

Accessory Splenic Mass Masquerading as Hepatocellular Carcinoma: A Diagnostic Dilemma

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1. Abstract

The spleen plays a pivotal role in our immune system by facilitating the proliferation and differentiation of lymphocytes and monocytes. Typically located in the left upper quadrant retroperitoneally, splenic tissue found outside of its usual position is termed ectopic spleen. When the tissue maintains its histological architecture and encapsulation, and receives blood supply from splanchnic vessels, it is called an accessory spleen. Although it commonly presents near the splenic hilum or pancreatic tail, rare instances have been reported in the gastric, liver, gastrosplenic/lienorenal ligaments, as well as thoracic and gonadal regions. However, the case of an accessory spleen, mimicking a hepatic lesion in the right diaphragm represents a novel observation.

2. Introduction

Accessory spleen, also referred to as polysplenia or splenunculus is a condition in which an additional spleen forms separately from the primary spleen due to congenital maldevelopment during embryogenesis. It is estimated that nearly 10-30% of the total population possesses an accessory spleen, and median diameter of the accessory spleen ranges from 2mm to

3.5cm[1–3]. Development of the primordial spleen commences from the splanchnic mesoderm during the 5th week of gestation. This condition is postulated to occur due to either the failure of embryonic splenic tissue to unite in the dorsal mesogastrium or the detachment of tissue from the main spleen due to extreme lobulation during embryonic development. Additionally, abnormal rotation of the stomach and migration of the dorsal mesogastrium during 7th and 8th week of embryogenesis may contribute to this condition. The most common location of an accessory spleen can be found in the hilum of the spleen, pancreas, the gastrosplenic ligament, lienorenal ligament and greater omental areas [1,4,5]. Herein, we would like to present a rare presentation of an accessory spleen located at the right diaphragm mimicking a hepatoma lesion, found incidentally in a healthy 39-year-old adult with no significant past medical history.

3. Case presentation

A 39-year-old male patient, with no significant past medical history, presented to the hospital due to an incidental finding of a liver mass during a work-up for hematuria. Although recently diagnosed with hypertension, he discontinued his antihypertensive medications after a week. He has no past history of hepatitis B or C infection or past surgical interventions, known drug allergies. He doesn't take any medications. Socially, he is married, a non-smoker, and abstains from alcohol. His family history reveals no cancer or liver-related issues in his parents and immediate relatives live in China. On physical examination, his vital were mostly within normal limits except mild elevation of blood pressure of 139/92 mmHg. His cardiac, chest, abdominal examinations were unrevealing so far. There is no skin rash or abnormal swelling or edema detected throughout his body.

Routine laboratory investigations including complete blood counts (CBC), basic metabolic panel (BMP), liver functional test (LFT), coagulation studies and hepatitis viral panels were unremarkable as well as the tumor markers such as alpha-feto protein (AFP), and chorionic embryonic antigen (CEA). Further investigations following a visit to his urologist for his hematuria and prescribed antibiotics for possible urinary tract infection together with an order of the CT scan of the abdomen and pelvis. The result revealed a 2-cm enhancing lesion in the right hepatic dome with moderate hepatomegaly and hepatic steatosis, which finding was recommended to repeat with an MRI scan. Patient returned to his primary care doctor and received MRI scan as requested. MRI scan result revealed a 2.3 cm right hepatic dome lesion that showed signal intensity on T2 weighted image with heterogeneous enhancement on arterial phase and persistent enhancement on venous phase. Additional 0.5cm lesion was found in the segment 6 of right hepatic lobe, showing both arterial and venous enhancement. Both of these lesions concluded indeterminate in pathology and recommended

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another MRI scan with gadoxetate disodium for better characterization of liver lesions. It unveiled a mildly T2 hyperintense 2 x 2 cm arterial enhancing mass in the right hepatic lobe showing washout at the venous phase and absent contrast uptake in the hepatobiliary phase with restricted diffusion pattern, concerning for malignancy (figure 1 and 2).

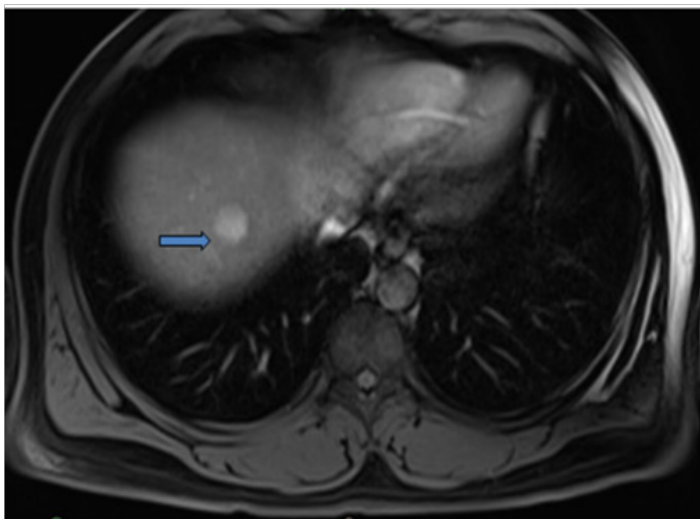


Figure 1: MRI of the liver in transverse section showing enhancing mass (blue arrow) during arterial phase

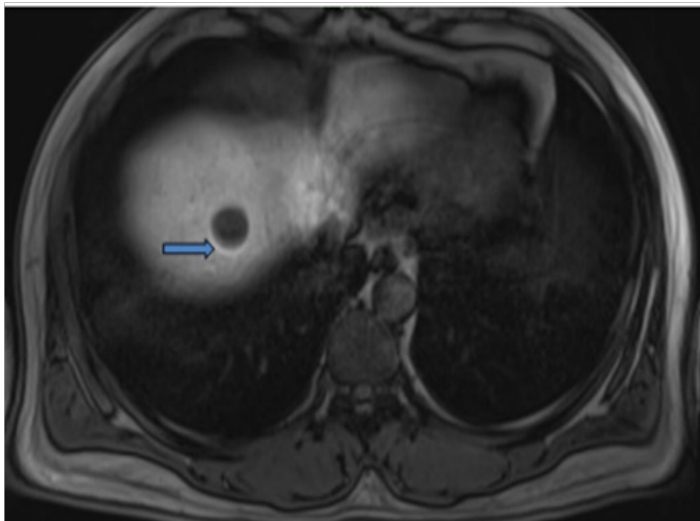


Figure 2: MRI of the liver in transverse section showing the same mass (blue arrow) manifesting washout phenomena during venous phase

Additional 0.7cm lesion in the segment 6 of the liver was identified as a benign focal nodular hyperplasia (FNH) or arterial portal shunt. Due to the concern for hepatocellular carcinoma (HCC), patient was referred to the intervention radiologist (IR) for biopsy after discussing the case and having consensus at the Hepatobiliary tumor board. The IR procedure was unsuccessful after finding difficult to identify the mass during the ultrasound-guided biopsy procedure, which led to a laparoscopic resection of the mass. He underwent laparoscopic resection of the right

diaphragmatic mass and mesh repair of the diaphragm defect as the mass was found to be distinct from the liver and attached to the right diaphragm during surgery. The surgical pathology revealed mature encapsulated splenic tissue with white and red pulps, confirming the diagnosis of accessory spleen (figure 3 and 4). He tolerated the procedure well without any complications. On follow-up visit at the clinic, he was doing well with minimal tenderness at the surgery site.

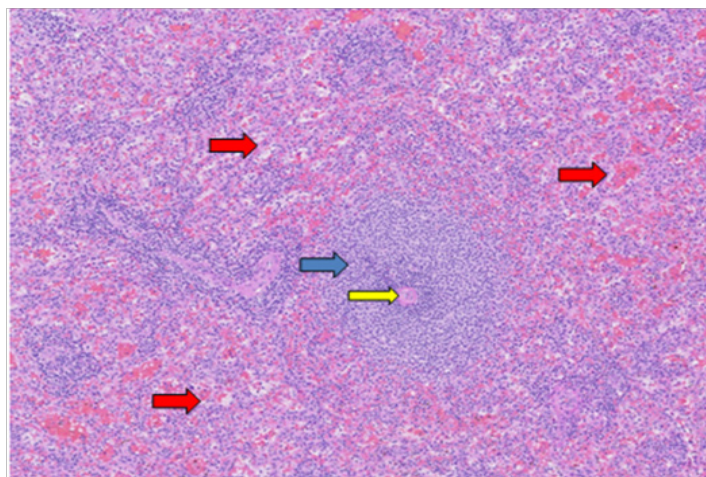


Figure 3: Splenic tissue histology at the medium power showing white pulp (blue arrow) surrounding a central arteriole (yellow arrow) and red pulps (red arrow) at peripherals

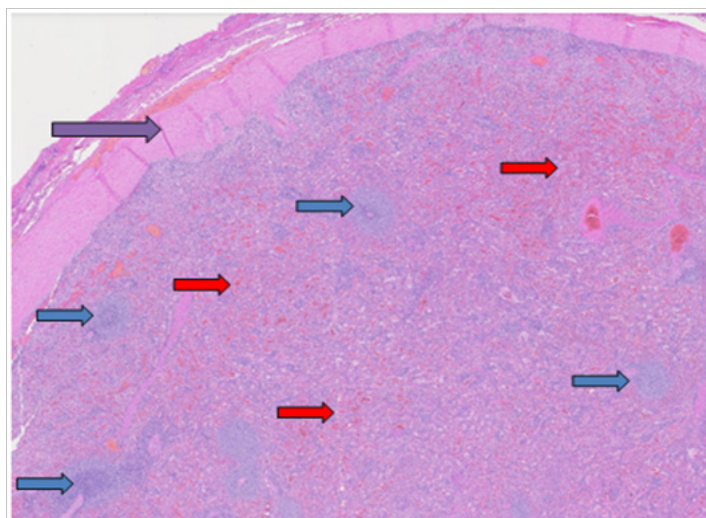


Figure 4: Splenic tissue histology at the low power demonstrating white pulps (blue arrow) and surrounding red pulps (red arrow) with splenic capsule (yellow arrow) encapsulating the bulk of splenic parenchyma the top of the figure

4. Discussion

From our literature review, the case of an accessory spleen located in the right diaphragm, mimicking an hepatocellular carcinoma (HCC) appears to be the first of its kind reported. The diaphragm originates from

multiple embryogenic sources, including the septum transversum, a mesodermal sheet, that initially separates the thorax and abdomen during early embryogenic development[6]. We hypothesize that our patient's accessory spleen, also of mesodermal in origin, may have migrated to the right hemidiaphragm due to anomalies during extreme lobulation of the primordial spleen in embryogenesis. However, the precise mechanism by which the accessory spleen in our patient's right diaphragm occurred remains unknown. Accessory spleens are typically incidental findings during radiological examinations conducted for other medical reasons and generally do not cause symptoms or complications. However, in some cases, they can lead to acute complications such as rupture due to trauma or torsion resulting in infarction [7–9]. Accessory spleen may also take over the role and responsibility of a normal spleen in individuals who have undergone splenectomy, as it possesses similar histological architecture and functional capacity. Therefore, in cases where diseases such as immune thrombocytopenia, or hereditary spherocytosis, recur or remain uncontrolled after splenectomy, the presence of possible functioning accessory spleen elsewhere in the body should be considered [11].

This kind of scenario is not possible in case of splenosis, which develops from a traumatic injury or prior surgery to the spleen and migrated to the other parts of the body, whereas accessory spleen arises from anomalies during embryogenesis. Furthermore, an accessory spleen maintains its blood supply from its mother splanchnic blood vessels with preserved normal splenic architecture and a complete capsule, whereas splenosis acquires its blood supply from local blood vessels, with distorted or incomplete splenic architecture [10]. In our patient, the biopsy results revealed an encapsulated mature splenic tissue confirming the accessory spleen. As CT scan is the most common imaging modality in clinical practice, we should be aware of the typical findings of the accessory spleen, as it can mimic the lymphadenopathy or malignancy if detected in other organs. According to Angarita et al., the sensitivity of a CT scan for detecting an accessory spleen is 60% and specificity is 95.6% [11]. Most accessory spleens exhibit homogeneous enhancement, with similar attenuation as the main spleen during contrast CT imaging, and typically display characteristic rounded, separate margins [2,12]. However, distinguishing these lesions from cancerous origins in most clinical settings proves challenging due to their small size, approximately 2cm, and their particular location within organs such as the liver or the pancreas. Differentiating them from HCC, the neuroendocrine tumor of the pancreas or metastatic cancer is particularly difficult [3,13]. Consequently, this often results in unnecessary biopsies and excision of the lesions. While histopathological examination remains the gold standard for diagnosis of an accessory spleen, it necessitates unnecessary excision or biopsy of the lesions. Currently, the preferred non-invasive test involves functional scans, including Tc-99m-labeled heat-denatured red blood cells (HRBC), and Tc-99m sulfur colloid scans. However, these scans require specialized expertise & facility and are not widely available like CT scans [13–15]. In our patient, concern over HCC persisted after MRI scan with gadoxetate disodium (Eovist), which showed arterial enhancement with washout phenomena in the venous phase as seen in HCC. Since repeated imaging studies failed to accurately diagnose the lesion pre-operatively, ultimately

resulting in laparoscopic removal of the accessory spleen.

5. Conclusion

In summary, our case underscores the diagnostic challenge posed by accessory spleen in the atypical sites such as the right diaphragm as observed in our patient. Despite advancements in imaging modalities such as MRI scan with Eovist, distinguishing accessory spleens from cancerous lesions remains a diagnostic challenge. Moving forward, heightened clinical suspicion and diagnostic awareness among clinicians and radiologists regarding the typical imaging features of accessory spleens are imperative to avoid misdiagnosis and unnecessary surgical interventions. This involves interdisciplinary collaboration and when necessary, the utilization of advanced imaging techniques to facilitate accurate diagnosis especially in young individuals with low risks for cancer.

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