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Case Report

Mirrizi Syndrome Mimicking Cholangiocarcinoma

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Abstract

Mirizzi's syndrome is a rare medical condition characterized by the occurrence of obstructive jaundice. Although infrequently observed, this clinical situation can evolve towards inflammation of the gallbladder and bile ducts, which can falsely mimic a cholangiocarcinoma. Medical history, clinical and biological evolution of jaundice, as well as Magnetic Resonance Imaging (MRI), are essential for making a diagnosis and, above all, preventing unnecessary and extensive surgery. We present a case of Mirizzi's syndrome in a 43-year-old man, who initially received a diagnosis of cholangiocarcinoma due to his clinical symptoms, elevated Cancer Antigen (CA)19-9 levels, and radiological result.

Introduction

Mirizzi's Syndrome (MS) was initially described in 1948 by the Argentinean surgeon, Pablo Mirizzi. It is an infrequently encountered condition characterized by the blockage of stones in the gallbladder's neck or cystic duct, leading to mechanical obstruction of the common hepatic duct and presenting clinically as intermittent or persistent jaundice [1]. The precise diagnosis of MS is of paramount importance for surgeons, as the condition may initially be mistaken for choledocholithiasis, bile duct stricture, or cholangiocarcinoma. Consequently, inappropriate surgical treatment poses a significantly heightened risk of inadvertent bile duct injury [2,3]. The objective of this study was to conduct a concise review of the literature and present a case of a 43-year-old man with MS initially misdiagnosed as cholangiocarcinoma due to his clinical symptoms, elevated Cancer Antigen (CA) 19-9 levels, and radiological result. The correct diagnosis was only established after surgery through anatomopathological examination

Case Reports

A 44-year-old man was admitted to the emergency department due to jaundice that had been present for a month. During the medical history interview, the patient reported an episode of acute abdominal pain occurring two months ago, which spontaneously resolved. Clinical examination revealed cutaneous-mucosal pallor, jaundice, and mild abdominal tenderness. The patient was afebrile and neurologically, hemodynamically, and respiratory stable. Considering the gradual progression of his symptoms, the patient was admitted to the gastrology department for further investigation of his jaundice.

Initially, a blood test was performed, revealing leukocytosis and biochemical evidence of cholestasis, characterized by elevated total bilirubin levels at 25.7 mg/dL (normal range: 0.1 to 1.2 mg/dL) due to direct bilirubin, as well as mild cytolytic damage indicated by serum glutamic oxaloacetic transaminase levels of 53 IU/L (normal range: 0 to 35 IU/L) and alkaline phosphatase levels of 248 IU/L (normal range: 45 to 129 IU/L).

An abdominal ultrasound was performed, revealing a sclero-atrophic gallbladder with dilation and wall thickening of the main bile duct and intrahepatic bile ducts.

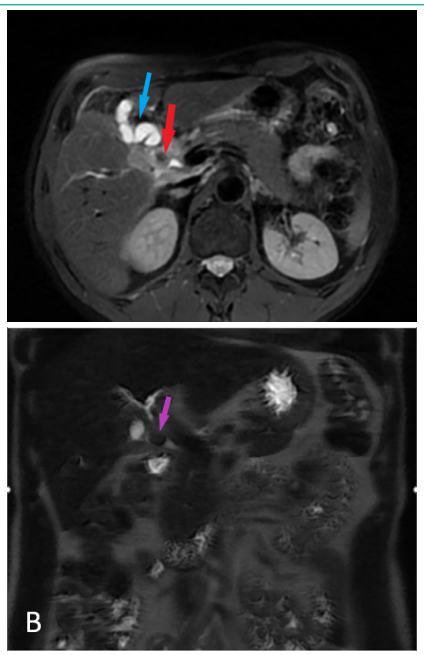

Figure 1: MRI Axial section (A) on T2 weighted gradient showing sclero-atrophic gallbladder, (blue arrow) with a trapped stone in the cystic duct (red arrow). MRI coronal section (B) with a trapped stone in the cystic duct (purple arrow).

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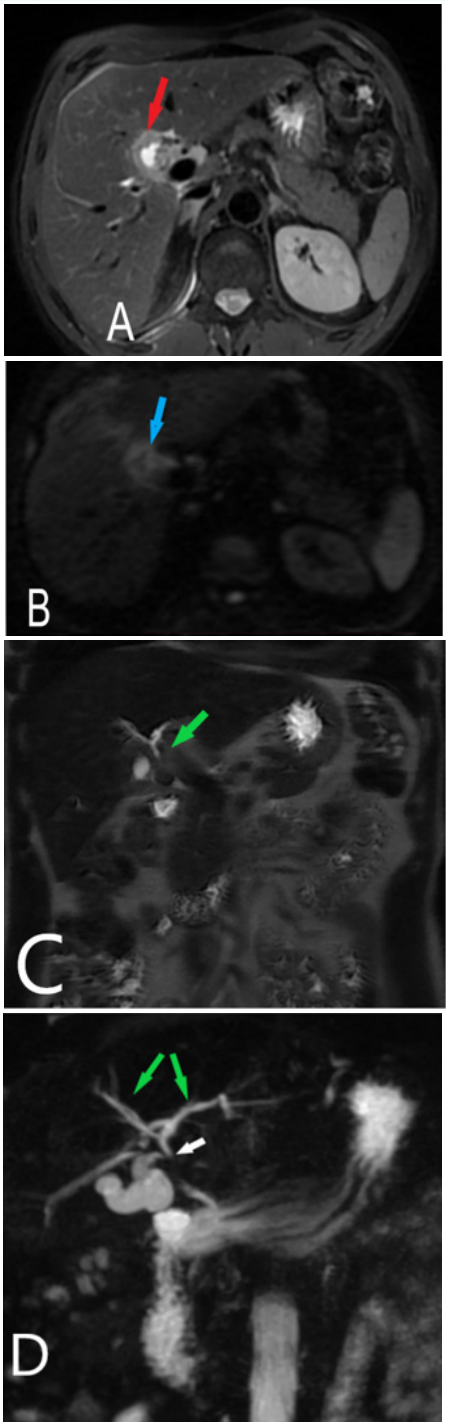

Figure 2: MRI Axial section on T2 weighted gradient showing thickening of the gallbladder wall (A, red arrow), with mild hyperintensity on diffusion-weighted imaging (B, blue arrow) extending to the main bile duct, the biliary bifurcation, and the intrahepatic bile ducts (C; green arrow). Coronal section on bili-MRI showing dilation of the intrahepatic bile ducts with image of sudden arrest at the level of the bile duct (D; green and white arrow).

A Magnetic Resonance Cholangiopancreatography (MRCP) was performed, revealing a sclero-atrophic gallbladder (Figure 1A) with a trapped stone in the cystic duct (Figure 1B) and thickening of the gallbladder wall, extending to the main bile duct, the biliary bifurcation, and the intrahepatic bile ducts (Figure 2). The findings

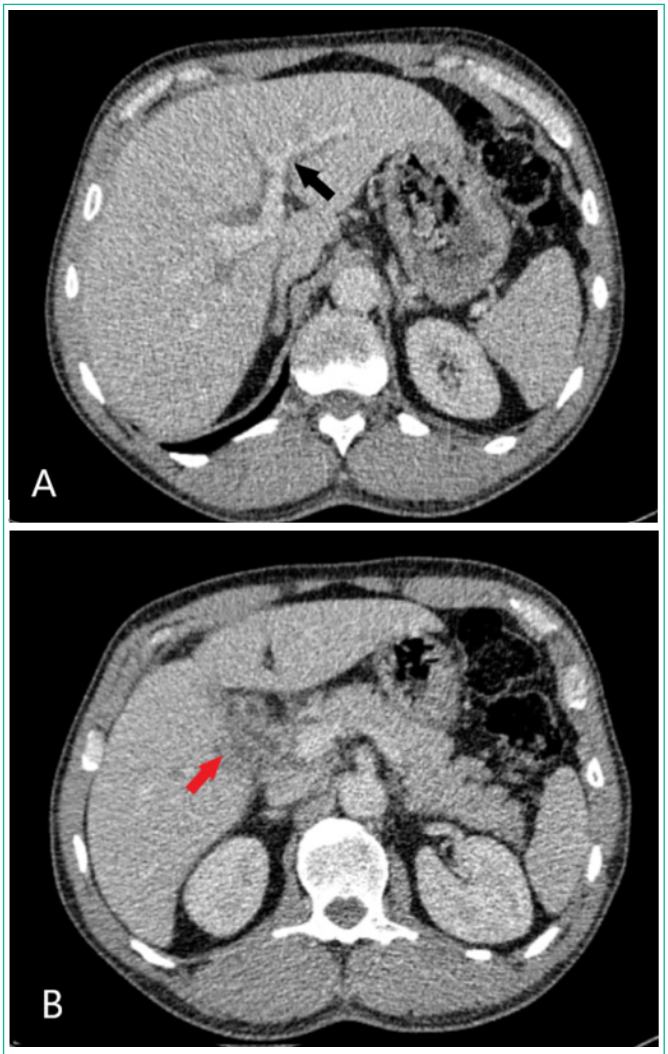

Figure 3: Axial CT section revealed moderate dilation of the intra-hepatic ducts up to the porta hepatis (A; black arrow), with soft tissue thickening affecting the gallbladder neck and proximal bile duct (B; red arrow).

showed hyperintensity on T2-weighted images, mild hyperintensity on diffusion-weighted imaging with homogeneous enhancement after contrast injection.In order to further explore the underlying pathology, a series of supplementary tests were performed. The patient's CA19-9 levels were found to be significantly elevated at 89 U/mL (normal range: 0 to 33 U/mL, median: 5.0 U/mL).

An abdominal Computed Tomography (CT) scan revealed moderate dilation of the intra-hepatic ducts up to the porta hepatis, with soft tissue thickening affecting the gallbladder neck and proximal bile duct, (Figure 3) as well as a few enlarged upper abdominal nodes. These findings suggested a neoplastic lesion involving the gallbladder neck and proximal common bile duct, leading to bile duct stricture. Considering the results from the MRCP, elevated CA19-9 levels, and CT scan findings, a tentative diagnosis of cholangiocarcinoma involving the gallbladder wall was established.

In light of the gradual decline in the biological cholestasis along with clinical improvement of the jaundice, the diagnosis of Mirizzi was suspected and confirmed during the intraoperative evaluation (Figure 4).

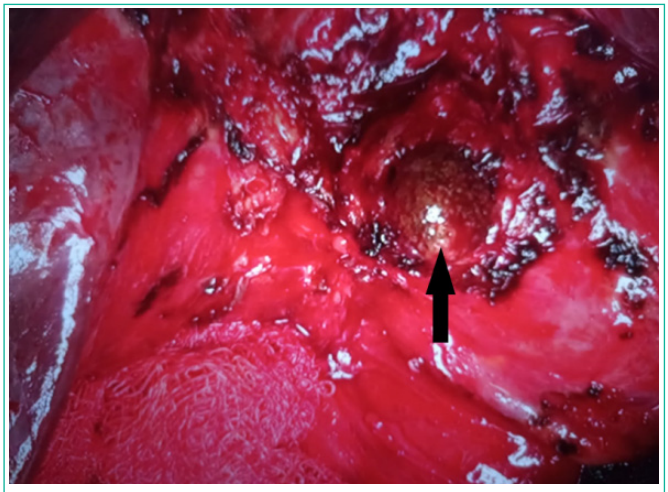

Figure 4: Intraoperative imaging showing the stone enclosed in the cystic duct (arrow).

Discussion

Mirizzi Syndrome (MS) was initially described by Kehr and Ruge in the early 1900s. However, it was in 1948 that Pablo Mirizzi provided a comprehensive description of this disorder and gave it its name [1,4]. It is due to acute and chronic inflammatory processes triggered by the lodgment of a large gallstone or multiple small gallstones in Hartmann's pouch of the gallbladder or the cystic duct, situated in close anatomical proximity to the common hepatic duct [5].

MS is a rare condition, with an incidence in patients undergoing biliary surgery varying from 0.7 to 1.4%, and age average from 53 to 70 years with around 70% of cases being in females. Nonetheless, it may occur at any age and in any patient with gallstones [4].

Predisposing factors for Mirizzi syndrome include a long intramural cystic duct or a low insertion of the cystic duct into the common bile duct [6].

Patients with Mirizzi syndrome typically present with clinical and biochemical signs indicative of biliary obstruction, which may sometimes occur in conjunction with acute cholecystitis, acute cholangitis, or pancreatitis. Usually, there is a longstanding history of biliary symptoms, and CA19-9 levels may show a moderate elevation, as observed in our patient [7-9].

Pre-operative diagnosis of Mirizzi's syndrome is of utmost importance to prevent complications arising from undetected cholecystobiliary or cholecystoenteric fistulas and inadvertent injury to the common hepatic duct during surgery. Achieving an accurate pre-operative diagnosis necessitates the application of combined imaging modalities, such as Ultrasonography (US) Computed Tomography (CT), and MRCP, to effectively assess the diagnosis [10].

On Ultrasonographic (US), typically we found is a large, immobile stone in the region of the neck of a shrunken gallbladder, with proximal dilatation of bile ducts. CT is not always specific to visualize gallstones, making difficult the diagnosis of Mirizzi syndrome [11].

MR cholangiopancreatography is very sensitive and specific in the detection of gallstone and bile duct stenosis. MR cholangiopancreatography can reveal the characteristic features of Mirizzi syndrome, including external compression and narrowing of the common hepatic duct, the presence of a gallstone in the cystic duct, dilation of the intrahepatic and common hepatic ducts, and a normal common bile duct .Nevertheless, it's important to note that the imaging findings may not always be specific and can sometimes be inconclusive .Rarely, inflammation around the common bile duct leads to stricture formation and thus resembles the periductal-infiltrating type of cholangiocarcinoma [11]. In our patient, considering the results from the MRCP, elevated CA19-9 levels, and CT scan findings, a tentative diagnosis of cholangiocarcinoma involving the gallbladder wall was established.

It is not an easy task to asses pre-operative diagnosis of Mirizzi's syndrome, it continues to be a challenge for the surgeon. Our patient was a postoperative diagnosis, made through anatomopathological examination, but in the intraoperative we also had this suspicion. Surgery is the primary treatment choice for MS [12]. It is based on extraction of the obstructing stone, removal of the gall bladder, and restoration of normal biliary drainage [10]. For our patient, the preoperative treatment raised suspicions of the diagnosis, which was later confirmed postoperatively through anatomopathological examination.

Conclusion

In summary, the rarity of Mirizzi syndrome and its absence of specific clinical features make it prone to misdiagnosis, often being confused with cholangiocarcinoma. Pre-operative diagnosis of Mirizzi's syndrome remains a challenging task, and MRI is an accurate method for preoperative diagnosis and surgical planning.

Author Statements

Guarantor of Submission

The corresponding author is the guarantor of submission.

Author's Contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Patient Consent Statement

Written informed consent for publication was obtained from patient.

Declaration of Interests

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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