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Dubin-Johnson syndrome as a laparoscopic finding

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Key words: Dubin-Johnson syndrome, conjugated hyperbilirubinemia.

CLINICAL CASE

We present the case of a 35-year-old woman with a history of polycystic ovary syndrome, treated with oral contraceptives who was under study for 9 months evolution pain in the right iliac fossa, associated with hyporexia and mild hyperbilirubinemia with a predominance of the conjugated fraction (total Bi 3.7 mg/dL, conjugated Bi 2.9 mg/dL). An abdominal computed tomography (CT) was performed showing homogeneous hepatosplenomegaly and adenopathies in both iliac chains, the biggest in the right external iliac chain of 1,6 x 3,6 cm. As a result, a laparoscopy was performed to biopsy the lymphadenopathies in the course of which it was observed a black liver surface (Fig. 1), which was biopsied. The anatomopathological study of the liver described brown pigment granules in centrilobular hepatocytes that stain irregularly with PAS, which were identified as lipomelanin, compatible with Dubin-Johnson Syndrome (DJS) (Fig. 2). The anatomopathological result of pelvic

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lymphadenopathies was non-necrotizing granulomatous lymphadenitis, whose study is being completed.

DISCUSSION

DJS is a benign autosomal recessive inherited disorder characterized by hyperbilirubinemia with a predominance of the conjugated fraction and an alteration in the metabolism of coproporphyrins (more urinary excretion of coproporphyrin I than III)[1]. It is caused by a mutation in a bilirubin transporter, called MRP2, which takes the conjugated bilirubin and introduces it into the bile duct[2]. Its prevalence has been estimated at less than 1 case per 100.000 inhabitants, being more prevalent in Iranian Jews[3]. The most common symptom is jaundice. The liver is characteristically black due to intracellular deposit of a melanin-like pigment. The main differential diagnosis is with Rotor Syndrome in which high urinary concentrations of both total coproporphyrin and coproporphyrin I are observed and the blackish pigment of the liver does not occur (Table 1). Due to its benign prognosis, no specific treatment is required.

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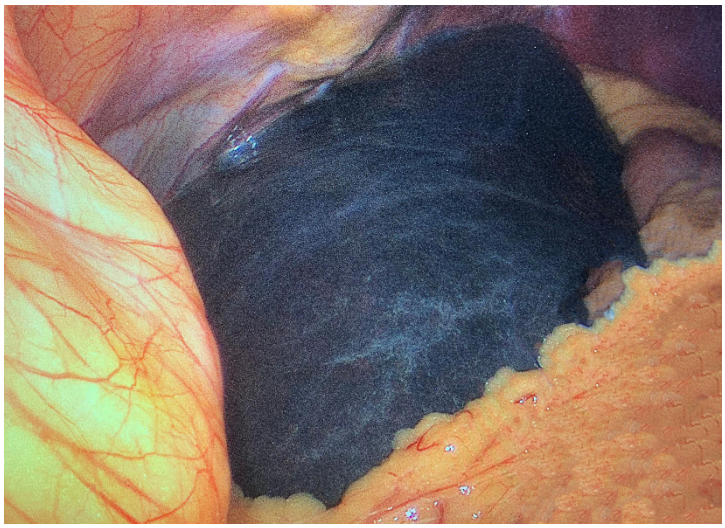

Figure 1. Laparoscopic image of the liver showing a regular surface and a blackish color of the parenchyma.

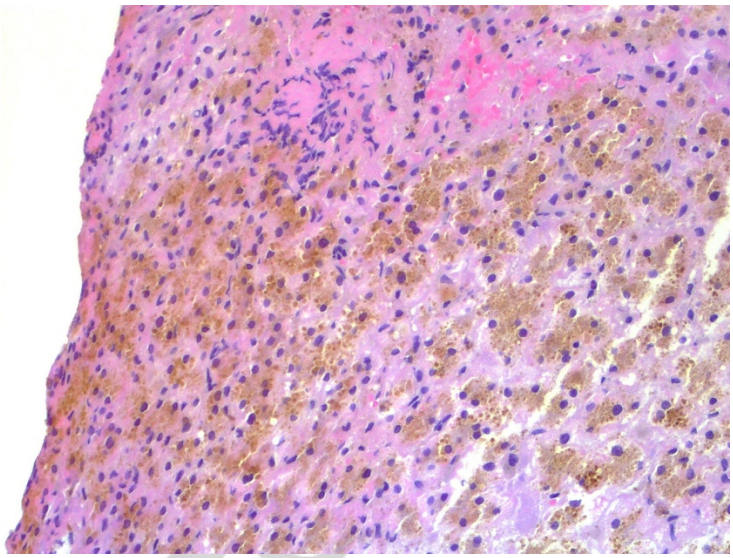

Figure 2. Liver biopsy: presence of brown intracytoplasmic pigment.

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	Dubin-Johnson	Rotor
Incidence	Rare	Exceptional
Inheritance	Autosomal recessive	
Bilirubin	2-5 mg/dl (60% conjugated)	
Symptoms	Asymptomatic jaundice	
Liver pathology	Presence of dark pigments in centrilobular areas	Normal
Urinary excretion of coproporphyrin	Total normal High isomer I	Total elevated High isomer I
Prognosis	Benign	Benign

Table 1. Differences between Dubin-Johnson Syndrome and Rotor Syndrome

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