Case Report: A rare case of a patient presenting with Dubin Johnson and Mirizzi syndrome

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Background

To gain a better understanding of the diseases and potential correlation we present the case of a patient with both Dubin Johnson syndrome and Mirizzi. To the best of our knowledge this is the first reported case of Dubin Johnsons syndrome and Mirizzi syndrome in the same patient.

Literature Review

Dubin Johnson syndrome is a rare hereditary autosomal recessive disorder of hyperbilirubinemia caused by a mutation in the MRP2/cMOAT transport proteins leading to an inadequate excretion of bilirubin.[1,2] This leads to a low grade increase in conjugated bilirubin resulting in hyperbilirubinemia and grossly black liver, which is usually incidentally found on routine tests or due to periods of major stress such as surgery, or infection.[3] It is diagnosed with elevated conjugated hyperbilirubinemia without other hepatic abnormalities, increased urinary coproporphyrin I compared to urinary coproporphyrin III, and centrilobular deposit of dark pigment on a liver biopsy.[1] It is a benign disease and does not require treatment as it does not lead to progression of fibrosis or cirrhosis.[3] Mirizzi syndrome is also a rare condition, which is due to either a single or multiple impacted gallstones in Hartman's pouch causing external compression and obstruction of the common bile duct or common hepatic duct.[4] Patients present similarly to cholecystitis but also have jaundice and can be confused for other obstructing conditions like choledocholithiasis or ascending cholangitis. It is difficult to diagnose preoperatively, but stent placement via ERCP prior to cholecystectomy can simplify the procedure and decrease the risk of intraoperative biliary injury.[5]

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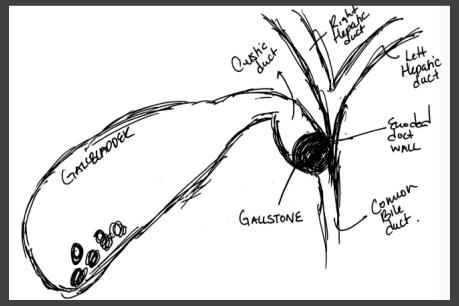
Case Presentation

We reported the case of a 53-year-old male who presented with suspected Mirizzi syndrome, with a past medical history of Dubin Johnson syndrome and alcohol abuse. Symptoms included right upper quadrant pain, elevated white blood cell count, and elevated total bilirubin. MRCP revealed a large gallstone in gallbladder neck secondary to gross inflammation of the gallbladder. Further imaging with ERCP and cholangiogram showed a patent common bile duct (CBD) and common hepatic duct (CHD), and a CBD stent was placed to assist with surgical dissection. Once pancreatitis was ruled out post ERCP, the patient ultimately underwent a successful open cholecystectomy, and a Jackson Pratt drain was left in place and removed once it remained serosanguinous. The gallbladder was sent to pathology, and the gross specimen was normal without signs of gallbladder adenocarcinoma. His white blood cell count down trended gradually but his total bilirubin remained elevated, due to the Dubin Johnson syndrome. He was successfully discharged without any complications.

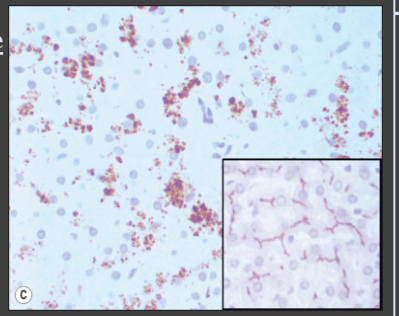
Mirizzi Syndrome

Image shows impacted gallstone in distal cystic duct causing external compression and obstructing the common hepatic duct.



Dubin-Johnson Syndrome

On liver biopsy of patient with Dubin Johnson, there is a complete absence of staining with immunohistochemical stain for cMOAT (MDR2)[6]



Conclusion

Dubin Johnson syndrome and Mirizzi syndrome are both rare syndromes, and can pose difficulty in getting a diagnosis and treatment, especially when found in the same patient. There is no data yet to suggest a correlation between the two syndromes, but perhaps more literature and evidence based cases are needed to identify one.

References

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