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GALLBLADDER ADENOCARCINOMA COURSE WITH MIRIZZI SYNDROME: CASE REPORT

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Abstract: Introduction: Gallbladder carcinoma (BVC) is the most common malignant neoplasm of the bile ducts, affecting more women in their sixties. It most commonly presents as an asymptomatic condition, but it can also present non-specific symptoms, or even mimic other biliary diseases, such as Mirizzi's syndrome (a biliary pathology characterized by obstructive jaundice). Tumor resection in early stages can be curative, but due to the lack of effective screening tests, its diagnosis usually occurs at a late stage, dramatically worsening the prognosis of the disease. In this case report, we approach a patient with gallbladder cancer associated with Mirizzi syndrome. **Case report:** Female patient, 59 years old, consulted at the surgery outpatient clinic of Santa Casa de Misericórdia de Fortaleza with complaints of jaundice associated with pruritus, fecal acholia and choluria. Initially, abdominal ultrasound was performed, which showed dilatation of the intrahepatic bile ducts, but without identification of obstructive factors. She was submitted to an resonance cholangiography which showed luminal narrowing at the level of the confluence of the hepatic ducts due to probable extrinsic compression of the gallbladder. Cholecystectomy was performed with partial hepatectomy sufficient to guarantee a surgical margin. During dissection of the hepatic pedicle, invasion of the left hepatic duct was observed, which was resected and a biliary anastomosis of the hepatic ducts with the jejunum was performed. Histopathological analysis of the gallbladder confirmed the diagnosis of adenocarcinoma. **Conclusion:** Concomitance of gallbladder adenocarcinoma with Mirizzi syndrome is rare. Detailed clinical history and imaging tests are sometimes not sufficient for diagnosis, requiring the association of histopathological methods to avoid underdiagnosis of this condition.

Keywords: Gallbladder adenocarcinoma; Mirizzi syndrome; jaundice; bile ducts.

INTRODUCTION

Gallbladder carcinoma is the most common malignant neoplasm of the bile ducts [2] and the seventh most frequent of the digestive tract. It is usually associated with a poor prognosis, due to its often late diagnosis [2, 14]. This is due to the absence of early symptoms of this condition and the lack of screening tests for this pathology. Treatment consists primarily of surgical removal of the lesion, but adjuvant therapies such as chemotherapy and radiotherapy are often used [2, 14].

Mirizzi syndrome is characterized by a pathology of the gallbladder causing benign obstructive jaundice [3,6]. The picture consists of a cystic duct that anatomically runs parallel to the common hepatic duct associated with an impacted stone in the neck of the gallbladder or in the cystic duct itself. This situation leads to an obstruction of the common bile duct caused by the impacted calculus or by the inflammatory response itself [1,5,6]. In this case report, we approach a patient with gallbladder cancer associated with Mirizzi syndrome.

CASE REPORT

Female patient, 59 years old, consulted at the General Surgery outpatient clinic of Santa Casa de Misericórdia de Fortaleza with a complaint of jaundice associated with pruritus, fecal acholia and choluria. She had been reporting bar pain in the upper abdomen for about a month, radiating to the right hypochondrium, which worsened with food. She denied weight loss and hyporexia.

Initially, abdominal ultrasonography was performed, which showed biliary sludge and intrahepatic bile duct dilatation, with no obstructive factors to the method. Then,

resonance cholangiography showed dilatation of the intrahepatic bile ducts (Figure 1), associated with luminal narrowing at the confluence of the hepatic ducts due to probable extrinsic compression of the gallbladder, a condition compatible with Mirizzi syndrome.

Because she had a condition compatible with gallstone disease associated with Mirizzi syndrome and without warning signs of neoplastic disease, the patient was initially submitted to videolaparoscopic cholecystectomy (Figure 2). After an inventory of the cavity with a 30o optic, a gallbladder was visualized with a petrified appearance, pearly color and well adhered to the common hepatic duct. So decided by conversion to open technique. Cholecystectomy was performed and surgical specimen was sent for intraoperative frozen section biopsy, whose result was suggestive of adenocarcinoma. We proceeded with dissection of the structures of the hepatic pedicle and due to the great adherence of the gallbladder to the common hepatic duct, a central hepatectomy was chosen (margin above 2 cm) with resection of the right and left bile duct 5 cm above the bifurcation of the hepatic ducts. reconstruction was performed with two right and left hepatic-jejunal anastomoses. In order to perform a surgery with a curative proposal despite the locally advanced disease, a lymphadenectomy of the hepatic hilum was also performed. The histopathological analysis associated with the immunohistochemical study corroborated the diagnosis of gallbladder adenocarcinoma.

DISCUSSION

Mirizzi Syndrome, described in 1948 by the surgeon Pablo Luis Mirizzi, is a rare complication that occurs in approximately 1% of patients with cholelithiasis. Its etiology is usually due to impaction of a gallstone at the level of the cystic duct or infundibulum associated with compression of the common hepatic duct, which can generate a cholecysto-choledocian fistula (the most common complication) [8]. The diagnosis of this syndrome can be very difficult [10], as it can be asymptomatic for a long period [9] or simulate other biliary diseases, such as gallbladder cancer itself [8, 9, 10], reported in this case.

A classification system was developed to categorize Mirizzi syndrome - In type I there is no fistula (it is subdivided into IA when the cystic duct is present and IB when there is obliteration of the cystic duct). Types II to IV are characterized by the presence of fistula and are grouped according to the degree of involvement of the common hepatic duct, with type II associated with a defect smaller than 33% of its diameter, type III with a defect of 33% to 66% of the diameter and type IV with a defect greater than 66%. Finally, type V is described when there is the presence of cholecystic fistula, with or without the presence of gallbladder ileus (types Va and Vb respectively) [10].

An early and accurate diagnosis has a major impact on the management and prevention of future complications. The initial screening test is abdominal ultrasound, which allows the visualization of signs indicative of such pathology, such as: impacted calculus in the infundibulum, contracted gallbladder with dilatation of the intrahepatic bile duct and common hepatic duct. However, endoscopic retrograde cholangiopancreatography (ERCP) remains the best method, allowing a better assessment of the anatomy. Other tests, such as magnetic resonance cholangiopancreatography, can be performed despite the ultrasound findings and according to hospital availability, but often this diagnosis ends up being performed intraoperatively [8,11]. Tumor markers, including Carcinoembryonic Antigen or Carcinogen Antigen 19-9, may be elevated

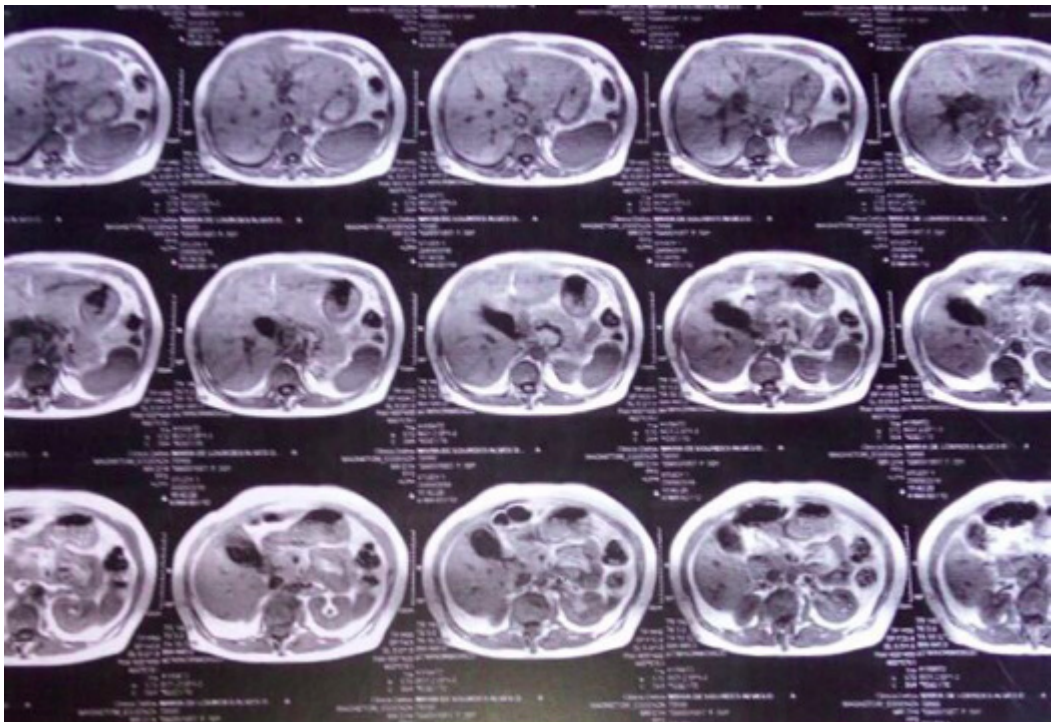

Figure 1: Image of cholangioresonance.

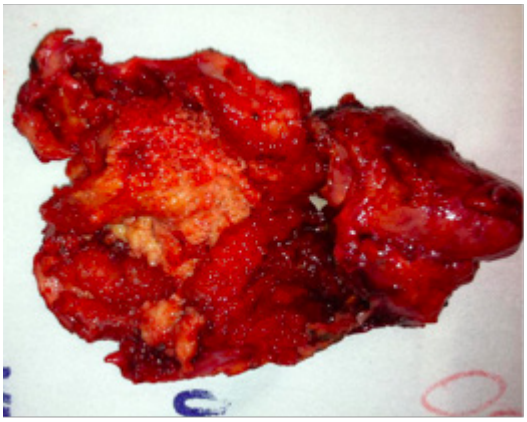

Figure 2: Surgical piece, gallbladder

in some cases but are not specific. Similarly, alkaline phosphatase or serum bilirubin may be elevated, but usually only when sufficient disease progression has occurred to facilitate bile duct obstruction [14]. In the case patient, abdominal ultrasonography was performed as a screening, but as no obstructive factor was observed in the method, magnetic resonance imaging of the biliary tract was chosen for ERCP, as it is less invasive, but without visualization of masses; as in statistics, the diagnosis was made intraoperatively.

Currently, a standard treatment for Mirizzi syndrome is not yet well defined, due to the heterogeneity of clinical presentation and anatomical distortion, and therefore, surgical treatment must be planned after a careful assessment of the stage of the disease and biliary anatomy, with the objective of main to avoid any damage to the bile ducts. The approach can be open or laparoscopic, depending on the stage and experience of the assistant surgeon [6, 9].

Gallbladder cancer (GBC) is an uncommon neoplasm, but one of the most life-threatening in the world. The histological type most frequently identified is adenocarcinoma, being more common in women and in their sixties [2,4]; it does not usually present with an exuberant clinic, being often insidious and with unusual presentations, as in the case in question where it manifested itself through Mirizzi syndrome. Among the factors related to its etiopathogenesis, cholelithiasis, mainly in coexistence with biliary sludge, is present in 96% of patients, being the main risk factor [13,14].

GBC is an aggressive neoplasm, with an overall 5-year survival ranging from 5 to 32%, and only 10% are diagnosed while the disease is restricted to the gallbladder. The standard treatment for early stages is simple cholecystectomy, which has an overall survival above 90%. In advanced cases, even when extended cholecystectomy with lymphadenectomy and resection of adjacent invaded organs is performed, 5-year survival remains low. [12] The median survival of patients with advanced stage disease is less than 6 months [13, 14].

The absence of early symptoms confers a reserved prognosis to patients, often with invasion of adjacent structures at the time of diagnosis [2,5,12]. Although in our case there was already some degree of invasion, it was still possible to completely resect the lesion, ensuring better patient survival.

CONCLUSION

Mirizzi syndrome and gallbladder adenocarcinoma are uncommon diseases, and their concomitance is extremely rare. The diagnosis of gallbladder neoplasia can have many presentations, including simulating unusual conditions such as Mirizzi Syndrome. Thus, a detailed clinical history and imaging tests may not be sufficient for the diagnosis of neoplasia, requiring the association of histopathological methods to avoid underestimation of the diagnosis.

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