Extramedullary hematopoiesis (EMH) has been reported in various organs. However, its manifestation as a focal tumor-like splenic lesion is rare. Here, we report a case of EMH in an 85-year-old female presenting as a well-defined intrasplenic mass of 12 cm on sonography and computed tomography. Splenectomy was performed under the impression of splenic tumor, and intrasplenic focal EMH was diagnosed histopathologically.

Key words: Computed tomography; Extramedullary hematopoiesis; Spleen; Sonography

CASE REPORT

An 85-year-old woman with the complaint of severe lower back pain was admitted to our hospital under the impression of acute sciatica, chronic anemia and impaired renal function. Laboratory findings showed red blood cell count $2.74 \times 10^6 /\text{ul}$, Hgb 5.8g/dl, MCV 71.6fL, platelet count 82000/ul, BUN 44mg/dl, and Cr 1.8mg/dl. Physical examination revealed pale conjunctiva, tenderness at the back region and mild limitation of movement of the left lower extremity. Patient had past history of thyroid goiter, cataract and hypertension without regular treatment. X-ray findings of lumbar spine and left hip showed only degenerative change.

On abdominal sonogram survey, splenomegaly and a heterogeneous echogenic splenic mass (Fig. 1) was identified incidentally. CT scan was then performed for further investigation. On nonenhanced CT, splenomegaly was noted with an intrasplenic ill-defined hypodense mass lesion (Fig.2A) measured approximately 12 cm in diameter. A small calcification
was identified at the margin of the lesion. Enhanced CT (Fig. 2B) showed minimal finger-like heterogeneous marginal enhancement.

Since the possibility of a malignant tumor growth could not be ruled out, splenectomy was performed. The intrasplenic tumor measured 12 × 7 × 7 cm in size and histological analysis (Fig. 3) revealed a picture of EMH with clustered megakaryocytes, fibrosis and occasional calcifications. Multiple infarcts in large area were also observed, and there was no evidence of malignancy.

**DISCUSSION**

EMH arises from pleuripotential stem cells distributed throughout the body. It is most common in patients with congenital hemolytic anemia, such as thalassemia, sickle cell anemia and hereditary spherocytosis as a response to ineffective red blood cell formation. It is also seen in patients with acquired marrow replacement disorders such as leukemia, lymphoma, carcinoma, and myelofibrosis [1]. The disease usually affects the spleen and liver in diffuse infiltration pattern microscopically. There may be focal mass-like involvement of these organs, however [2-7].

In our case, the etiology of the patient’s focal splenic EMH is not clear. However she was anemic with low MCV only. Further investigations such as bone marrow biopsy, hemoglobin electrophoresis and osmotic fragility tests were not available because of the patient’s rejection. Thalassemia group may be the most possible cause in consideration.

The sonographic, CT and magnetic resonance (MR) features of focal splenic EMH have been reported in several cases [3, 5, 6, 7]. For the sonographic finding, hyperechoic mass without or with central necrosis has been described, in contrast to our case of mass with inhomogeneous echogenicity.

The CT appearances of focal splenic EMH are typically well-defined hypodense mass with internal mosaic attenuations in the nonenhanced and enhanced CT scans. However, minimal finger-like marginal enhancement with a tiny peripheral calcification were noted in our case.

The reported MR findings of EMH mass at various sites are mostly intermediate signal intensity (SI) on T1-weighted images (T1WI) and high SI on
T2-weighted images (T2WI) with slight enhancement after infusion of gadolinium [9,10]. Low SI hematopoietic mass on T2WI MR images has also been reported [11]. Gabata et al [6] reported the only MR findings of focal splenic EMH showing low SI on T1WI, high SI on T2WI, and progressive enhancement on dynamic scans. These SI and enhancement patterns may suggest an active hematopoietic process, while older inactive lesions may show low SI on both T1- and T2WI due to iron deposition, or high SI on both sequences due to fatty infiltration [12].

There are a long list of differential diagnosis for a focal splenic mass, including hemangioma, hamartoma, lymphangioma, lymphoma, metastasis, angiosarcoma, atypical infarction and infection. Although the imaging appearance of splenic EMH is not specific, it should always be suspected in the setting of a known hematological disorder clinically.

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REFERENCES


Figure 3. Microscopically, the resected splenic tumor showed a picture of extramedullary hematopoiesis with marked red pulp infiltration by myeloid cells (arrows) and megakaryocytes (arrowhead). (HE stain, X400)
侷限性腫瘤樣之脾臟骨髓外造血：病例報告

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骨髓外造血可以發生在許多器官中，然而以侷限性腫瘤表現在脾臟中則相當罕見。在此，我們報告一脾臟骨髓外造血病例，患者為一名八十五歲女性，超音波及電腦斷層影像檢查上發現脾臟中出現一個十二公分大小空腔性病灶，由於無法排除腫瘤的可能性，患者接受了脾臟切除手術，並證實病灶為脾臟中侷限性骨髓外造血。

關鍵詞：電腦斷層、骨髓外造血、脾臟、超音波