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PSEUDOTUMORAL PARACOCCIDIOIDOMYCOSIS OF THE BILE DUCTS: CASE REPORT

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Abstract: Paracoccidioidomycosis is a systemic granulomatous disease restricted to Latin America, caused by the dimorphic fungus *Paracoccidioides brasiliensis*. It typically occurs in chronic form, affecting the lungs, skin and lymph nodes. Rarely, paracoccus infection can simulate a neoplasm, especially in abdominal organs, and must be considered as a differential diagnosis of cholangiocarcinoma, especially in patients coming from endemic areas, even in those without routine exposure to contaminated soil. We report two cases of paracoccidioidomycosis simulating distal and perihilar cholangiocarcinoma.
Keywords: Paracoccidioidomycosis; Cholangiocarcinoma; Biliary tract; Liver.

INTRODUCTION

Paracoccidioidomycosis, also known as Lutz-Esplendore-Almeida disease or South American blastomycosis, is a systemic granulomatous disease caused by thermodimorphic fungi that currently comprise two species: *Paracoccidioides brasiliensis* and *Paracoccidioides lutzii* [1], endemic in Latin America, mainly in Brazil, Colombia and Venezuela. It was described by Adolfo Lutz in 1908 [2], and its incidence varies from 3 to 4 cases per million, up to 1 to 3 cases per hundred thousand inhabitants per year in endemic areas, with an annual mortality of 1.45 per million inhabitants, the highest rate being observed among systemic mycoses [1]. It mainly affects male adults, between 30 and 60 years of age, with two forms of presentation: the juvenile form, constituting 3 to 5% of cases, which affects the reticuloendothelial system, and the chronic form, more common, which comprises 90%. of cases, mainly affecting the skin, lymph nodes and lungs [1]. It is a disease with an important social and economic impact, as it mainly affects individuals of working age. It is believed that around 50% of inhabitants of endemic areas have been exposed to *P. brasiliensis*, however, only a small proportion of exposed individuals present any clinical manifestations. When not diagnosed and treated, it can lead to severe and lethal disseminated forms, with abrupt and progressive involvement of the lungs, integument, ganglia, spleen, liver and lymphoid organs of the digestive system. Paracoccidioidomycosis can simulate periampullary [3] or colon [4] neoplasms, being an important differential diagnosis of abdominal tumors to be considered in endemic areas [5].

This mycosis must be seen as an important Public Health problem due to its high disabling potential and the number of premature deaths it causes, mainly for specific social segments, such as rural workers, since the management of soil contaminated with the fungus constitutes the main risk factor for infection, which can manifest itself many years after infection [1].

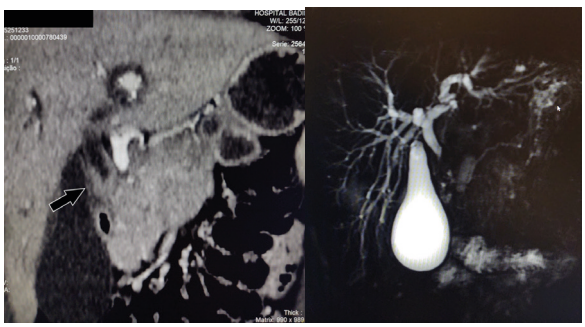
CASE REPORT

PATIENT 1

A 34-year-old male patient, born in Rio de Janeiro, Brazil, sought care in the emergency department of our hospital presenting with jaundice that had lasted for a week, associated with choluria and fecal acholia that had started fifteen days ago. He reports a similar episode two months earlier, after copious ingestion of fatty foods, associated with malaise, constipation and a feeling of gastrointestinal fullness. He also reports asthenia and weight loss of approximately 8 kg in two months. The physical examination revealed significant jaundice (+3/4), painless abdomen on palpation, with a palpable and painless gallbladder in the right hypochondrium (Courvoisier-Terrier sign). Upon admission, the patient had a serum bilirubin of 5.14 mg/dL, at the expense of direct bilirubin (4.86 mg/dL), in addition to an increase in canalicular

enzymes and transaminases. Abdominal computed tomography without contrast showed cholelithiasis, moderate dilation of the intra- and extrahepatic bile ducts and an apparent increase in the dimensions of the pancreatic head, with blurring of its contours. Aortic and inter-aorto-caval lymph node enlargement in the upper abdomen, the largest on the left measuring approximately 2.7 x 1.9 cm, were also observed.

A hypodense image was also evident with liquid areas anterior to the left psoas muscle, measuring approximately 2.1 x 1.4 cm.



Aiming to study the bile ducts, magnetic resonance cholangiography was performed, where thickening of the distal common bile duct could be observed, with interruption of biliary drainage and dilation of the intra- and extrahepatic bile ducts upstream. The patient underwent ERCP, and dilation of the intra- and extrahepatic bile ducts was observed, with a diameter of 10 mm, associated with abrupt interruption of biliary drainage in the middle third of the common bile duct. Deep catheterization of the bile duct was performed, with maintenance of the guide wire in the intrahepatic biliary branches and biopsy of the lesion with a cytology brush, in addition to a wide papillotomy, with the introduction of a 10 Fr x 9 cm plastic biliary prosthesis. Serum levels of CA 19.9 and CEA during hospitalization were 2.0 and 1.46, respectively.

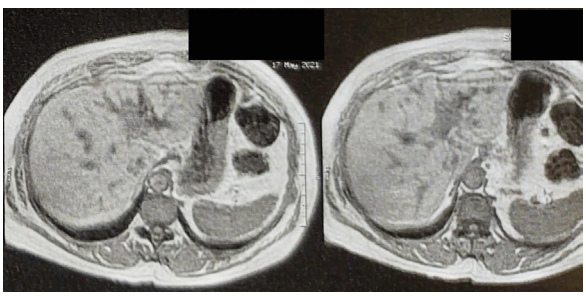
With early-stage distal cholangiocarcinoma as the main diagnostic hypothesis, surgical treatment was indicated. The patient underwent a gastroduodenopancreatectomy, with Roux-en-Y biliodigestive reconstruction, showing good clinical evolution postoperatively. We obtained the results of the histopathological analysis ten days after surgery, where the presence of numerous granulomas containing the fungus *Paracoccidioides* brasiliensis was observed in the wall of the distal common bile duct, using Grocott staining. No sign of malignancy was observed in the surgical specimen. With diagnostic confirmation, treatment with intravenous Amphotericin B was started during hospitalization. After completion of parenteral therapy, the patient was discharged and was prescribed treatment with oral Itraconazole.

PATIENT 2

A 63-year-old female patient, born in Rio de Janeiro, Brazil, was admitted with a history of jaundice, abdominal pain, nausea, vomiting and weight loss of approximately 10 kg in two months. On examination, she was jaundiced (+1/4), with a painless abdomen, without palpable masses. An MRI cholangiography revealed the presence of an infiltrative-looking lesion, with ill-defined edges and contrast enhancement, located in the central portion of the transition of hepatic segments II/IVa and III/IVb, determining dilation of the intrahepatic bile ducts, mainly from the left hepatic lobe. Lymph node enlargement was also evidenced near the hepatic hilum, measuring approximately 1.8 x 1.6 cm, and peripancreatic lymph nodes measuring 1.2 x 1.0 cm, associated with prominent pancreaticoduodenal, portocaval, near the gastric fundus and pericardiophrenic lymph nodes.

There was a slight increase in transaminases and direct bilirubin (1.2 mg/dL), associated with a significant increase in alkaline phosphatase and gamma glutamyl transferase.

As advanced Klatskin Tumor was the most likely diagnostic hypothesis for the case described, we proceeded with a surgical approach aiming to drain the bile duct. The patient underwent a Hepp-Couinaud biliodigestive shunt, and a biopsy of the retrocholedochal lymph node was performed. With good clinical evolution postoperatively, the patient was discharged from hospital eight days after the procedure. Histopathological analysis showed a chronic granulomatous inflammatory process with necrosis, multinucleated giant cells and numerous rounded structures containing *Paracoccidioides* brasiliensis, after research using Grocott staining. After confirmation of chronic paracoccus infection, treatment with parenteral Amphotericin B was indicated, complemented by oral Itraconazole after hospital discharge.



DISCUSSION

The involvement of the bile ducts by Paracoccidioidomycosis can occur in chronic multifocal forms, progressing with jaundice of obstructive origin, which comprises three pathophysiological mechanisms: extrinsic compression of the bile duct due to lymph node enlargement, hepatitis and/or blastomycotic pancreatitis, and intraluminal blastomycotic granuloma of the common bile duct. In the reported cases, the presence of consumptive syndrome associated with cholestatic syndrome, as well as imaging tests showing thickening of the distal common bile duct, presence of an intrahepatic mass and lymph node involvement, led us to consider cholangiocarcinoma as the main diagnostic hypothesis. Despite being a rare presentation, biliary paracoccidioidomycosis must be considered as a differential diagnosis in the presence of obstructive jaundice and radiological findings compatible with intra-abdominal tumors when found in endemic areas, even in the absence of pulmonary involvement and peripheral lymph node enlargement.

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