Yemen Journal of Medicine

https://yemenjmed.com

Article information

DOI: 10.63475/yjm.v4i1.0011

Article history:
Received: 29 March 2025
Accepted: 12 April 2025
Published: 28 May 2025

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How to cite this article
Wani I, Aga N, Bhat J , Hamdani N , Lone W, Younus U. A black liver in Dubin-Jonson syndrome: Does it matter?. Yemen J Med. 2025;4(1):200-201

Clinical Image

A black liver in Dubin-Johnson syndrome: Does it matter?

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A 28-year-old female presented with recurrent chronic abdominal pain. An abdominal ultrasound revealed cholelithiasis, characterized by two large stones and several smaller ones. Laboratory tests including liver function tests were within normal limits. A laparoscopic cholecystectomy revealed an enlarged dark black liver, with round margins suggestive of Dubin Johnson syndrome (Figure 1), while the Gallbladder was distended with multiple calculi inside. Successful laparoscopic cholecystectomy was done with a liver biopsy. Histopathology of the gallbladder revealed chronic cholecystitis. A liver biopsy confirmed the diagnosis of Dubin-Johnson syndrome. The follow-up was uneventful.

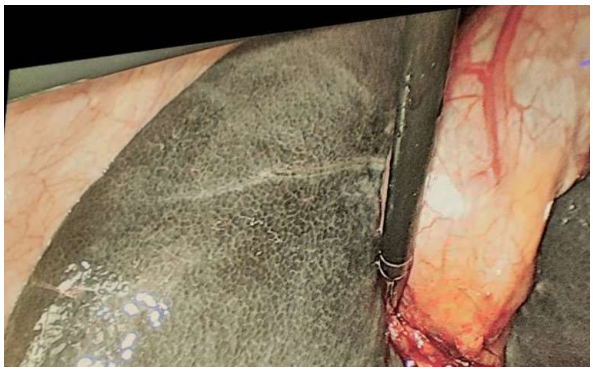

Figure 1
Laparoscopic images showing an enlarged deep black- liver having rounded edges with gallbladder at the liver bed

Dubin Johnson syndrome (DJS) is an autosomal recessive disorder caused by MRP2 [2] protein dysfunction. This originates from a mutation in the ABCC2 gene which provides instructions to produce a protein called MRP2. This MRP2 acts as a transporter protein for the excretion of conjugated bilirubin from liver cells into the biliary ducts.

This syndrome occurs among all races, and nationalities, and equally among both males and females, although it manifests earlier in men. DJS typically manifests in adolescence or young adulthood. Around 80 to 99% of people with DJS have jaundice, abnormal urinary color, conjugated hyperbilirubinemia, and biliary tract abnormality. Rarely, patients may present with abdominal pain, fatigue, liver enlargement, or dark urine.

Conjugated hyperbilirubinemia and a dark liver, whose color varies from black, dark green, and purple, to slate gray, are characteristic features of DJS.[3] The black liver is a unique feature of this disease and results from the deposition of coarse, dark, granular pigment in the centrilobular liver cells. Despite chronic hyperbilirubinemia, the black liver in DJS exhibits a normal liver parenchymal pattern.

Various radiological examinations, including oral cholecystography and hepatobiliary iminodiacetic acid scintigraphy (HIDA), have been performed to confirm DJS. [4] The gold standard for the diagnosis of DJS is liver biopsy. [5] Microscopically, brown pigment granules are detectable in the centrilobular hepatocytes. The definitive diagnosis to confirm DJS is molecular genetic testing of the ABCC2 gene.

Learning points

- In JDS, black liver, which ranges in color from black, dark green, and purple to slate grey, is attributed to the deposition of a dark granular pigment in the hepatic centrilobular cells.
- The condition is benign with a normal life expectancy and requires no special treatment.

CONSENT

Written informed consent was obtained from the patient for the publication of this case and the accompanying images.

AUTHORS'S CONTRIBUTION

All authors contributed to the completion of this work. The final manuscript was read and approved by all authors.

SOURCE OF FUNDING

None.

CONFLICT OF INTEREST

None.

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