Liver Scintiscan in Budd Chiari Syndrome

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Abstract

Budd Chiari Syndrome is a rare disease and can be difficult to diagnose. Most of the investigations to diagnose this disease are invasive. Liver scintiscanning is not only non-invasive, it also shows a typical pattern in this condition. Good correlation exists between Liver scanning and other procedures. A case of Budd Chiari Syndrome with typical clinical and scan findings is described (JPMA 34: 375, 1984).

Introduction

Symptomatic occlusion of hepatic veins, Budd Chiari Syndrome, is an infrequently diagnosed condition. Clinically it presents with abdominal pain and rapidly accumulating ascites. The commonest causes are polycythaemia rubra vera, nocturnal haemoglobinuria, hepatic webs, generalized thrombotic tendency, oral contraceptives and secondary hepatic involvement from hypernephromas. The cause is not clear in 50-70% of the cases. The investigations used to diagnose this condition include hepatic scintigraphy, inferior venacavography, hepatic vphography, direct hepatography, splenoportagraphy and liver biopsy. A case with typical clinical and liver scan findings is reported here.

Case Report

A 45 year old male presented with rapidly accumulating ascites. There was no history of previous jaundice, melaena or haemetemesis. On examination the patient had ascites, the abdomen was tense with fluid. No mass could be felt and there was no evidence of chronic liver dysfunction like spider naevi palmer erythema, gynaecomastia and testicular atrophy. The chest radiograph showed a normal heart size and clear lung fields. There was no calcification in the pericardium. Ascitic fluid was haemorrhagic out there was no evidence of malignant cells. Scintiscan showed an enlarged, low lying medially displaced liver with a triangular midline area of increased uptake which was posteriorly oriented in the right lateral view. Spleen was not enlarged but there was increased uptake by the spine and ribs (Fig 1 and 2).
A diagnosis of hepatic vein obstruction was made and the patient was advised further investigations. Unfortunately the patient did not return and could not be followed up.
Discussion

Hepatic venography and hepatic biopsy with typical findings of centrizonal venous congestion, haemorrhage and necrosis in the presence of normal right atrial and jugular venous pressures correlates well with radiocolloid scanning. Both hepatic venography and biopsy are relatively invasive procedures and not easily available in our setting. Hepatic scintiscanning on the other hand though technologically a more sophisticated investigation, is easily available in or near almost all large medical centres of the country and is almost totally non-invasive.

The typical findings of radionuclide imaging are diminished peripheral activity in the right and left lobe with a triangular midline area of normal or increased uptake\(^3\). Posteriorly oriented as seen in the right lateral view\(^2\), this central localization may be the predominant feature or it may be accompanied by a patchy uptake by the rest of the liver. Normally the area of maximum uptake is to the right of the midline. This scan finding is suggestive enough to warrant a first diagnosis of Budd Chiari Syndrome\(^3\).

The dimple sign has also been mentioned, appearing as an exaggeration of the normally present defect in the mid portion of the superior border of the liver due to the imprint of the two hepatic veins\(^4\), a defect in this area is usually seen due to the cardiac imprint and the presence of this finding in a previously normal liver scan when associated with a normal cardiac shadow on the chest radiograph and central localization may increase the specificity of the scan\(^4\). The reason for this localization has been attributed to the caudate lobe of the liver which is not totally dependent upon the hepatic veins for drainage and has direct drainage channels into the inferior vena cava. This means more efficient flow to the lobe is maintained while the rest of the liver flow is compromised in hepatic vein obstruction. This ‘sparing’ along with the hypertrophy which this lobe undergoes in this pathology, as evidenced by inferirbr vénacavography and autopsy\(^2\) is responsible for the increased uptake seen in the scintiscan. The ‘hot area’ is not an exclusive finding seen only in Budd Chiari Syndrome. Other pathologies described to give this appearance include haemangiom\(^5\), focal nodular hyperplasia and hepatic adenoma\(^6\) and abscess\(^7\) though these hot areas are not confined to any location. A picture similar to Budd Chiari Syndrome is seen in constrictive pericarditis and advanced cirrhosis but the distribution of colloid is different being largely perihilar without a prominent midline uptake and with more irregularity throughout the liver and with a more intense splenic uptake\(^2\).

In our patient the diagnosis of hepatic vein obstruction was suggested by the history, clinical examination, laboratory and scintiscan findings. Liver scanning is easily available in this country and the diagnostic yield of this infrequently diagnosed condition might improve with greater awareness of scan findings in this pathology. This is especially relevant because treatment of Budd Chiari Syndrome is different from cirrhosis with the option of surgery being a very important consideration.

References
