Clinical Case Report

Focal nodular hyperplasia on an accessory liver lobe
A case report and literature review

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Abstract
Introduction: Accessory liver lobe (ALL), an autonomous island of normal liver parenchyma, is a rare congenital anomaly that is difficult for preoperative diagnosis and often identified incidentally. It can also be accompanied with benign or malignant diseases, which is extremely rare. There are only 3 cases of focal nodular hyperplasia (FNH) detected on ALL reported by previous literature.

Patient concerns: A 33-year-old woman was incidentally diagnosed with a mass in left upper quadrant abdomen by a routine ultrasound examination. Doppler ultrasound revealed that the mass was attached to left liver lobe with a vascular pedicle. A spoke-wheel artery with diffuse enhancement during hepatic arterial phase was visualized on contrast-enhanced ultrasound, and the mass was continuously hyper-enhanced with a hypo-enhanced intralesional scar during the portal and delayed phase. And contrast-enhanced computed tomography showed a similar enhancement mode of the mass.

Diagnosis: The mass was resected and postoperative histopathologic result of the lesion revealed a nodular hyperplastic parenchyma with a central fibrous scar, without tumor cells. And a final diagnosis of FNH on ALL was determined accordingly.

Interventions: Mass resection was conducted according to patient’s demand.

Outcome: After general postoperative administration, the patient was discharged. Then, she had been undergoing regular serological tests and imaging examinations in our hospital for 24 months.

Conclusion: The finding of a mass connecting with liver by a stalk should alert the clinician of the possibility of ALL, as well as benign or malignancies on an ALL. This is the first case of FNH on ALL preoperatively confirmed by contrast-enhanced ultrasound. We suggest that an integrated radiologic approach is crucial to evaluate an incidentally detected, asymptomatic abdominal focal mass.

Abbreviations: ALL = accessory liver lobe, CEUS = contrast-enhanced ultrasound, CECT = contrast-enhanced computed tomography, FNH = focal nodular hyperplasia, HAP = hepatic arterial phase, MSCTA = multi-slice spiral computed tomographic angiography.

Keywords: accessory liver lobe, case report, focal nodular hyperplasia, integrated radiologic approach

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1. Introduction
Accessory live lobe (ALL) is a rare congenital liver anomaly, whose incidence is 0.09% based on data described in studies of laparoscopy and autopsy.[1] Preoperative diagnosis of ALL is highly challenging, as most of them are asymptomatic unless a pedicle torsion, compressive effect, or neoplastic change occurs.[2–4] ALL may have pathologic features similar to anatomically normal liver, thus leading to the incidence of benign or malignant diseases that appear in liver tissues, which is an extremely rare condition. Benign diseases on ALL, including hemangioma, focal nodular hyperplasia (FNH) and adenoma, have lower incidence than that of malignancies. Only 3 FNH on ALL cases have been reported in English literature.[1–5] Herein, we presented case of an asymptomatic mass that was verified as FNH on an ALL finally in this study, and we paid special attention on its imaging presentations.

2. Case presentation
A 33-year-old woman was incidentally diagnosed with a mass in left upper quadrant abdomen by a routine ultrasound scanning, with no specific personal and family history. For physical
examination, there was no palpable mass, tenderness, or rebound
tenderness. Laboratory findings including HBV and HCV tests,
amylase, liver function tests autoantibodies and tumor markers
(serum α-fetoprotein, CA 199 and CA 125) were normal.
Doppler ultrasound demonstrated an enlarged and distorted
artery in her left liver lobe. By tracing the vessel, a well-defined
hypoechoic mass was found outside the left liver lobe, 83 mm ×
59 mm × 56 mm in diameter (Figs. 1A and 1B). The mass was
close to the spleen, relative movement was present during
respiration (Fig. 1C), and the mass was highly vascularized by
Doppler (Figs. 1D and 1E).

We performed Contrast-enhanced ultrasound (CEUS) using an
intravenous dose of 2.4 mL SonoVue (Bracco Inc., Milan, Italy)
for further investigation. Regarding the normal liver tissue as
control, the mass had a remarkable quick wash-in during hepatic
arterial phase (HAP), with a centrifugal enhancement pattern
(Fig. 2A). Continuous hyper-enhancement with a hypo-enhanced
intralesional scar was seen in the portal and delayed phase
(Fig. 2B). A second dose of 1.2 mL of SonoVue was then
administered, the blood supply of the mass was found arising
from a vascular pedicle, which originated from distorted artery
inside the left liver. Furthermore, the venous flow of the mass was
found to drain into enlarged left hepatic vein through the
connected stalk.

Contrast-enhanced computed tomography (CECT) confirmed
the presence of a large and well-defined abdominal mass between
spleen and diaphragm, attached to the left liver lobe with a stalk.
In the arterial phase, the mass was avidly and homogeneously
enhanced with a central irregular hypodense area. Moreover,
multi-slice spiral computed tomographic angiography (MSCTA)
and volume rendering technique images suggested that the blood
supply of the mass originated from a single distorted artery
directly from the aorta (Fig. 3A), with venous flow draining into
left hepatic vein (Fig. 3B).
Initially diagnosing the mass as FNH in an ALL according to the imaging manifestation, we performed laparotomy subsequently which revealed that the mass was located in the left liver triangular ligament, connecting to the lateral segment of the left hepatic lobe through a pedicle. The mass was covered with a fibrous capsule and multiple blood vessels. A central fibrous scar was also found by laparotomy (Figs. 3C and 3D). Postoperative histopathology of the mass demonstrated a nodular hyperplastic parenchyma with a central fibrous scar, containing proliferation of small bile ducts and irregular tortuous arteries with thickened walls, veins, and capillaries, and no tumor cell was detected by histology (Fig. 3E). Therefore, the histopathological result confirmed the diagnosis of classical FNH on an ALL.

After general postoperative administration, the patient was discharged and received regular serological tests and imaging examinations in our hospital for 24 months. The patient has provided informed consent for publication of the case.

3. Discussion

Accessory liver tissue is an autonomous island of normal liver parenchyma. They may be located at various sites, most frequently on the inside of the gallbladder, and may also be involved with ligaments around liver, spleen, portal vein, umbilicus, adrenal grand, pancreas, esophagus, diaphragm, and thoracic cavity.[2,5]

There are 2 types of accessory liver lesions. The first is an accessory lobe connected to the liver, named as accessory live lobe (ALL), which was seen in 0.09% of patients. And the second is the ectopic liver completely separated from the liver, which was seen in 0.47% of patients.[1] However, it is difficult to make a clear distinction between the 2 types in many cases, while the precise incidence is unknown.

ALL is subject to the same benign or malignant diseases as the original liver tissues,[4] which may be caused by abnormalities of vascular supply and biliary drainage.[3] It is reported that ALL associated with benign lesions are rarer than with malignancies, benign diseases that have been reported including hemangioma, FNH, and adenoma.[4]

In this case, an abdominal mass was found between left liver lobe, spleen, and diaphragm during routine examination. Grey scale and Doppler ultrasound revealed that the mass was attached to left liver lobe by a vascular pedicle, with its characteristics unknown. Furthermore, CEUS showed a spoke-wheel artery with diffuse enhancement during HAP, and the mass was continuously hyper-enhanced with a hypo-enhanced intralesional scar in the portal and delayed phase. These radiologic findings highly suggested the diagnosis of FNH. As the mass was outside but connected to liver by a stalk, it was considered as FNH on an ALL, which was finally proved by pathology.

We searched the database, including PubMed, Scopus, and Excerpt Medica Database (EMBASE), to identify studies evaluating FNH on ALL. According to previous literature, only 3 cases of FNH on ALL have been reported in English literature (Table 1). These 4 patients had different signs when referring to hospital, of whom 2 had abdominal pain, 1 had an isolated increase in GGT. These lesions were found in areas between spleen and diaphragm, between kidney and liver, and in the pelvis. Clinical manifestations were depended on compression of adjacent organs, the location, and pedicle torsion of ALL.[6,7]
Demonstrated by imaging results, the 3 lesions and the case in our study were all connected to liver tissue by a vascular pedicle. They had large hepatic artery branch and a prominent vein, which drained directly into the hepatic vein or inferior vena cava. Specific imaging features were visualized in CEUS and CECT imaging, such as diffuse hyper-enhancement during HAP, continuous hyper-enhancement with hypo-enhanced intralobular scar in the portal and delayed phase. Totally, all the 4 lesions can be considered as FNH on an ALL by imaging examinations before surgery.

To our knowledge, we present the first case of FNH on ALL pre-operatively which was confirmed by CEUS. CEUS showed a blood vessel arising from aorta, distorted within the liver, and then connected to the extrahepatic mass by a vascular pedicle, consistent with intra-operative observation. However, MSCTA could only indicate the vascular pedicle arose from the aorta, and the vessel distortion through the liver could not be seen, for liver tissue could not be visualized by MSCTA. CEUS is a minimally invasive method for preoperative evaluation of abdominal masses. They can show a real-time vascular map of the mass, also the relationships with organs, thus indicating the characteristics of the blood supply. Moreover, the microbubbles used as the contrast agents for CEUS possess high safety, and seldom cause hepatotoxicity and allergies. Therefore, as a non-ionizing and non-radiative imaging method, CEUS can be an alternative to CT/CECT, especially for pediatric patients, for whom radiative imaging methods are not suitable. Experienced radiologists are needed to perform CEUS, which is the main limitation of the novel method, but its wide clinical utilization can still be expected.

4. Conclusion

In conclusion, we supposed that FNH on an ALL can be diagnosed by integrated radiological examinations before surgery. When a mass is found in a special location of the abdominal or pelvic cavity, it is necessary to use integrated imaging approaches to evaluate the lesion. If the mass is outside but attaches to the liver with a vascular pedicle, it is highly indicated as an ALL. In the meanwhile, we should better maintain an awareness of the possibility of benign or malignancies on an ALL. Further diagnostic confidence may be achieved with the use of CEUS, which provides clear vascular map in a non-invasive way.

Author contributions

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Table 1

| Study          | Yr   | Patient | Clinical manifestation | Radiological examination                                                                 | Location            | Size        | Connection with mother liver                  |
|----------------|------|---------|------------------------|------------------------------------------------------------------------------------------|---------------------|------------|-----------------------------------------------|
| Leone N, et al [4] | 2005 | 61/F    | None except increased GGT | US: a pedunculated hypoechogenic lesion with rich arterial vasculatization, MRI: thin stalk, central scar | Between right kidney and hepatic right lobe | 6 cm       | Attached to the right liver by a slim stalk   |
| Ruiz Hierro C, et al [3] | 2013 | 13/F    | Abdominal pain/vomit   | US: a homogeneous and high vascularized mass, CECT: a homogeneous mass attached to liver by a pedicle, with hypodense areas | Pelvis              | 10.7 x 10 cm | Attached to the left liver by a broad vascular pedicle |
| Dreizin D, et al [5] | 2014 | 45/F    | Abdominal pain         | CECT: HAP: avidly enhancing, homogeneous mass with central low attenuation, with a hepatic artery branch and prominent vein drained into IVC, MRI: thin stalk | Between spleen and diaphragm | /          | Attached to the left liver by a thin stalk    |
| Na             | 2020 | 33/F    | None                   | US: hypoechogenic mass outside the liver, highly vascularized CEUS: quick wash-in and centrifugal enhancement in HAP, no early wash-out CECT: homogenously enhanced with a central irregular hypodense area MRI: blood supply originated from aorta, and venous flow drained into left hepatic vein | Between spleen and diaphragm | 8.3 x 5.9 x 5.6 cm | Attached to the left liver with a stalk |
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