobin between 8 and 9 g/dL. She was sent to our hospital emergency room with significant acute back pain that was lateralized to the left and accompanied by some abdominal discomfort. There were no signs of lymphadenopathy, pallor, or icterus. Her motor and sensory func-

Abbreviations: EMH, extramedullary hematopoiesis; PRBC, pure red blood cell; CT, computed tomography.

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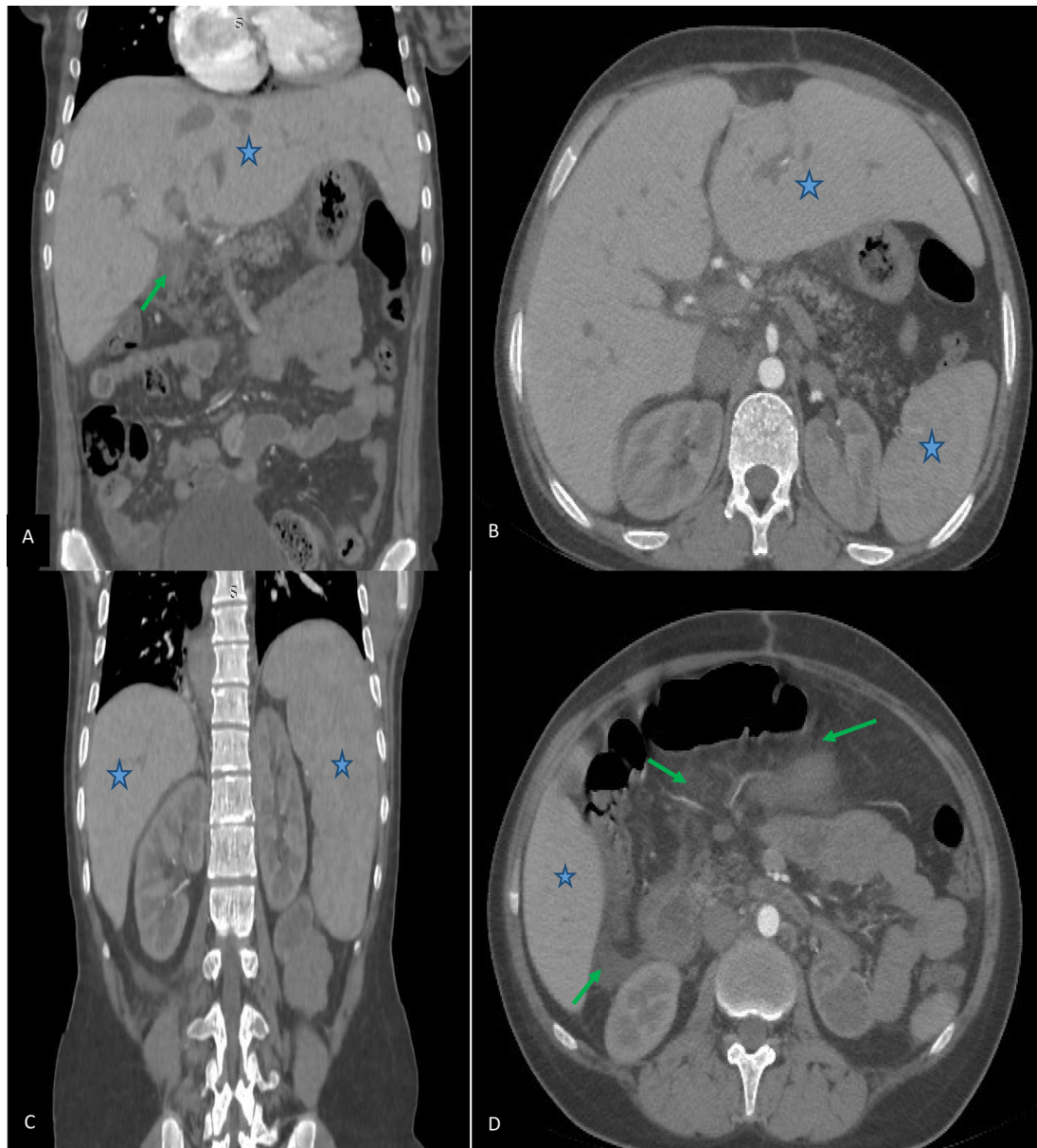


Fig. 1 – Coronal (A, C) and axial (B, D) CT images demonstrating a homogeneous hepatosplenomegaly (stars) with focal peritoneal nodularity and effusion (yellow arrows) (Color version of figure is available online).

tions were also in good working order. During an abdominal examination, a substantial splenomegaly measuring 6 cm below the left costal boundary was detected. Her treating physician suspected a splenic infarction based on the clinical symptoms. As a result, an abdominal angioscan was performed, which revealed homogenous hepatosplenomegaly with focal peritoneal nodularity and effusion (Fig. 1). The computed tomography (CT) scan revealed bilateral mediastinal paraspinal soft tissue masses with smooth surfaces on the thoracic level, which were moderately enhancing following injection (Fig. 2). A posterior epidural enhancing soft-tissue bulk in direct touch with the spinal cord was also seen (Fig. 3). The patient was referred to the hematology department and received 2 transfusions over the course of 6 months, dur-

ing which time she showed no signs of motor or sensory impairment.

Discussion

Extramedullary hematopoiesis is known to be caused by hemoglobinopathies such as thalassemia. The first case of spinal cord compression caused by EMH was documented in 1954. EMH most commonly affects the reticuloendothelial system (liver, spleen, and lymph nodes) and paraspinal parts of the thorax [3]. There has been evidence of diffuse microscopic infiltration of the liver and spleen, which can be

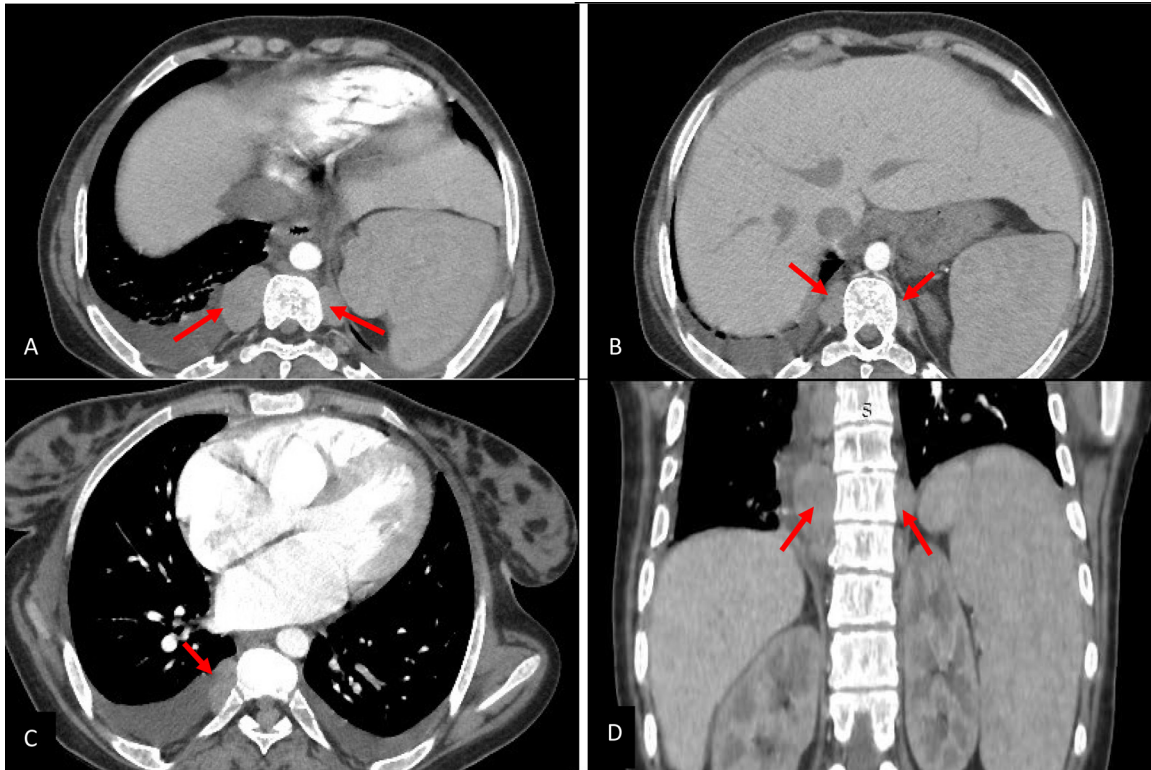


Fig. 2 – axial (A-C) and coronal (D) enhanced CT images of bilateral mediastinal paraspinal soft tissue masses with smooth surface (red arrows) (Color version of figure is available online).

seen on radiographs as hepatosplenomegaly or isolated mass lesions [3,4]. Pleura, lungs, gastrointestinal tract, breast, skin, brain, kidneys, and adrenal glands are all impacted in a less common way. The central nervous system, the head and neck (including the spine and suprasellar region), and the central nervous system can all be affected [3]. In rare cases, it might result in spinal cord compression, pleural effusion, severe bleeding, and respiratory failure [5]. Our case illustrates a multifocal involvement of EMH with paraspinal mediastinal, hepatic, splenic, peritoneal and spinal involvement.

EMH is usually asymptomatic and symptoms are usually caused by the mass effect [6]. Our patient was experiencing minor orienting symptoms, such as back pain and abdomen tenderness.

The CT scan reveals tissue masses with lobulated contours that are well-limited, mildly enhanced after injection, and contain some fatty portions [2]. Calcifications are quite uncommon. The EMH shows as a solid mass with interior vascularity on ultrasonography [2]. In our case, the masses were purely tissue-based, with no fat or calcium.

Magnetic resonance imaging is the preferred examination. It investigates epidural extension and its impact on the medullary cord. The masses have a variable signal based on the proportion of active red and inactive yellow marrow, thus if the area of involvement is active, they have an intermediate signal on T1 with a hyper signal in T2, which increases after injection. If it is inactive, they present a T1 and T2 hypo signal with no enhancement following injection. The fatty fraction

exhibits a T1 and T2 hyper signal, which is suppressed on the saturated sequences.

EMH's paraspinal thoracic masses are typically bilateral and have a smooth surface. They are much more common in the thorax than in the abdomen or pelvis [2].

Intraspinal EMH has only been documented in a few cases, and it is usually asymptomatic until there is spinal cord compression [7]. The preferred evaluation is an magnetic resonance imaging, which may reveal the size of the tumor as well as meningeal and epidural involvement [7]. On CT, active lesions have a soft tissue-like density. Inactive lesions, on the other hand, are either hyperdense due to iron deposits or hypodense due to fat deposits [7]. Compression of the spinal cord must be managed as soon as possible. Surgery, radiation, and red blood cell transfusion are all alternatives for treatment [7]. The existence of a peripheral fatty rim in some intraspinal masses has been documented, and it helps inside the differentiation of EMH from metastatic masses [8]. It isn't always present, though [8].

The liver and spleen are the most common abdominal sites of EMH. Usually with diffuse microscopic involvement manifesting as visceromegaly. Infrequently, focal masses may be seen. Periportal and peribiliary hepatic involvement may also occur [2].

Ascites or peritoneal implants are examples of peritoneal involvement [9]. The parietal or visceral peritoneum, as well as the larger omentum, may acquire these implants [9]. Intestinal stenosis or blockage may result from implants in

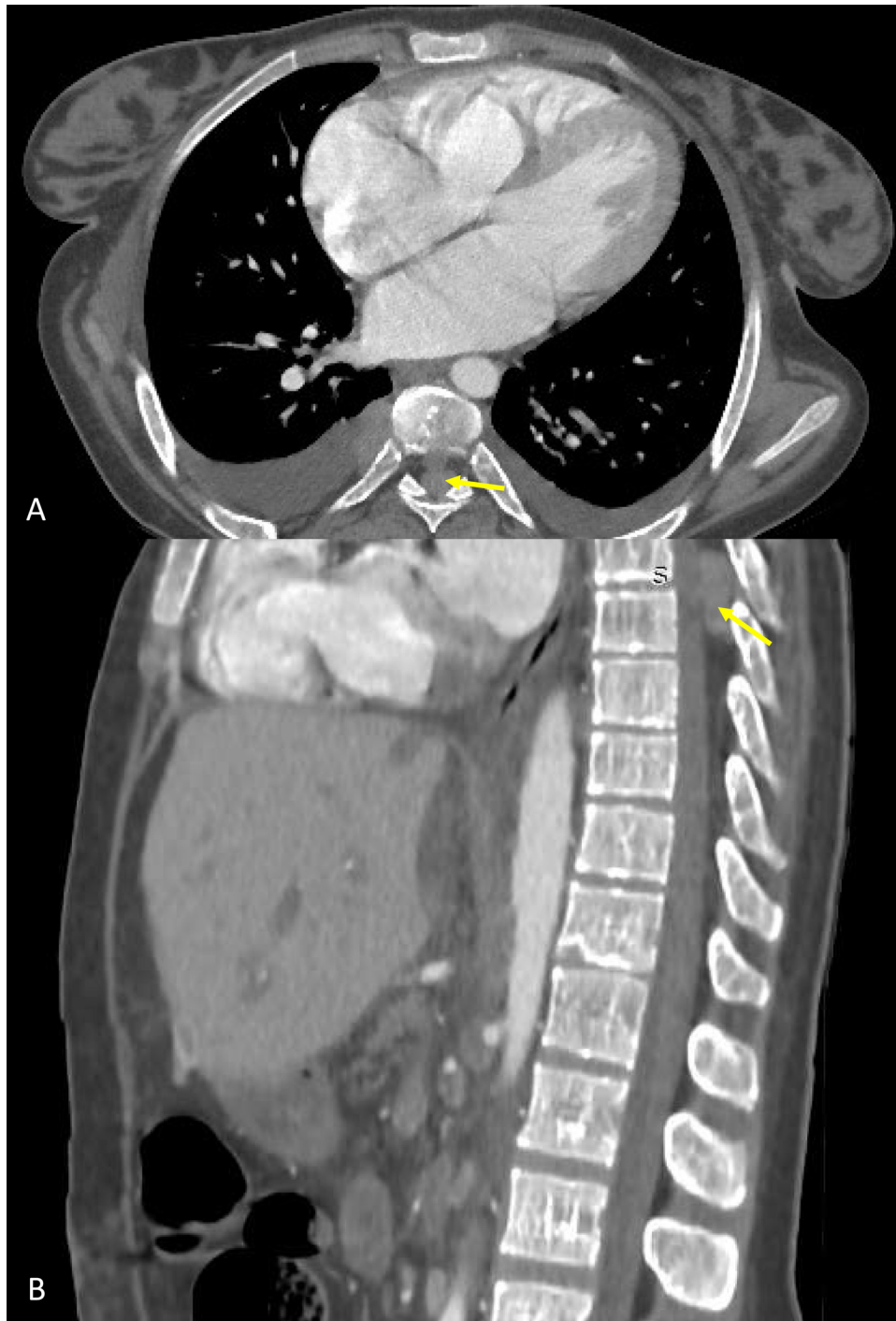


Fig. 3 – axial (A) and sagittal (B) CT images showing posterior epidural enhancing soft-tissue masse (yellow arrows) in immediate contact with the spinal cord (Color version of figure is available online).

the visceral peritoneum [9]. Peritoneal tuberculosis and peritoneal carcinomatosis are the most common differential diagnoses [9].

The evolution is generally towards regression of the masses with correction of the hematopoietic disorders. It is therefore important to be able to suggest the diagnosis in the presence of hematopoietic dysfunction and typical imaging findings.

Conclusion

In conclusion, our case was a rare case of extramedullary hematopoiesis with multifocal involvement. Knowledge of the many locations and forms of EMH is critical for thinking about the diagnosis and guiding management.

Authors' contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Patient consent

Written informed consent for publication was obtained from patient.

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