Left hepatectomy for cholangiocarcinoma: case report and literature review

Hepatectomia a esquerda por colangiocarcinoma: relato de caso e revisão de literatura*

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ABSTRACT

Cholangiocarcinoma is a rare biliary tract malignant tumor that remains a major challenge for surgeons. It is associated with poor prognosis and high morbidity and mortality rates even after surgical treatment. Diagnosis by non-invasive methods (3D CT, MR cholangiography) enables precise tumor staging in addition to correctly selecting patients for surgery. The general consensus has shown that surgery by completely resecting the tumor and leaving free surgical margins is the treatment that offers the best prognosis and the only chance of cure. Chemotherapy and radiotherapy in the postoperative period do not seem to change survival or recurrence rates, even in cases in which complete resection is performed. Despite all diagnostic and therapeutic advances, the 5-year survival rate remains 7-33%. Up to now, surgery is the treatment that offers the best chances of cure. In the present study, we describe the surgical treatment of a patient with cholangiocarcinoma with a satisfactory outcome and present a review of the current treatment in international literature for patients with cholangiocarcinoma.

Keywords: Cholangiocarcinoma/surgery; Hepatectomy; Biliary tract/pathology; Biliary tract surgical procedures; Drug therapy

INTRODUCTION

Cholangiocarcinoma is a rare biliary tract malignant tumor that remains a major challenge for surgeons. It is associated with poor prognosis and high morbidity and mortality rates even after surgical treatment. Little is known about its etiology and pathogenesis (1).

It classically occurs in individuals aged over 60 years and is more frequent in males, in a ratio of 3:2.

Clinically, patients present progressive jaundice associated with abdominal pain and weight loss, and the physical examination is generally inconclusive. The diagnosis is made through imaging examinations, such as ultrasound, computerized tomography (CT) and magnetic resonance cholangiography (MRC). The only curative treatment is surgery, by resection of the biliary tract associated with an occasionally extensive hepatectomy, and cholangio-enteric bypass. In cases in which surgical resection is contraindicated, palliative measures, such as endoscopic stenting in the biliary tract or percutaneous transhepatic biliary drainage, are carried out in order to improve jaundice and/or cholangitis and extend survival.

Despite all diagnostic and therapeutic advances, the 5-year survival rate remains 7-33%. So far, surgery is the treatment that provides the best chances of cure (2-4).

CASE REPORT

We treated a Caucasian, 74 year-old patient, with a history of intense epigastric pain associated with fever and chills for a
week. On the initial physical examination, the patient had fever, pain at palpation during deep breath (Murphy sign) and was clinically anicteric. These findings let to a presumptive diagnosis of cholangitis and laboratory tests and imaging examinations were requested. The tests confirmed the diagnosis of cholangitis and aroused the suspicion of biliary tract neoplasm. The MRC showed an intraductal growing nodular lesion at the intrahepatic bile duct projection of the IV-A segment, associated with intrahepatic biliary tract dilation, retraction and edema of this segment, leading to the diagnosis of an expanding lesion of peripheral intraductal origin. Initially, cholangitis was treated followed by surgery to resect the expanding lesion. The surgical treatment consisted of expanded left hepatectomy (segments I, II, III and IV), performed with temporary clamping of the hepatic hilum. The gallbladder and the extrahepatic biliary tract were also resected due to proximity of the lesion and as a surgical tactics. The biliary tract was reconstructed with hepaticojejunostomy using a jejunal Roux-en-Y loop and non-absorbable suture (prolene 6.0). The surgical procedure was carried out uneventfully, and the patient is clinically well ten months later. The histological examination of the surgical specimen confirmed the diagnosis of multifocal papillary adenocarcinoma of the biliary tract, infiltrating adjacent connective tissue. The surgical margins of the bile duct and liver parenchyma showed no signs of malignancy.

**DISCUSSION**

Resection of biliary tract tumor remains a challenge for surgeons and numerous studies recently published show that there are few cases of cure\(^{(2)}\). Cholangiocarcinoma presents typically as a sclerosing adenocarcinoma that can extend from the liver parenchyma to the distal bile ducts. Some signs suggest poorer prognosis: obstructive jaundice, serum albumin levels < 35 g/L, need for extensive liver and/or vascular resections, and a location proximal to the bile duct\(^{(3,4)}\). Non-invasive diagnostic methods (3D CT, MR cholangiography) enable precise tumor staging in addition to correct selection of patients to whom surgery is indicated\(^{(5)}\). Some centers point out the importance of gene p53 mutation in the pathogenesis of cholangiocarcinoma and emphasize its identification in routine staging\(^{(6)}\).

The general consensus has shown that surgery with complete tumor resection and free surgical margins is the treatment with the best prognosis and the only chance of cure\(^{(7)}\); postoperative chemotherapy or radiotherapy do not seem to change survival or recurrence rates, even in cases with complete resection\(^{(8)}\). On the other hand, radical surgical treatment may be contraindicated in some situations, such as presence of distant metastases, invasion of the portal vein or adjacent vascular structures or need for extensive liver resections\(^{(9)}\). These situations occur in most cases in which palliative measures should be performed, such as endoscopic stenting in biliary tree, percutaneous transhepatic biliary drainage and cholangio-enteric bypass; these procedures aim at relieving jaundice and/or cholangitis, improving clinical status and extending patient survival\(^{(2,6)}\).

Survival rates have improved as surgical interventions have progressively become more aggressive. However, surgeries with extensive resections have increased hospital morbidity and mortality rates\(^{(10)}\). Due to its local invasive nature, most specialized centers have performed partial hepatectomies in 50 to 100% of the cases\(^{(5)}\). A large series assessed in Italy showed the need for caudate hepatectomy as a statistically significant factor for 5-year survival rate (25% for patients submitted to caudate hepatectomy and for those not submitted it was zero)\(^{(7)}\). In several centers, the 5-year survival rate for patients submitted to curative surgery varied from 7 to 33%\(^{(2,10)}\).

Prognostic factors related to longer survival are associated with free surgical margins, absence of nodal metastases, tumor histology, absence of gene p53 mutation and no invasion of adjacent vascular structures\(^{(1,2,10)}\).

A recent study carried out in Japan using tumor necrosis factor (TNF) receptors, showed its promising role as an antitumor agent for treating human cholangiocarcinoma\(^{(11)}\). Further clinical trials are required to better establish the role of this agent.

In the case reported in this study, the only sign of poor prognosis presented was its location close to the bile duct. During
surgery, it was possible to resect the tumor with free and expanded margins and to verify there no local lymph nodes involved. Comparing the case with literature reports, the chances of a favorable long-term outcome are promising.

CONCLUSION

Early diagnosis by non-invasive methods and expanded local resection with free surgical margins associated with partial hepatectomy yield the best chances of survival for patients with cholangiocarcinoma. Further studies are required to better clarify the pathogenesis of cholangiocarcinoma. So far, surgical treatment seems to be the only alternative to effect cure.

REFERENCES