Malignant lymphoma primarily originating from the extrahepatic bile duct is extremely rare, and preoperative diagnosis is difficult because symptoms and imaging are nonspecific [1, 2]. The most common symptoms of primary biliary lymphoma are abdominal pain, weight loss, fever, and obstructive jaundice [1, 2]. A total of 23 cases have been reported in literature [1–5], and, to the best of our knowledge, this is the fourth case of diagnosis made preoperatively and the first report of primary biliary lymphoma with acute pancreatitis and cholangitis as the first clinical manifestations.

A 30-year-old male patient was referred to our hospital for acute pancreatitis. On admission he had severe abdominal pain, fever, and jaundice. Laboratory findings were: total leukocyte count, 15700/µL; C-reactive protein, 7.89 mg/dL; aspartate aminotransferase, 271 U/L; alanine aminotransferase, 428 U/L; alkaline phosphatase, 580 U/L; γ-glutamyl transpeptidase, 428 U/L; serum total bilirubin, 9.1 mg/dL (direct 8.5 mg/dL); amylase, 951 U/L; and lipase, 1837 U/L. Transabdominal ultrasonography revealed only a dilated common bile duct (CBD) with sludge in the gallbladder. An urgent endoscopic ultrasound showed a stricture in the distal portion of the CBD, with proximal bile duct dilatation (Fig. 1).

During the same endoscopic session, endoscopic retrograde cholangiopancreatography (ERCP) and cholangiography confirmed the stricture in the lower portion of the CBD, with proximal dilatation (Fig. 2a); sphincterotomy and forceps biopsies were carried out (Fig. 2b), and finally a plastic stent was placed for drainage (Fig. 2c).

Histological examination (Fig. 3a) and immunohistochemistry (Fig. 3b–d) reveal...
vealed a large B-cell-type malignant lymphoma.

Total body computed tomography (CT) showed a well-circumscribed, heterogeneous, enhancing mass in the lower-mid portion of the CBD, without evidence of metastases (Fig. 4).

The patient was symptom-free and scheduled for surgical resection. Examination of the surgical specimen confirmed the diagnosis of large B-cell-type malignant lymphoma, without metastasis in the lymph nodes included in the resection. The patient was referred to an oncologist for chemotherapy (rituximab, cyclophosphamide, vincristine, doxorubicin, and prednisolone [R-CHOP]) and after 6 months of follow-up he remains asymptomatic.

Competing interests: None

References