

CASE REPORT



Gilberts Associated with Dubin Johnson Syndrome in 50 Years Old Male Patient

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Received: 11-10-2019; Revised: 18-11-2019; Accepted: 26-11-2019.

ABSTRACT

A rare case was reported with Gilbert's syndrome in association with Dubin Johnson Syndrome in the rare age group. A 50 years male patient admitted in the hospital in Hyderabad with chief complaints of upper right quadrant pain, yellowish colour skin and whites eyes. His was severely exerted with office works. He drinks 6-7 units of alcohol per day. His abnormal lab investigations were precisely with elevated levels of neutrophils, depleted levels of monocytes. His urine examination was with cloudy, epithelial cells and pus cells. Liver function tests were shown as elevated levels of total bilirubin, direct bilirubin. Initially, the likely patient was diagnosed with Gilbert's syndrome. Belatedly it was once more diagnosed as Gilbert's syndrome in association with Dubin Johnson Syndrome with identification of black pigmentation of the liver and understood as liver enlargement, He promptly went with symptomatic drug treatment, additionally with Phenobarbitone drug to decrease bilirubin levels and Tetracycline and Streptomycin for active infections in urine.

Keywords: Gilbert's Syndrome, Dubin Johnson Syndrome, Serum bilirubin, UGT1A, MRP2, Phenobarbitone.

INTRODUCTION

Gilbert's disorder represents an innocuous hereditary condition which happens because of hepatic enzyme abnormality which leads to mellow elevations of bilirubin. The liver doesn't appropriately process bilirubin, and it is delivered by the breakdown of red blood cells¹. The condition has additionally been alluded to as sacred hepatic dysfunction and familial nonhemolytic jaundice². The key gene ordinarily controls an enzyme UGT1A (called Uridine Diphosphate Glucuronosyltransferases that are significant for bilirubin digestion) which situates on chromosome II³⁻⁵. Gilberts Syndrome is because of genetic mutation in the promoter locale of a gene. An ineffectual gene (naturally acquired from the parent) lead to excessive bilirubin typically develops in the blood⁶. Dubin Johnson disorder represents an uncommon, autosomal latent, kindhearted issue⁷ that causes an isolated increment of conjugated bilirubin in the serum. Classically, the condition causes a dark liver because of the pigment deposition like melanin⁸. This condition is related with a rare deformity in the capacity of hepatocytes to emit conjugated bilirubin into the bile. It is generally asymptomatic, yet might be analyzed in early outset dependent on laboratory tests. No treatment is typically required⁸. The conjugated hyperbilirubinemia is a consequence of a damaged endogenous and exogenous transfer of anionic conjugates from hepatocytes into bile¹⁰. Debilitated biliary discharge of bilirubin glucuronides is because of a mutation in the canalicular multiple drug resistance protein 2 (MRP2) genes. A hazily pigmented liver is expected to polymerized epinephrine metabolites, not bilirubin¹¹. Dubin Johnson disorder is because of an

imperfection in the MRP2 genes, located on chromosome X.⁷

CASE PRESENTATION

An unusual case report was identified with Gilbert's syndrome in association with Dubin Johnson Syndrome in a rare age group. A male patient with age group of 50 years admitted in general ward in Hyderabad with a chief complaint of upper right quadrant pain, yellowish colour skin and whites eyes. His was stressed with severe office works. He acquires a habit of alcohol consumption of 6-7 units per day. His abnormal lab investigations were found with elevated levels of neutrophils (79%) count, depleted levels of monocytes (17%). His urine examination was with cloudy, epithelial cells 1-2/HPF & pus cells 2-3/HPF. His liver function tests were shown as elevated levels of total bilirubin 4.5mg/dl, direct bilirubin 1.2mg/dl. At this stage, the patient was diagnosed as Gilbert's syndrome. When the patient's liver biopsy test was identified as black pigmentation of the liver and understood as liver enlargement, then it was again re-diagnosed as Gilbert's syndrome in association with Dubin Johnson Syndrome. He went with symptomatic drug treatment, additionally with phenobarbitone drug to decrease bilirubin levels and Tetracycline and Streptomycin for infections in urine.

DISCUSSION

This is an extremely rare case because Gilbert's syndrome in association with Dubin Johnson syndrome is unusual and Dubin Johnson occurrence in 50 years age is identified as very new based on literature survey. The patient's chief complaints of upper right quadrant pain, yellowish colour



skin and whites eyes were usually indicated initially as symptoms of jaundice¹². The patient has social habit alcohol consumption 6-7 units per day and one study says that alcohol consumption elevates the serum bilirubin levels¹³. As the patient urine appearance was founded with cloudy, pus cells and neutrophils count was with elevated levels, it has been considered the patient with urinary infection and it was treated with streptomycin and tetracycline drugs. The patient was diagnosed as Gilbert's syndrome which was assessed with elevated levels of bilirubin levels. This may be due to improperly processed bilirubin produced from the red blood cells break down. The previous study says that an infective gene inherited from parent leads to excessive increase of bilirubin in the blood due to genetic mutation in the promoter region of a gene which controls an enzyme UGT1A⁶. Belatedly the patient was also diagnosed again as Dubin Johnson syndrome which was assessed with an additional lab report of the liver biopsy which revealed as black pigmentation of the liver and enlarged liver. Dubin Johnson syndrome condition will also reveal with elevated levels of unconjugated serum bilirubin. A study reveals that impaired biliary excretion of bilirubin glucuronides is due to a mutation in the canalicular multiple drug-resistance protein 2 (MRP2) genes and darkly pigmented liver is due to polymerized epinephrine metabolites¹¹. The treatment for elevated serum bilirubin levels has gone with phenobarbitone. Phenobarbitone increases the clearance of bilirubin by enhancing UGT1A activity¹⁴ which is effective for Gilbert's syndrome. Phenobarbitone also enhances the rate of glucuronide formation and increase the canalicular bile flow¹⁵ which is effective for Dubin Johnson treatment. Ultimately, the patient was recovered.

CONCLUSION

This is an unusual case of Gilbert's syndrome in association with Dubin Johnson syndrome in 50 years male patient. Typically, no effective treatment is required for Dubin Johnson Syndrome, as the patient was with an associated condition of Gilbert's syndrome, he treated with phenobarbitone. Close monitoring of serum bilirubin levels is required for this specific type of conditioned patients.

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Source of Support: Nil, Conflict of Interest: None.

