Hepatic Peliosis Associated with a Paraganglioma

Mahbuba Shirin

Associate Professor, Department of Radiology & Imaging, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh

Abstract
Peliosis of the liver and spleen is a rare benign condition characterized by dilatation of sinusoidal blood-filled spaces. The imageology of hepatic peliosis resembles hepatocellular carcinoma, hepatic metastases and hemangioma. We report a case of hepatic peliosis imitating metastases on CT. The patient was 26-year-old man who previously had a splenectomy and retroperitoneal paraganglioma surgically removed. Peliosis must be considered a potential differential diagnosis of hypodense foci of the liver seen on CT.

Keywords: Peliosis, CT scan, FNAC

Background
Peliosis is characterized by blood-filled thin-walled vessels/cavities with a cystic appearance often found throughout the parenchyma of the organ involved [1]. “Peliosis” is a term derived from the Greek pelios, which means “dusky” or “purple,” referring to the color of the liver parenchyma with peliosis. The lesion may be found in liver, and may be seen in the spleen, lymph nodes, and other organs (including the bone marrow, lungs, pleura, kidneys, adrenal glands, stomach, and ileum) [2]. Disseminated peliosis involving several organs has been described [1].

Although peliosis has been associated with malignant tumors, drugs, toxins and infections, the etiology is unknown. As the causes are varied, the demographics will reflect the underlying cause [3].

We report a case of peliosis of the liver occurring in a patient previously operated for a retroperitoneal paraganglioma and splenectomy about 3 years back. When the lesions were detected in liver by CT 3 years after surgery, they were supposed to be metastases and FNAC was performed.

Case report
A 26-yr-old man was admitted to the hospital with a large lump in left upper abdomen. Ultrasound and CT revealed a large (12 x 10 x 9 cm) retroperitoneal lesion between left kidney and aorta. Ultrasound also revealed multiple echogenic lesions within the spleen. The splenic lesions were hypodense in non-contrast CT scan and were heterogeneously enhancing after giving IV contrast. Size of the splenic lesions were variable, largest one measuring 6 x 5 cm.

Figure 1: Sonographic study shows multiple hypoechoic lesions of variable sizes disseminated throughout hepatic parenchyma.
Later, CT scan of abdomen with triphasic contrast was performed. CT scan showed enlarged liver (22 cm in craniocaudal extension). Left lobe of liver was also enlarged, occupying the splenic region in left sub-diaphragmatic region. There were numerous disseminated ill-defined hypodense lesions of variable sizes (few millimeters to 4.8 cm) throughout hepatic parenchyma in unenhanced scans (Figure 2). After IV contrast, there was strong rim enhancement of the lesions in arterial phase. Some small sized lesions also showed strong homogenous enhancement pattern in arterial phase (Figure 3). In portal venous phase, there was incomplete washout of the contrast (Figure 4). And in venous phase, most of the lesions showed complete contrast washout and some incomplete (Figure 5).

**Figure 2:** Axial unenhanced CT images of upper abdomen

Numerous hypodense lesions of variable sizes noted in the liver parenchyma.
Enlargement of left lobe of liver is noted, occupying splenic region.

**Figure 3:** Axial and coronal CT images in arterial phase of contrast enhancement

Some lesions showing strong rim enhancement and some small-sized lesions showing strong homogeneous enhancement patterns.

**Figure 4:** Axial and coronal CT images in portal venous phase of contrast enhancement

Lesions showed incomplete washout of contrast.

**Figure 5:** Axial CT image in venous phase of contrast enhancement

Most of the lesions showed complete washout of contrast and some lesions showed incomplete washout of contrast.

Among the hematological and biochemical reports, hemoglobin, WBC count, platelets, ESR, serum bilirubin, SGPT, SGOT, alkaline phosphatase and AFP (alpha fetoprotein) were within normal limits as shown in (Table 1).

<table>
<thead>
<tr>
<th>Parameters</th>
<th>Values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>11.50 g/dl</td>
</tr>
<tr>
<td>White blood cell count</td>
<td>8200/ml</td>
</tr>
<tr>
<td>Platelets</td>
<td>209000/ml</td>
</tr>
<tr>
<td>ESR</td>
<td>15 mm in 1st hour</td>
</tr>
<tr>
<td>Serum bilirubin (total)</td>
<td>0.8 mg/dl</td>
</tr>
<tr>
<td>SGPT/ALT (Alanine transaminase)</td>
<td>31 U/L</td>
</tr>
<tr>
<td>SGOT/AST (Aspartate aminotransferase)</td>
<td>28 U/L</td>
</tr>
<tr>
<td>Alkaline phosphatase</td>
<td>120 U/L</td>
</tr>
<tr>
<td>Alpha fetoprotein</td>
<td>4.2 ng/ml</td>
</tr>
</tbody>
</table>

Then, FNAC (fine needle aspiration cytology) from hepatic lesion under ultrasound guidance was performed. The cytosmears showed cords and trabecula of regular hepatocytes and blood. No malignant cell was seen in the smears examined.

**Differential diagnosis**

Differential diagnosis for hypoechoic lesions in ultrasound involving hepatic parenchyma, and which appeared peripherally rim enhancing hypodense lesions in CT scan are hypervascular metastases, hemangioma and hepatocellular carcinoma.

Although some hypervascular metastases with fibrotic change can show mild hyperattenuation in the delayed phase, hypervascular metastases are usually totally hypo- or iso-attenuating in the delayed phase of contrast enhancement because of the rapid washout of contrast material. Thus, in general, peliotic lesions are rarely confused with hypervascular metastases [2].

The typical enhancement pattern of hemangiomas (i.e., peripheral ring or globular enhancement with centripetal progression) is opposite of peliosis hepatitis, and therefore differential diagnosis can be achieved in most patients. In addition, hemangiomas may be rather large lesions with a mass effect on the hepatic vessels, whereas
peliotic lesions usually show no mass effect on hepatic vessels [2].

Hepatocellular carcinoma is usually hyperattenuating in the arterial phase with rapid washout in the portal venous phase and iso- or hypoattenuation in the delayed phase [2]. This patient had history of resected retroperitoneal paraganglioma 3 years back and no other primary malignancy. Case reports of peliosis of liver and spleen associated with paraganglioma have been published before [1].

And Ultrasound guided FNAC from hepatic lesion showed cords and trabecula of regular hepatocytes and blood. And no malignant cell was seen in the smears examined 3 years back, the splenic lesions that were homogeneously enhancing hypodense in CT and confirmed as benign vascular lesions composed of endothelial lined blood vessels histologically. These lesions also correlate with the findings of peliotic lesions. So, those splenic lesions mimicking metastases could be peliotic lesions. These all findings favor this patients case with hepatic peliosis associated with paraganglioma.

**Discussion**

Peliosis means “purpura” or “extravasated blood” and refers to the macroscopic appearance of blood-filled cystic cavities [1]. Histologically, peliosis is characterized by multiple mottled blood-filled cyst-like spaces. These vary in size from few millimeters to several centimeters in diameter. The cysts may show thrombosis of varying degree [3]. Peliosis was initially described in the liver. Involvement of the spleen is generally seen in connection with hepatic peliosis. However, cases in which peliosis involves the spleen alone have been published [4]. Peliosis involving multiple organs of the same patient has been documented [2].

The cause of peliosis hepatis can be related to drugs (including anabolic steroids, oral contraceptives, corticosteroids, tamoxifen, diethylstilbestrol, azathioprine, 6-thioguanine, 6-mercaptopurine, and methotrexate); toxins (polyvinyl chloride, arsenic, and thorium oxide); chronic wasting diseases (e.g., tuberculosis, leprosy, and various malignancies, particularly hepatocellular carcinoma); and infection in AIDS (so-called bacillary peliosis caused by Bartonella henselae and Bartonella quintana). In addition, several other conditions are described as associated with peliosis hepatis, including sprue, diabetes mellitus, necrotizing vasculitis, and hematologic disorders. Moreover, peliosis hepatis may develop after renal or cardiac transplantation. In 20–50% of patients, no associated condition is identified. Cases with sudden rupture of hepatic & splenic peliosis causing intrahepatic, intrasplenic and intraarterial hemorrhages have been published before [4,5].

On imaging studies, various patterns are characteristic. On unenhanced CT, peliotic lesions appear as iso- or hypodense. On contrast-enhanced CT, peliotic lesions can be hypoattenuating to liver parenchyma in the early acquisitions and tend to become progressively isodense with time. In addition, some lesions can also show areas of increased attenuation. Notably, larger cavities communicating with sinusoids display the same attenuation of blood vessels, whereas thrombosed cavities have the same appearance as nonenhancing nodules. More often, during the arterial phase of contrast enhancement, peliotic lesions typically show early globular enhancement (vessel-like enhancement) and multiple small accumulations of contrast material in the center of the lesions (the so-called target sign). During the portal venous phase, a centrifugal progression of enhancement without a mass effect on hepatic vessels is usually observed, however, a centripetal progression of enhancement can also be seen. On the delayed phase, late diffuse homogeneous hyperattenuation can also be seen in some cases of hepatic peliosis (because of the lack of hemorrhagic parenchymal necrosis). This accumulation of contrast material in the delayed phase can be useful in the differential diagnosis with other focal hepatic lesions that do not show blood pooling. In some instances, small (< 2 cm) peliotic lesions may also show hyperattenuation on both arterial and portal venous phase images [2,6].

**Summary**

Hepatic peliosis is a rare benign tumor. Peliosis can also occur in spleen, lymph nodes, and other organs (including the bone marrow, lungs, pleura, kidneys, adrenal glands, stomach, and ileum). On imaging, hepatic peliosis can resemble hepatocellular carcinoma, hypervascular hepatic metastases and hemangioma. So, hepatic peliosis should be considered in cases presenting with multiple unknown liver tumors that are revealed as atypical on radiological images [2,6-9].

**References**


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