LIVER SCAN IN BUDD-CHIARI SYNDROME

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Seventeen liver scans were performed in seven patients with occlusion of hepatic veins: the Budd–Chiari syndrome. When some, but not all liver veins were occluded, markedly diminished uptake over the affected segments was usually seen (absence of uptake may occur in acute infarction and in chronic cases). When all major liver veins are occluded, markedly diminished uptake is seen over the peripheral parts of the right and left lobes with a triangular midline area of normal or excessive activity. The latter effect is probably caused by uptake in segments surrounding the inferior vena cava (frequently the caudate lobe) that have direct venous drainage. This liver-scan appearance is characteristic enough to warrant consideration of the Budd–Chiari syndrome as the first diagnosis. Good correlation existed between selective venography and liver-scan findings. The usefulness of liver scans in the followup after portocaval shunting is illustrated in one patient.

Symptomatic occlusion of hepatic veins, the Budd–Chiari syndrome (BCS), has been regarded as an infrequently diagnosed condition (*1*). Recent evidence suggests that its incidence may be on the increase, particularly in women taking oral contraceptives (*2,3*). Diagnosis of BCS in life is usually made by hepatic venography or liver biopsy (*3,4*). More recent reports of BCS have included descriptions of liver scans, but their diagnostic significance has not been adequately considered (*3–8*).

We have studied 17 liver scans in seven patients with confirmed BCS over the last 7 years and we have reviewed the current literature in an attempt to assess the usefulness of hepatic scintigraphy in the diagnosis and followup of this syndrome.

CASE REPORTS

Case 1. A 36-year-old woman had been taking oral contraceptives. Hepatic venogram showed occlusion of all major liver veins. A liver scan with ^{198}Au-colloid (July 22, 1968), only showed activity in the superior medial segments of the right lobe. There was uptake in the bones; the spleen measured 14 × 6 cm. A side-to-side portocaval shunt was performed, but the patient died 2 days later.

Case 2. A 49-year-old man presented with polycythemia rubra vera. Hepatic venography confirmed occlusion of all hepatic veins. A liver scan with ^{198}Au-colloid showed absent activity over the periphery of both hepatic lobes with much activity in the area between the two lobes. The patient failed to respond to heparin therapy, and a repeat liver scan showed further reduction in uptake. He died 3 weeks later on Sept. 20, 1968.

Case 3. A 36-year-old woman had been taking oral contraceptives. Laparotomy with biopsy confirmed occlusion of main hepatic veins. Liver scan with ^{99m}Tc-sulfur colloid (May 6, 1970), showed uptake mainly in the midline, probably in the caudate lobe and the superior median segment of the right lobe; the spleen measured 14 × 8 cm. She died in hepatic coma 2 weeks later.

Case 4. A 38-year-old woman presented with polycythemia rubra vera. A ^{99m}Tc-sulfur colloid liver scan showed markedly diminished activity at the periphery of both lobes with uptake in the midline region, probably the caudate lobe and the medial parts of the right lobe. She failed to respond to anticoagulants. Budd–Chiari syndrome was confirmed at autopsy.

Case 5. A 25-year-old woman had been taking oral contraceptives. Selective venography showed occlusion of all major hepatic veins. A ^{99m}Tc-sulfur colloid liver scan (Fig. 1), taken on Aug. 5, 1973, showed markedly diminished uptake in the right lobe and the lower segments of the left lobe with a "heart-shaped" area of normal or excessive uptake in the

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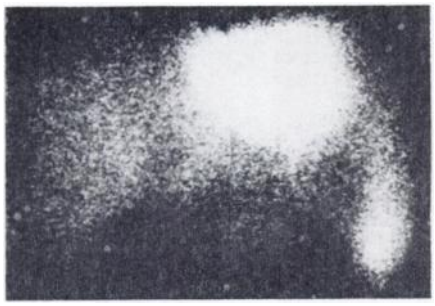

FIG. 1. Anterior scintigram of liver in Case 5 with BCS shows "heart-shaped" area of relatively increased uptake in midline and left lobe region.

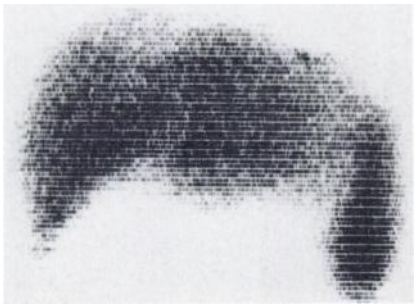

FIG. 2. Scan in Case 5, 2 months after mesocaval anastomosis, shows improvement of liver uptake over Fig. 1.

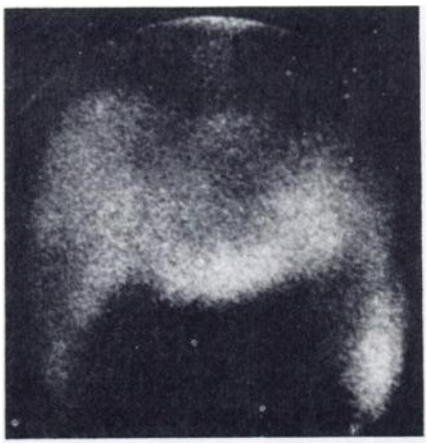

FIG. 3. Repeat scan in Case 5, 14 months after mesocaval anastomosis, shows slightly uneven uptake in most parts of liver, except midline areas which now show markedly diminished activity.

midline; the spleen measured 17 × 7 cm. When her condition worsened, a repeat scan showed uptake in the ribs and spine. A ^{99m}Tc-sulfur colloid scan (Fig. 2), 2 months after side-to-side mesocaval anastomosis, showed improvement of uptake over the periphery of the liver, and the midline region of excessive uptake was no longer present. For about 1 year she was in good health and a liver biopsy showed improvement, but as clinical deterioration reappeared her liver scan (Fig. 3) showed diminished and uneven uptake in both lobes and no activity in the midline area roughly corresponding to the "heart-shaped" hot area in Fig. 1. She died 4 months later on June 18, 1975. At autopsy the mesocaval shunt was thrombosed and both lobes of the liver showed nodules of regeneration, fibrous replacement, and paracentral necrosis.

Case 6. Selective venography on a 43-year-old woman showed occlusion of the right and median veins and partial occlusion of the left hepatic vein. Liver biopsy showed dilatation of sinusoids and central veins. Arteriogram showed atrophy of the right lobe and enlargement of the left and caudate hepatic lobes. A ^{99m}Tc-sulfur colloid scan 12 days after a splenorenal shunt showed no activity over the right lobe and normal uptake over the enlarged left lobe; the spleen measured 18 × 7.5 cm. A repeat scan 1 year later showed clearing of uptake from the ribs and spine. She was still in good health 4 years later.

Case 7. Liver biopsy on a 45-year-old woman suggested BCS, and selective venography showed all but the median hepatic vein to be occluded. A ^{99m}Tc-sulfur colloid scan (Fig. 4), taken on July 4, 1973, showed liver enlargement, with diminished uptake over the periphery of both lobes with normal uptake in the midline area. Scanning with ^{131}I-rose bengal showed an activity distribution similar to that of the ^{99m}Tc-sulfur colloid scan. The patient remains in fairly good health on low sodium diet.

DISCUSSION

The blood leaves the liver by three main veins to enter the inferior vena cava. The left hepatic vein drains most of the left lobe, the middle (or median) vein carries blood from the central parts of the liver

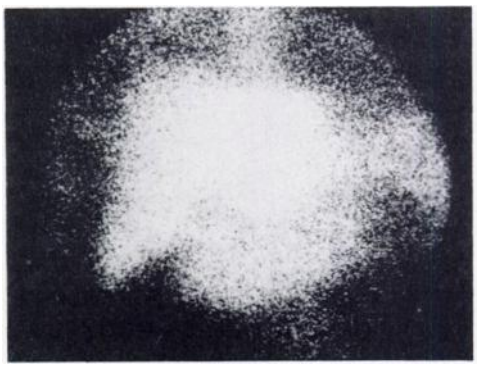

FIG. 4. Anterior liver scintigram in Case 7, in which only the median hepatic vein is patent, shows diminished uptake at periphery of right and left lobes and increased uptake near midline.

including the quadrate lobe, and the right vein drains the right lobe. The left and middle veins often enter the inferior vena cava by a common trunk. The caudate lobe is drained by branches of the left and right hepatic veins and by several small veins opening directly into the inferior vena cava; the same small veins also drain parts of the posterior right lobe (*9*).

When more than one main vein is occluded, BCS results. The syndrome comprises hepatomegaly, abdominal pain, ascites, and hepatic histology showing centrizonal sinusoidal distension, hemorrhage, and necrosis. Mortality is high in acute illness, but chronic cases may survive for months, and rarely years, showing varying degrees of liver failure and complications related to portal hypertension. The caudate lobe often remains intact and shows hypertrophy (*5,10*). Budd–Chiari syndrome is associated with polycythemia, use of oral contraceptives, malignancy, trauma, and congenital abnormalities of the hepatic veins or inferior vena cava. Treatment is directed toward the underlying abnormality, and surgical correction or bypass is attempted when technically feasible (*3,10*).

In our series Cases 1–5 showed complete Budd–Chiari syndrome, i.e., all three main hepatic veins were occluded. Liver scans in all five showed hepatomegaly, markedly diminished uptake over the left and right hepatic lobes involving the right more than the left, and a persisting midline region of normal or increased uptake. Excellent correlation existed between the venous obstruction shown venographically and the markedly diminished uptake seen in liver scans over the right and left lobes in Cases 1–5 and 7. Case 6, with long-standing BCS, differed a little from this pattern in that uptake was relatively absent over the occluded lobe. Cases 1–6 also showed obstruction of the middle hepatic vein, but only three clearly showed diminished uptake over the lateral parts of the quadrate lobe (the area between the gallbladder and the midline in the anterior view) because of the abnormally increased uptake in the midline region. This triangular or heart-shaped area of increased uptake is the most striking and constant scintigraphic feature in patients with complete BCS; it may be seen also to a lesser extent in incomplete BCS [Case 7 (Fig. 4), but not Case 6]. Sherlock (*10*), Caroli (*5*), and Tavil et al (*8*) have published scintigrams of this unusual appearance in Budd–Chiari syndrome; they believe that the caudate lobe is relatively spared in this condition and may take up radiocolloid normally or excessively. In posterior, lateral, and oblique views in four of our patients, this area of increased uptake appeared to lie in the postero-inferior midline region of the liver and, therefore, probably in the caudate lobe. Part of this uptake was found to be in the area of the superior anteromedial segments of the right lobe in three patients and in the upper part of the left lobe in one. We have concluded, therefore, that this midline activity is located in the small segments of the liver, sometimes in the caudate lobe, surrounding the inferior vena cava. These segments have probably retained or developed venous drainage into the inferior vena cava and, because of the increased portal pressure in BCS, received excessive amounts of portal blood, leading to their hypertrophy. These functioning liver segments may account for survival for months or years in chronic cases of BCS. The segments may become palpable clinically, may cause narrowing of the inferior vena cava, may show up on venocavography (Cases 5 and 7), and may mimic a tumor (*5*). A liver scan showing increased uptake over the palpable liver mass clearly points to hypertrophy as opposed to a tumor.

This liver-scan appearance is not seen exclusively in BCS. It has been reported in one of constrictive pericarditis (*7*) and one case of postnecrotic cirrhosis (*11*). We have seen it in two scans in a series of 200 patients with cirrhosis of the liver (patent hepatic veins were confirmed by selective venography in both of these patients). Excessive uptake in the midline with markedly diminished activity at the periphery may be the first clue that BCS is present, and since this condition requires urgent management different from that for cirrhosis, Budd–Chiari syndrome should be considered as the first probable diagnosis when such a scan is encountered.

Cases 6 and 7, with incomplete BCS, showed absent and diminished uptake, respectively, in the scans over the segments with occluded veins. Some cases of chronic incomplete BCS with atrophy of the affected lobe (Case 6) and some cases of acute incomplete BCS with infarction (*6*) show no uptake in the liver scan over the obstructed segments. The differential diagnosis of liver scans in patients with incomplete BCS should include partial hepatectomy, radiation injury, fortuitous segmental involvement by diffuse or focal liver disease, and (very rare) hepatic artery occlusion (*12*).

Two months after a portocaval shunt procedure, Case 5 showed clinical recovery and the repeat liver biopsy showed less necrosis and a larger number of normal liver cells. The hepatic scintigram showed fairly normal uptake over all parts of the liver (Fig. 2). This probably results from the better perfusion of the obstructed segments after the shunt procedure: hepatic artery flow to the affected lobules increases as the portal pressure falls, with the portal venous branches acting as the outflow route with reversed

flow. The marked drop in portal pressure causes underperfusion of the hypertrophied midline segments so that at 2 months (Fig. 2) the uptake there is similar to the rest of the liver. At 14 months this midline area shows no uptake (Fig. 3), probably because of its atrophy.

As in other conditions with deterioration of liver function, radiocolloid uptake appears in the ribs, spine, and lung (Cases 5 and 7); this uptake recedes when liver function improves.

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