CLINICAL IMAGE

Contrast-enhanced ultrasonographic features of primary splenic angiosarcoma

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A 32-year-old man presented with dull pain and discomfort in the left upper abdomen lasting for over 1 month. Physical examination revealed an enlarged, hard, uneven, nodular spleen. Laboratory tests showed anemia (red blood cell count, 3.93×10^{12}/l, hemoglobin, 112 g/l, white blood cell count, 11.13×10^{9}/l, platelet count, 91×10^{9}/l), while the levels of tumor markers for digestive tract cancer were within the reference range. The patient underwent several imaging tests, including ultrasound (US), contrast-enhanced ultrasonography (CEUS), contrast-enhanced computed tomography, and positron emission tomography /computed tomography (PET/CT), which revealed an enlarged spleen with multiple masses (FIGURE 1A–1F). Laparoscopic splenectomy was performed. Grossly, the spleen was enlarged, with numerous hemorrhagic or necrotic nodules. Histological examination revealed that the tumor cells were positive for CD31, CD34, and coagulation factor VIII, and the expression of Ki-67 was over 40%, indicating splenic angiosarcoma (FIGURE 1G–1H).

Primary splenic angiosarcoma has a higher incidence in men. Patients of any age may be afflicted, and the age distribution ranges from 14 months to 89 years. Although there is no specific clinical presentation of this disease, it is prone to distant metastases. The prognosis is poor; death was reported from 17 days to 29 months after diagnosis, regardless of treatment.[1] The worst prognosis is associated with spontaneous or traumatic rupture of the spleen, which could cause sudden death from hypovolemic shock and disseminated intravascular coagulopathy.[2] To date, more than a year after discharge, our patient still has not come to the hospital for a follow-up visit, so his current status remains unknown.

There are many reports on the features of splenic angiosarcoma on traditional US, CT, or PET/CT; however, CEUS findings of this disease are rare. The dynamic real-time imaging characteristics of CEUS dramatically improve the sensitivity and specificity of diagnostic ultrasonography; its ability to diagnose the most frequent focal liver lesions with high accuracy is similar to that of magnetic resonance imaging.[3] In contrast, the available data on the usefulness of this modality in the differential diagnosis of focal splenic lesions are sparse, possibly owing to the low frequency of occurrence.[4] Although CEUS shows an equipollent diagnostic performance in terms of differentiating splenic lesions compared with CT and magnetic resonance imaging, ultrasonic contrast agents are associated with a very low rate of anaphylactoid reactions and do not excrete through the kidneys. Therefore, this method has an excellent safety profile in both adult and pediatric populations, particularly in multimorbid patients as well as those with renal impairment or thyroid hormone disturbances.[5] According to Stang et al,[3] CEUS is clinically recommended for patients with no history of cancer, in whom isoechoic or hypoechoic splenic lesions are found by US. Typically, benign lesions initially showed isoenhancement or hyperenhancement followed by slow and incomplete washout, while malignant tumors appeared early as hypoenhancing defects with rapid and complete washout.[3] The predictive ability of CEUS was higher for lesions with isoechoic or hypoechoic appearance than for those with hyperechoic appearance on US.[3] In our patient, US revealed an enlarged spleen with multiple, ill-defined, heterogeneous, hypoechoic masses, whereas CEUS showed uneven and slight enhancement in the arterial phase, then rapidly cleared and showed low echogenicity in the parenchymal phase, suggesting that the lesions are malignant. Therefore, CEUS may be a useful imaging modality in the evaluation of primary splenic angiosarcoma, but more experimental data are needed to confirm its utility.

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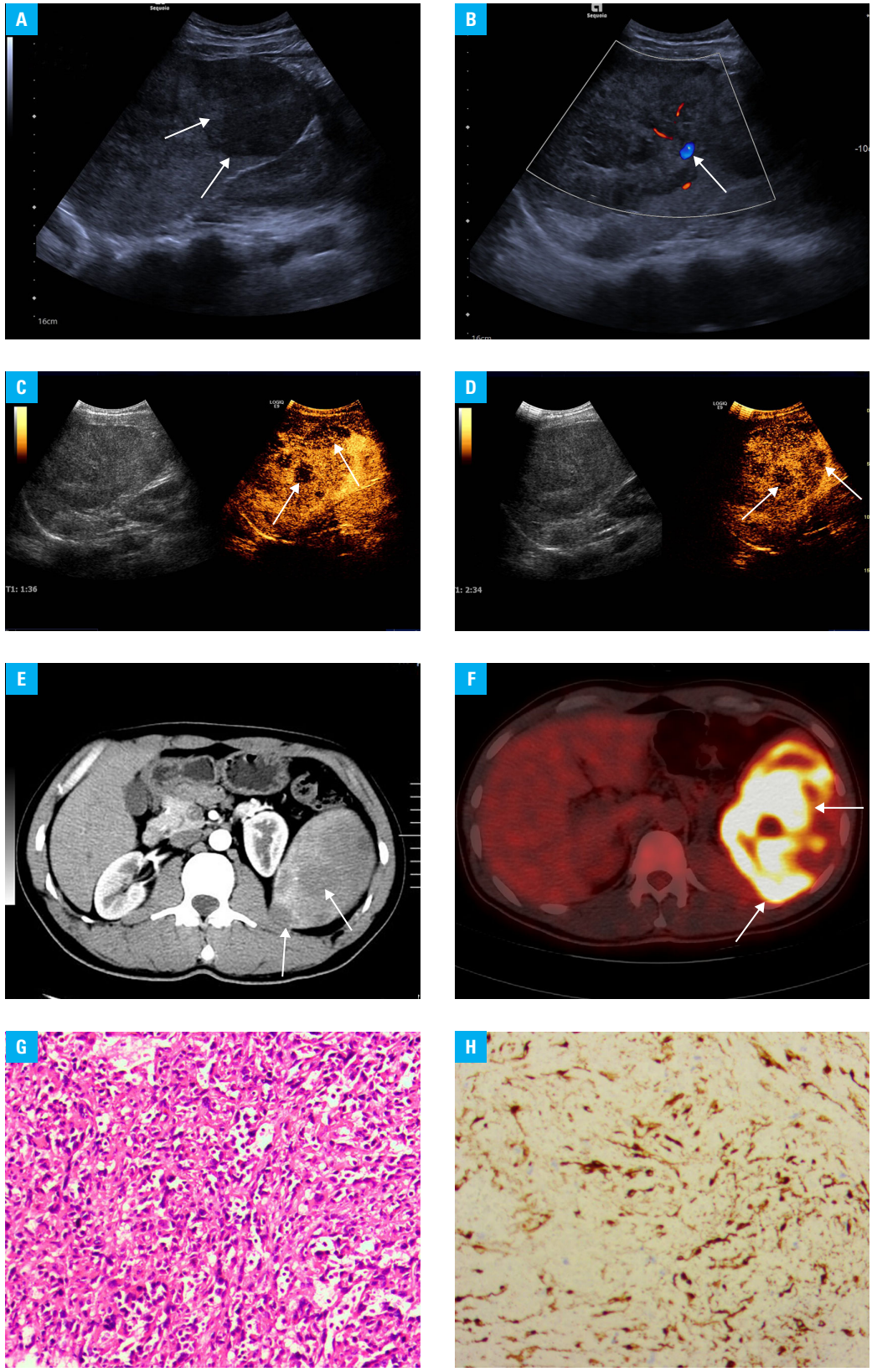

FIGURE 1 **A** – ultrasound image showing an enlarged spleen with multiple, ill-defined, heterogeneous, hypoechoic masses, some of which containing internal echoes; **B** – color Doppler ultrasound image showing increased blood flow in the nodules; **C**, **D** – contrast-enhanced ultrasonography showing uneven and slight enhancement in the arterial phase, not apparent in the central anechoic region (**C**) as well as rapid clearance and low echogenicity of the masses in the parenchymal phase (**D**); **E** – contrast-enhanced computed tomography imaging showing multiple complex low--attenuation lesions in the enlarged spleen with slight heterogeneous enhancement; patchy unenhanced areas are also visible. **F** – fused image showing heterogeneously increased fluorodeoxyglucose uptake with diffuse distribution in the spleen (maximum standardized uptake value, 9.13); arrows on panels **A–F** indicate splenic nodues. **G** – microscopic image of the lesion (hematoxylin and eosin staining, magnification × 200) showing disorganized spindle cells, atypia, nuclear fission, and necrosis; **H** – tumor cells positive for CD34 (immunohistochemical staining, magnification × 200)

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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