Letter to the Editor

Multimodality imaging findings of splenic littoral cell angioma: a case description and analysis of literature

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Introduction

Littoral cell angioma (LCA) is a rare primary splenic tumor originating from littoral cells lining the splenic red pulp sinuses (1). Here, we present a case of pathologically confirmed proven LCA in a 19-year-old asymptomatic male patient, who underwent laparoscopic surgery and recovered. To the best of our knowledge, this case report contains the most comprehensive imaging information reported to date. Multimodality imaging can provide increased information from different perspectives to identify LCA in clinical practice. The merit of this case report lies in the rarity of the condition and the richness of the imaging data presented.

Case presentation

LCA is a rare primary vascular neoplasm of the spleen, which is thought to originate from littoral cells lining the splenic red pulp, and it has features of both the endothelial and histiocytic immunophenotypes. The exact pathogenesis is still unclear, but it has been suggested that the pathogenesis is associated with malignancy, immune-dysregulation or chronic inflammation (1).

Here, we report a rare case of LCA with accessory spleen in a 19-year-old male, whose hemoglobin, number of erythrocytes, hematocrit, and lymphocyte ratio had declined slightly. All the tumor markers examined were negative. Abdominal ultrasonography showed a hypoechoic nodule (size: 4.3 cm ×3.7 cm) in the spleen, which had a rich color flow signal. There was also an isoechoic nodule in the splenic hilus (*Figure 1A*). The nodule in the spleen appeared homogeneously blue (soft) on the shear wave elastography (SWE) map. The maximum Young's modulus of the splenic nodule was 22.6 kPa and the value of its surrounding spleen was 23.6 kPa. The stiffness value of the splenic lesion revealed a soft mass using SWE (*Figure 1B*). Subsequently, the patient received contrast-enhanced ultrasound (CEUS). First, the splenic nodule displayed rapid hyper-enhancement compared to the surrounding spleen parenchyma in the early phase 10 seconds after injection (*Figure 1C*). Then, the central part of the splenic nodule appeared slightly washed-out during the parenchymal phase (after 180 seconds) (*Figure 1D*). The nodules in the splenic hilus showed homogeneous iso-enhancement. According to the CEUS pattern, the sonographers investigating the case had diverse opinions regarding the diagnosis: some considered that the tumor was malignant, whereas others considered it benign. Regarding the nodules in the splenic hilus, the diagnosis was accessory spleen. On magnetic resonance imaging (MRI) scans, the

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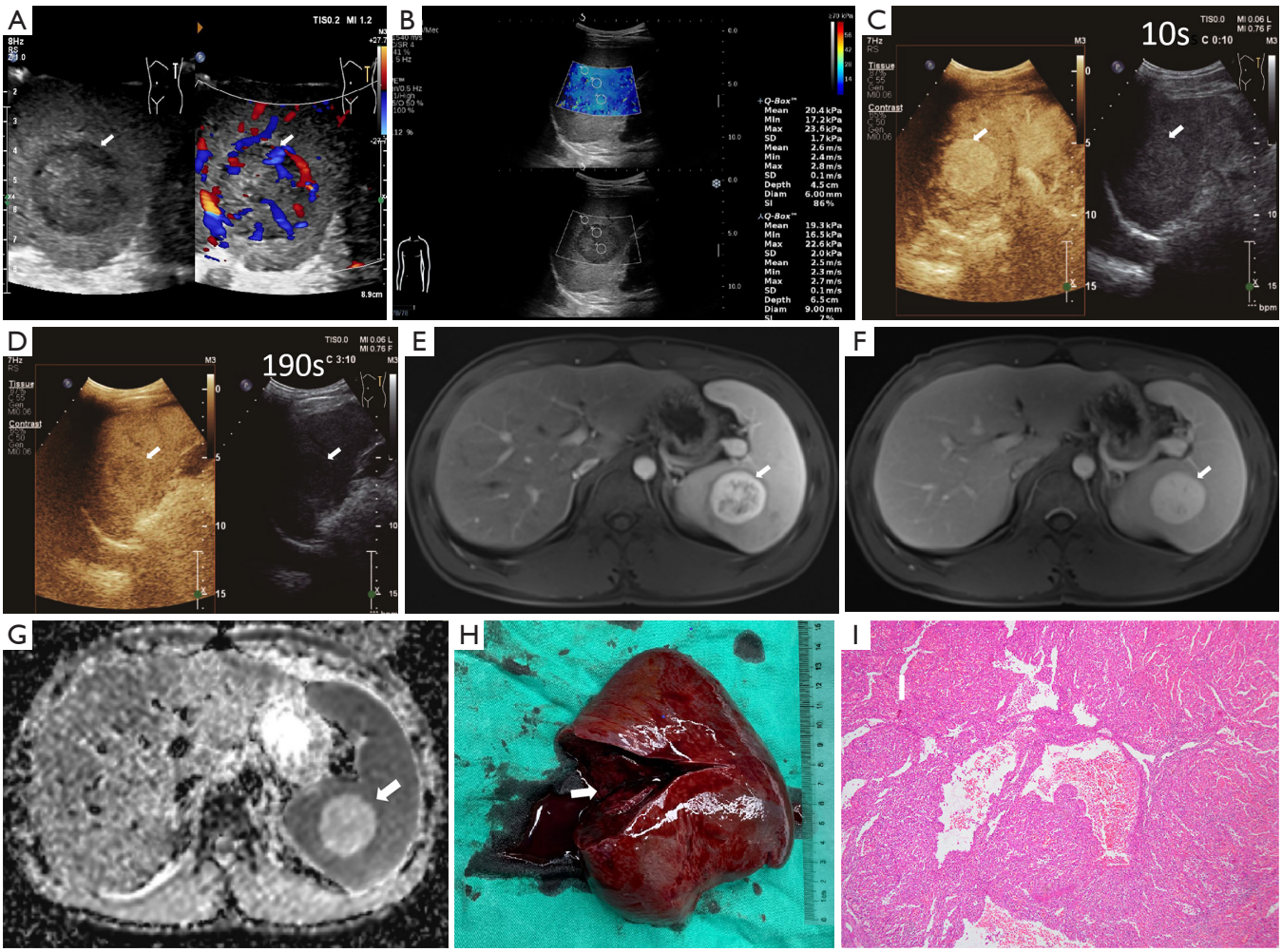

Figure 1 Multimodality imaging findings of splenic littoral cell angioma in this study. (A) The ultrasound images showed a hypoechoic nodule (arrows) in spleen, with rich color flow signal. (B) SWE reveals the splenic nodule was soft. (C,D) The CEUS demonstrated the different time points (10 s, 190 s) of blood perfusion. The arrows point to splenic lesion. (E-G) MRI showed the enhanced pattern of the lesion (arrows) in all phases (E) arterial phase; (F) portal phase; (G) apparent diffusion coefficient. (H) Gross pathological specimen shows an enlarged spleen containing a solitary nodular lesion (arrow); (I) The lesion consisted of dilated anastomozing vascular channels, with multiple papillary projections and cyst-like space (HE, ×40). SWE, shear wave elastography; CEUS, contrast-enhanced ultrasound; MRI, magnetic resonance imaging; HE, hematoxylin-eosin staining.

mass exhibited relative hypointensity on T1-weighted imaging and hyperintensity on T2-weighted imaging, without restricted diffusion. On enhanced scan, centripetal enhancement appeared within an area of the lesion, and the nodule showed hypointensity on diffusion-weighted imaging (DWI). The splenic nodule showed increased apparent diffusion coefficient (ADC) values compared with the surrounding splenic tissue (*Figure 1E-1G*). MRI images suggested a lesion of a "vascular" nature. Integration of these findings pointed towards a benign diagnosis of this lesion.

The patient declined percutaneous biopsy because of the high risk of bleeding complications. Then, he underwent laparoscopic splenectomy. He was placed in a right semi-decubitus position and his abdomen was opened at the upper umbilicus, subxiphoid position, subxiphoid position, left midaxillary line, rectus incision and mid-abdominal incision after general anesthesia. The gastrosplenic, splenocolic ligaments and splenic artery were divided gradually, and the splenic hilum was visualized and transected. The spleen and an accessory spleen were placed in a retrieval bag and

retracted via the midabdominal incision. Finally, a surgical drain was placed in the splenic fossa. The total operating time was less than 120 minutes with a blood loss of 50 mL. He was discharged from the hospital without any complications a few days later. The cut section showed a circumscribed grey-red mass measuring 3.5×3.5 cm (*Figure 1H*). Histopathologically, the lesion was characterized by dilated anastomosing vascular channels with cystic dilatation and multiple intraluminal papillary protrusion. The vascular channels had a focal pseudopapillary appearance, and the surface was lined with a monolayer of endothelial cells without significant cytological atypia (*Figure 1I*). Partial cells displayed hemosiderin and hemophagocytosis. Immunohistochemical studies exhibited expression of FVIII, ERG, CD31, CD 34, and CD 68, and both endothelial and histiocytic cells, but CD8 was not detected. The postoperative histopathological analysis confirmed LCA. Cellular atypia was not found. During a telephone follow-up 6 months later, it was confirmed that the results of an ultrasound at the local hospital had been clear.

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was provided by the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Discussion

LCA usually manifests as multifocal nodules in the spleen, and only a few cases involving a solitary lesion have been reported in the literature. LCA may occur at any age in patients of either gender, and it clinically manifests as abdominal pain, pyrexia, splenomegaly, anemia and pancytopenia. Approximately 27% of patients who are diagnosed with LCA have coexistent visceral malignancies and 17% of patients have an immune disorder in the form of an autoimmune condition (2). To date, relatively little research is available on SWE and CEUS in LCA. SWE can assess the stiffness of splenic nodules, and CEUS can display their parenchymal microvasculature. The CEUS pattern of our case showed a rapidly hyperechoic enhancement pattern with short filling time in the early phase. Then, the central part of the splenic nodule appeared slightly washed out. However, progressive enhancement from the periphery was detected when a slow-motion videotape was replayed. In contrast to previous related studies, our MRI images showed a relatively hyperintense splenic lesion on T2 signal intensity (3,4). We suggest that the difference is due to the varied content of hemosiderin in the hemophagocytic cells. In addition, previous research reported that vascular lesions have increased ADC values, and solid splenic tumors, fungal infections and abscesses have reduced ADC values (5). The MRI images suggested a "vascular" nature of the splenic lesion.

Hypersplenism, splenic soft nodule, progressive enhancement pattern and increased ADC values maybe helpful in the diagnosis of LCA. The diagnosis of LCA is difficult to make preoperatively, because the imaging patterns of splenic lesions may exhibit similarities. Primary differential diagnoses include lymphomas, angiosarcoma, hemangioma and harmatoma. Lymphomas are the most common malignancy of the spleen. Discrete lesions appear as hypoechoic targetoid masses on ultrasound. The typical CEUS pattern is characterized by marked inhomogeneous or homogeneous contrast enhancement during the early arterial phase followed by rapid wash-out during the parenchymal phase (6). Lymphomatous deposits are hypoenhanced compared with the surrounding splenic parenchyma in the portal venous and delayed phases on MRI enhanced scan. Splenic lymphoma may show diffusion restriction on DWI sequences. Spleen angiosarcoma is a very rare, malignant neoplasm of vascular origin and is more prone to metastasis and rupture. The ultrasound images showed an enlarged spleen containing solitary or multiple poorly circumscribed heterogeneous echogenic mass(es), with anechoic areas due to the presence of necrosis. Using CEUS, angiosarcoma may show hypoenhancement in the arterial phase, and the lesions may display rapid wash-out in the parenchymal phase. On MRI, the lesions usually appear with mixed T1 and T2 signal intensity due to necrosis and hemorrhage. In addition, contrast enhancement is variable, and heterogeneous enhancement may be demonstrated (7). Hemangiomas are the most common benign vascular tumors and showcase a well-demarcated homogenous hypoechoic or hyperechoic lesion on ultrasound. The common enhancement pattern of splenic hemangioma on CEUS has been shown to be early peripheral nodular enhancement followed by centripetal fill-in or homogeneous hypervascularized pattern in all phases (6). On MRI, hemangiomas are iso- to hypo-intense on T1-weighted imaging and hyperintense on T2-weighted imaging. The contrast MRI images of hemangiomas are characterized by a gradual, progressive enhancement in the portal venous and delayed phases. DWI sequences show the nodule to be

hyperintense (7). Hamartomas are rare, benign lesions of the spleen and showcase a well-demarcated homogenous lesion with or without cystic center on ultrasound. MRI will show a lesion, which is hypointense on T2-weighted sequences and hyperintense on T1-weighted sequences. The mass shows a relatively homogeneously enhancing mass in the spleen on contrast MRI images (7). Splenic hamartomas show hypointensity on DWI. Ultrasound and MRI imaging techniques provide a non-invasive and valuable evaluation for the diagnosis and differential diagnosis of focal splenic lesions. LCA is often viewed as a benign tumor, but malignant LCAs have been reported. Fernandez *et al.* published a case report of a patient with metastatic lesions in the liver and retroperitoneum 4 years after splenectomy for LCA (8). Due to its potential malignant biological behavior, splenectomy is recommended for treatment. It has been recommended that patients with a malignant tendency of the LCA undergo postoperative checks at 6 months and 1 year. Some case reports show malignant transformation which is alarming and may necessitate a close follow-up with repeat diagnostic imaging at 6 to 12 months. Of course, patients should undergo prudent long-term follow-up after an operation. Patients receiving conservative therapy may require close follow-up with repeat diagnostic imaging at 3 to 6 months (4).

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-897/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was provided by the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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