Cholescintigraphy in Dubin-Johnson syndrome

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Introduction

Hereditary or familial hiperbilirubinemia comprises a group of syndromes (Dubin-Johnson's, Rotor's, hepatic storage disease) in which hyperbilirubinemia, predominantly unconjugated or conjugated, occurs as an isolated biochemical abnormality without evidence of either hepatocellular necrosis or cholestasis. We present a patient with Dubin-Johnson syndrome, one of the familial disorders associated with conjugated or mixed hyperbilirubinemia in the absence of cholestasis (1, 2, 3).

Case report

We present a 67-year-old male patient with Dubin-Johnson syndrome. The diagnosis was based on isolated hyperbilirubinemia and typical liver biopsy appearance. The diagnosis was missed 18 years ago when the patient had unnecessary laparotomy because of suspected cholelithiasis as he had a negative oral cholecystogram, which is a typical finding in this syndrome. Patient had repeated pain in the upper abdominal quadrant, radiating to the right scapular region and lasting for a few hours. Physical examination showed slight hepatosplenomegaly and mild tenderness below the right costal margin. Laboratory analyses were within normal range, including serum markers for hepatitis B and C, as well as antimitochondrial, antinuclear and smooth muscle antibodies. Urine analyses were normal, except positive reaction to bilirubin, while serum bilirubin was elevated with BSP retention in plasma. Ultrasound showed a stone in the gallbladder, while i.v. cholangiocholecystography and ERCP confirmed this finding and excluded choledocholithiasis. The distribution of the pigment in the liver was diffuse without predominant centrilobular distribution usually found in Dubin-Johnson syndrome. This heavy pigment burden might be an explanation for some of the discrepancies with classical findings in this syndrome: 1. high level of direct reacting serum bilirubin (79–89%), 2. high initial BSP retention (43.4% at 45 min) and 3. postponed accumulation of ^{99m}Tc-EHIDA in the liver. Hepatobilliary scintigraphy (^{99m}Tc diethyl IDA) showed delayed but uniform and complete uptake of the radiopharmaceutical by the hepatocytes, with very low and slow excretion in hepatobiliary system (Figures 1, 2).

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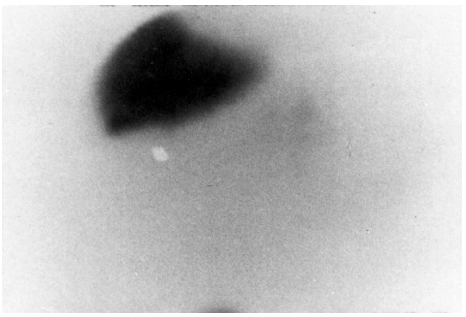

Figure 1. Sequential static scintigram (60 seconds duration) obtained 60 minutes after intravenous application of ^{99m}Tc diethyl iminodiacetate acid, showing uniform hepatic uptake, with very discrete excretion in hepatobilliary system (discrete gallbladder visualisation).

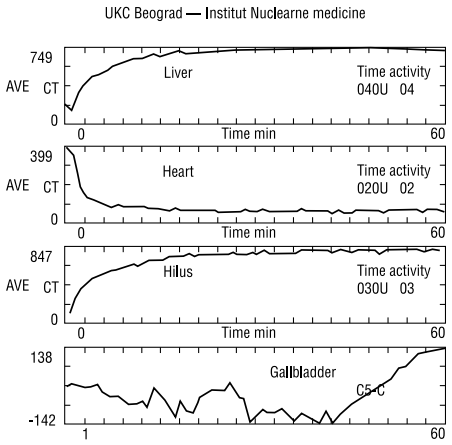

Figure 2. Liver time/activity curve shows delayed uptake of radiopharmaceutical (maximal parenchymal activity at 38th min; physiological values 10–15 min) and very slow excretion from the hepatocytes (5% for 60 min, physiological values 50% for 25 min); Time activity curve over hilus region shows delayed maximal activity (43rd min, physiologically at 25th min), and extremely slow outflow (3% for 60 min, physiologically 50% for 45 min). Accumulation of radioactivity in the gallbladder region is very discrete, at 40th minute (physiologically in 15th min), while intestinal activity is not visualised at all during 60 minutes.

Conclusion

Up to now, there has not been much data about hepatobiliary scintigraphy in those patients with inherited disorders characterised by conjugated hyperbilirubinemia. However, our results show certain discrepancies with the results of other authors. Accumulation of the radiopharmaceutical in the liver was considerably postponed in comparison with the results of Zimacek et al. (4) and Bar-Meir et al. (5) where liver uptake was homogeneous, rapid and intense. Biliary excretion was impaired in our patient similarly to those investigated by Zimacek et al. (4), while according to Bar-Meir et al (5), intestinal activity was registered within one hour. Similar to our results, Bujanover et al. (6), demonstrated intense prolonged homogeneous visualisation of the liver, together with delayed visualisation of the gallbladder and extrahepatic ducts, which has not been described in any other hyperbilirubinemic state. On the contrary, in patients with Rotor's syndrome, as well as in jaundiced patients with hepatobiliary disorders, liver, gallbladder and biliary tree are not visualised at all, without intestinal accumulation of radioactivity over 24h. Instead, visualisation of the kidneys was intense, indicating selective excretion of the radiopharmaceutical by this route (5, 7–9). We can conclude that hepatobiliary scintigraphy can be a useful method in the detection (4, 10, 11) and evaluation (6) of the patients suspected of having Dubin-Johnson's disease.

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