Ultrasound scanning and ^{99m}Tc sulphur colloid scintigraphy in diagnosis of Budd-Chiari syndrome

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SUMMARY Ultrasound scanning and ^{99m}Tc sulphur colloid scintigraphy are widely used in the diagnosis of the Budd-Chiari syndrome and have been compared at the time of presentation in 18 patients in whom the diagnosis was subsequently confirmed by histology and hepatic venography. Ultrasound was diagnostic in 16 (87%). The findings seen most often included hepatic vein abnormalities, caudate lobe hypertrophy with decreased reflectivity and compression of the inferior vena cava. Additional information not shown by scintigraphy included intracaval tumour, or thrombosis, and concomitant portal vein thrombosis. Although scintigraphic abnormalities were present in all patients, only in three (17%) was the 'classical' appearance of increased uptake and/or enlargement of the caudate lobe present. In one patient with non-specific abnormalities on ultrasound, scintigraphy gave a positive diagnosis and it is in such cases that scintigraphy should continue to be used.

The diagnosis of the Budd-Chiari syndrome is at times difficult because of the variability of presentation which may mimic cirrhosis, intra-abdominal malignancy, tuberculous peritonitis or even acute hepatitis.¹ Indeed, since 1972, we have had nine patients referred to this Unit in whom hepatic vein thrombosis had been unsuspected and an exploratory laparotomy had been carried out. In each instance this was followed by serious postoperative complications including one death. Analysis of these cases indicated inadequate preoperative investigation, in particular, by scanning techniques. The characteristic appearances of the Budd-Chiari syndrome with enhanced caudate lobe uptake on colloid scintigraphy are well described² and the patency or otherwise of the hepatic veins and inferior vena cava can be established by grey scale ultrasound scanning.³⁻⁵ As little has been published on the diagnostic accuracy of the two techniques, we have studied their use in 18 patients with the Budd-Chiari syndrome of varying aetiology.

Methods

PATIENTS

The 18 patients were aged 13-46 years and, in all

cases, the diagnosis was confirmed by hepatic venography and liver histology. Aetiological factors in two cases included polycythaemia rubra vera with single instances of essential thrombocythaemia, paroxysmal nocturnal haemoglobinuria, right atrial tumour, hepatic abscesses with hepatic vein compression, the use of the contraceptive pill and thrombosis of the inferior vena cava in a liver transplant recipient. No specific cause could be identified in the remaining 10. The presentation in 17 was with an acute illness of eight weeks or less duration. The remaining patient was being investigated three years after the first symptoms with an apparent diagnosis of cryptogenic cirrhosis. Both types of scan were reported on by different members of the respective departments before the diagnosis had been proven by biopsy and hepatic venography.

Ultrasonography was carried out using a real time machine (Diasonics DRF1) and ^{99m}Tc sulphur colloid scintigraphy using either an IGE or an Elscint CE-1 gamma camera. The distribution of uptake within the liver outline and presence of extrahepatic uptake by the spine and spleen were noted. Splenomegaly was considered significant when the long axis exceeded 13 cm or when splenic width exceeded 7 cm.

Results (Table)

Ultrasound examination was indicative of the Budd-

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Table Features on ultrasound scanning and ^{99m}Tc sulphur colloid scintigraphy of 18 patients with the Budd-Chiari syndrome

Patient	Age	Sex	Primary condition	Ultrasound scan				Colloid scintigraphy			
				Caudate lobe hypertrophy	Decrease in caudate lobe reflectivity	Absent normal hepatic veins	Compression of inferior vena cava	Additional findings	Central localisation	Uptake by right lobe reduced	Extrahepatic and splenic uptake
1	36	F	Idiopathic	+	+	+	+	Ascites Patent PV	-	+	+
2	46	M	Hepatic abscesses	-	-	-	-	Ascites Multiple hepatic abscesses	-	Filling defects	+
3	26	F	Polycythaemia rubra vera	+	+	+	+	Ascites	-	Patchy appearance with 'hot spots'	+
4	39	M	Idiopathic	-	-	+	-	Ascites	-	+	+
5	23	F	Idiopathic	+	+	+	+	Ascites Patent PV	-	+	-
6	23	F	Idiopathic	+	-	+	Thrombosed	Ascites Patent PV	-	+	+
7	29	F	Polycythaemia rubra vera	+	-	-	-	Ascites Patent PV	-	+	+
8	33	F	Idiopathic	+	+	-	+	Ascites Patent PV	-	+	+
9	37	M	Paroxysmal nocturnal haemoglobinuria	+	-	+	-	Ascites Blocked PV	-	+	+
10	21	F	Right atrial leiomyosarcoma	+	-	+	Intraluminal tumour	Ascites Patent PV	-	Patchy uptake with 'hot spots'	-
11	41	M	Idiopathic	+	+	+	+	Ascites Patent PV	+	+	+
12	38	M	Idiopathic	-	-	-	-	Ascites Patent PV	-	+	+
13	26	F	Contraceptive pill	+	+	+	+	Ascites Patent PV	+	+	+
14	24	F	Idiopathic	-	-	-	+	Ascites	+	+	+
15	20	M	Idiopathic	-	-	+	-	Ascites	-	+	+
16	44	F	Idiopathic	+	+	+	+	Ascites Patent PV	-	+	+
17	35	M	Post liver transplant	-	-	+	Thrombosed	Ascites	-	+	+
18	41	F	Essential thrombocythaemia	+	+	+	-	Ascites Blocked PV	-	+	+

Chiari syndrome with failure of visualisation of the hepatic veins in 13 (72%) of the 18 cases. In three patients this finding was the only abnormality. Caudate lobe hypertrophy was present in 12 (67%) with decreased caudate lobe reflectivity in eight of them (Fig. 1). In eight (44%) the inferior vena cava was compressed as a consequence of the hypertrophy (Fig. 2). Intraluminal tumour or thrombosis

of the inferior vena cava throughout its entire length was present in three (17%). In 12 of the 18 cases in whom the portal vein was specifically identified, patency was demonstrated in 10 and blockage by thrombosis in two (Fig. 3). In one patient with the Budd-Chiari syndrome and multiple hepatic abscesses, clear evidence of hepatic vein thrombosis was not obtained but, one year later, a repeat scan

Fig. 1

Fig. 2

Fig. 3

Fig. 1 In the Budd-Chiari syndrome, the caudate lobe is enlarged (arrowed) and of decreased reflectivity in comparison with the adjacent parenchyma within the left and right lobe.

Fig. 2 In the Budd-Chiari syndrome, hypertrophy of the caudate lobe results in compression of the inferior vena cava (arrowed).

Fig. 3 In this patient with the Budd-Chiari syndrome, portal vein thrombosis is present with demonstration of collateral channels (arrowed) and no normal main portal vein.

showed narrowing of the hepatic veins proximal to their junctions with the inferior vena cava, this probably being the site at which obstruction occurred. An additional feature observed in all of the 18 patients was the presence of gross ascites.

On colloid scintigraphy, the 'classical' appearance of relative increase in uptake by the caudate lobe in comparison with the right and left lobes was found in only three (17%) patients (Fig. 4). In 14 of the remaining 15 patients, the findings were abnormal but non-specific with reduced uptake by the right lobe in 12 (67%) and non-uniform uptake giving the appearance of 'hot spots' in two (11%) (Fig. 5). Increased skeletal and splenic uptake was present in 16 (89%) and in 13 of them the spleen was also enlarged.

When the value of ultrasound and scintigraphy as primary diagnostic investigations was compared, the former was consistently better, with 16 (87%) reported as 'diagnostic' but only three (17%) with scintigraphy. In one patient with a 'diagnostic' scintiscan, there was no abnormality on ultrasound scanning other than ascites.

Discussion

The findings show the marked superiority of ultrasound scanning over scintigraphy in the diagnosis of the Budd-Chiari syndrome. Reporting of the scans was carried out by different members of the respective departments and therefore did not reflect the special expertise of just one member. The low level of diagnostic accuracy for scintigraphy was surprising in view of the findings in the report of Tavill *et al.*,² in which 15 of 19 patients showed central localisation of colloid to be a predominant feature together with reduced uptake in the right lobe, while in the other eight, it was present to a lesser degree in association with patchy uptake by the rest of the liver. 'Classical' appearances on scintigraphy were also reported by us in 16 (46%) of a series of 35 patients studied retrospectively.⁵ Most of these had long preceding histories and in those with a duration of symptoms of less than eight weeks the only abnormality detected was some enlargement of the right lobe with normal uptake of colloid. In the present series, most of the patients had histories less

Fig. 4 'Classical' appearance of the Budd-Chiari syndrome showing reduced hepatic uptake of isotope except for the caudate lobe. The spleen is enlarged.

Fig. 5 In this patient with the Budd-Chiari syndrome, the anterior view shows focal areas of apparently normal uptake of sulphur colloid. The spleen is moderately enlarged.

than eight weeks in duration and the scans were reported on before the diagnosis had been finally established. Consequently, increased midline uptake was more likely to be attributed to skeletal rather than caudate lobe intake in the present series. Of interest was the finding, not previously described, of multiple 'hot spots' in two of the cases. These appearances might represent focal preservation of the blood supply or possibly nodular regeneration which, in some patients with the Budd-Chiari syndrome, may be quite marked.

Ultrasound scanning was of greater diagnostic value not only in giving information regarding the size and reflectivity of the caudate lobe but also the presence and, occasionally, the cause of the hepatic vein obstruction—that is, thrombosis or tumour within the inferior vena cava and whether the portal vein was also thrombosed. The latter is a not infrequent complication of hepatic vein thrombosis⁵ and is a contraindication for decompressive shunt surgery. In the small number of cases in whom ultrasound scanning did not provide a definite answer, colloid scintigraphy is worth carrying out as

this led to a positive diagnosis in one of the patients in the present series.

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