

Case Report

Magnetic resonance imaging findings of ectopic liver in the adrenal gland with associated hypertension: a case report and literature review

Yanxiu Han¹, Jie Zhang¹, Haili Wang¹, Mengyuan Zhao², Yan Li³, Zhaoqin Huang¹

Departments of ¹Radiology, ²Pathology, Shandong Provincial Hospital Affiliated to Shandong University, Jinan, Shandong, China; ³Department of Ultrasound, The Second Hospital of Shandong University, Jinan, Shandong, China

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Abstract: Ectopic liver is a rare developmental abnormality. Here, we report the magnetic resonance imaging (MRI) findings of ectopic liver in the adrenal gland that was misdiagnosed as pheochromocytoma before surgery in a 41-year-old woman. This case is unique in some ways. MRI of ectopic liver in the adrenal gland has not been described before. Also, the patient had clinical symptoms of hypertension, which completely disappeared after tumor resection.

Keywords: Adrenal gland, ectopic liver, magnetic resonance imaging (MRI), hypertension

Introduction

Ectopic liver tissue can be found at various sites in the body, usually in the abdomen, retroperitoneum and the chest [1-4], especially around the liver. This is a rare disease, with an incidence of about 0.24%~0.47% [5]. The presence of ectopic liver tissue is usually asymptomatic, but occasionally, it causes unexpected problems such as hepatocarcinogenesis [6]. We describe a case of symptomatic ectopic liver tissue, which was resected postoperatively to prevent the potential risk of malignant transformation.

Case report

A 41-year-old woman presented with a six-month history of hypertension accompanying vague chest discomfort, who was admitted to our hospital on March 12, 2015. Her highest blood pressure value observed was 160/110 mmHg and was controlled by oral nifedipine sustained-release tablets and phenoxybenzamine. Furthermore, there was a family history of hypertension in her mother. The patient was diagnosed as not suffering from diabetes, coronary heart disease nor hepatitis. Physical ex-

amination found nothing unusual. The laboratory tests, including complete blood count and liver function, were in the normal range. Hepatitis B virus (HBV) and hepatitis C virus (HCV) tests were negative.

Magnetic resonance imaging (MRI) showed a round, well-defined soft-tissue mass in the right adrenal gland, with a size of 5.8 × 3.9 × 6.2 cm. The normal adrenal gland could be seen partially, and the boundary between the lesion and the distal branch was unclear (**Figure 1C**). The lesion gave a heterogeneous signal that was different from the normal liver parenchyma on T1-weighted imaging (WI) and T2WI sequences on MRI. Compared to the liver parenchyma, the lesion showed higher signal intensity on the T2WI and similar or slightly lower signal intensity on the T1WI (**Figure 1A, 1B**). Linear and spot hyperintensity was observed in the lesion on T2WI, which showed hypointensity on T1WI. The mass showed heterogeneous, significant enhancement during the arterial phase (**Figure 1E**) and continued enhancement during the venous phase and delayed phase (**Figure 1F, 1G**). The spot or line-like hypointensity was completely filled with higher signal during the delayed 10-min period (**Figure 1H**). The lesion

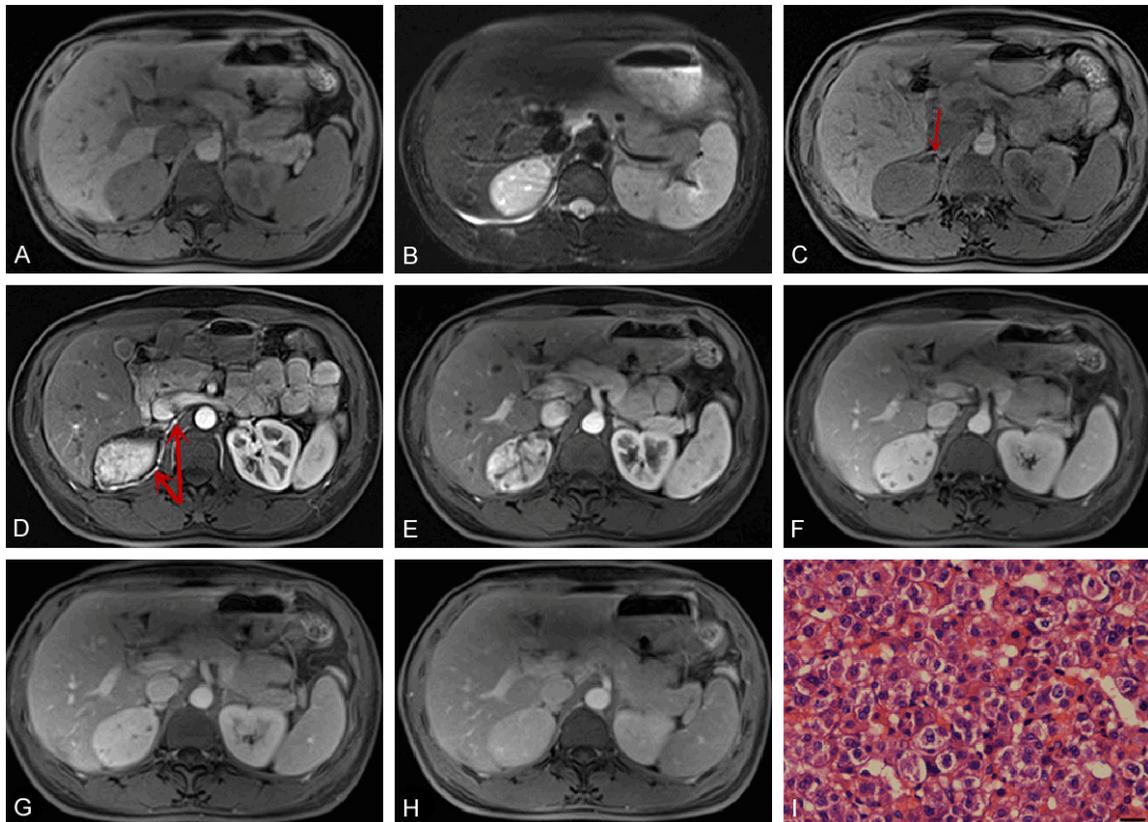


Figure 1. A, B. Magnetic resonance imaging (MRI) showed the mass is in the right adrenal gland around the liver. Compared with the liver, there were higher signal intensity on the T2WI and similar or slightly lower signal intensity on the T1WI. C. Normal adrenal gland is partial visible, and the distal branch and the lesion are unclear (red arrows). D. Blood supply of blood vessels is visible around the lesion (red arrows). E. The mass showed heterogeneous, significant enhancement during the arterial phase. F, G. Continued enhancement during the venous phase and delayed phase. H. Spot or line-like hypointensity areas within the lesion were completely filled with a higher signal during the delayed 10 min period. I. Normal liver cells are arranged in a slight disorder (Hematoxylin-eosin staining; magnification $\times 400$; Bar = 50 μm).

was rich in blood supply, being surrounded by blood vessels from the abdominal aorta (**Figure 1D**). The initial impression of the imaging, combined with the clinical history, was pheochromocytoma.

A right adrenal tumor resection was performed on March 19, 2015. During the surgery, the mass was confirmed to be in the right adrenal gland. Although the mass adhered to the surrounding tissues of the kidney due to chronic inflammation, it was completely separated from the liver. Macroscopic pathological examination of the specimen showed a dark red and tough texture with a cutting area of 6×4 cm. Immunohistochemical staining revealed focally expressed Hepper-1 and was negative for Syn, CgA, Me1nnA and CK7. The pathological diagnosis was ectopic liver located in the right adre-

nal gland (**Figure 1I**). Postoperative follow-up on November 3rd, 2018 showed that the patient had no symptoms, normal blood pressure control and normal liver function.

Discussion

Collan et al [7]. defined ectopic liver as liver tissue lying obviously outside the liver without any connection with the liver, which is confirmed by macroscopic and microscopic pathology. In contrast, accessory liver is described as liver tissue connected to the liver, often by a stalk. The incidence of ectopic liver has been reported to be between 0.24% and 0.47% [5], but more modern studies have documented a higher incidence during laparoscopic surgery, about 0.7% [8]. Reviewing embryologic hepatogenesis can help explain the wide range of ectopic liver

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locations. Hepatic diverticulum, which can develop into liver, is derived from endothelial cell proliferation in the terminal ventral wall of the foregut [9]. Ectopic liver may be due to the initial hepatic cells differentiating into an independent hepatic tissue during embryonic development, usually near the liver. Common locations include the gallbladder, spleen, retroperitoneum, pancreas, adrenal gland, portal vein, diaphragm, thorax, gastric serosa, umbilical cord, and placenta [10-16]; gallbladder is the most common site of occurrence.

It is important to consider ectopic liver in the assessment of perihepatic lesions. Statta [17] believed that diagnosis should be based on a biopsy of the ectopic liver or an MRI with liver-specific contrast. In theory, the ectopic liver should be consistent with the normal parenchymal liver signal [18, 19], but this was not the case in our report. Compared to the liver, there was a higher signal intensity on the T2WI and a similar or slightly lower signal intensity on the T1WI, which may be associated with abnormal blood supply. In addition, this case was difficult to distinguish from adrenal pheochromocytoma. When the tumor of pheochromocytoma is larger, the signal is usually less uniform, due to bleeding, necrosis and cystic change; the T2WI signal is significantly hyperintense due to the rich water and blood sinuses; diffusion is limited; and the enhanced arterial phase shows significantly heterogeneous enhancement. Although our case exhibited significant heterogeneous enhancement and showed a high signal on T2WI, diffusion was not limited. In addition, a round-like hyperintense region was observed on T2WI, showing a roundish cystic hypointensity in the arterial phase, but it was filled with contrast agent in the delayed phase, indicating that the region was not cystic or necrotic. The T2WI coronal display showed that the area was a strip, similar to the intrahepatic bile duct structure signal.

Hepatocytes in ectopic livers usually mimic normal liver cells and show relatively similar pathological pattern [20]. Therefore, ectopic liver tissues usually display normal histological structure and are subject to the same risk factors and pathological processes as native liver tissue, such as chronic hepatitis (HBV, HCV infection), steatosis, adenoma, hemangiomas, focal nodular hyperplasia, and even cancer [21]. However, benign lesions do not seem to be fre-

quent; the higher frequency of malignancies may be due to many benign lesions remaining undiagnosed because they are asymptomatic. Moreover, due to their abnormal locations, asymptomatic lesions may be misdiagnosed in the absence of histology.

Ectopic liver tissue is more predisposed to malignancy than normal liver tissue [6, 22]. However, the reason why ectopic liver is particularly susceptible to tumor degeneration is still unclear. Carcinogenesis is a complex multi-step process involving multiple factors. Ectopic liver tissue may be metabolically defective due to the lack of normal vascular and catheter systems, which lead to a longer exposure to various carcinogenic factors [6, 20]. The potential microenvironment may lead to sustained cellular stress, which may lead to cell death and compensatory cell proliferation [6, 20]. Increased cell renewal may lead to genetic mutations and subsequent development of cancer [21, 22]. Although the pathology of our case shows bile ducts, there is no normal bile duct drainage system. The vascular system was similar. The mother liver had portal vein drainage, but the drainage system in the case of the ectopic hepatic tissue was not obvious. In most cases, malignant tumors are found in ectopic livers, but no malignant tumors were found in the mother liver, and even hepatitis and cirrhosis were not manifest [6, 22]. Ectopic liver usually has no clinical symptoms, but it can cause unexpected clinical problems in rare cases, when its torsion, infarction, compression of adjacent organs and rupture lead to intraperitoneal bleeding [23, 24].

Fortunately, this case did not develop any type of pathology. However, the lesion was large and the blood supply was abnormally abundant. We cannot predict its subsequent development, nor can we completely rule out whether it would have caused complications. Coupled with the patient's symptoms of hypertension, surgical treatment was necessary. As far as we know, our case is the first case of adrenal ectopic liver with hypertension. In short, despite the low incidence of ectopic liver and the few complications, it is necessary to maintain awareness of this possibility in addition to the characteristics of the site of the disease. More importantly, the potential risk of malignancy of ectopic liver tissue is a basis for a complete resection.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Zhaoqin Huang, Department of Radiology, Shandong Provincial Hospital Affiliated to Shandong University, Jingshi Road 9677#, Jinan 250000, Shandong, China. Tel: 15-168887535; E-mail: devin813@163.com

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