

## CHOLANGITIS AS AN UNUSUAL CLINICAL PRESENTATION OF PANCREATIC LYMPHOMA: CASE REPORT

*Colangite como uma manifestação clínica atípica de linfoma pancreático: relato de caso*

Cleber **SOARES-JUNIOR**, Carlos Augusto **GOMES**, Rodrigo de Oliveira **PEIXOTO**, Luciano Augusto **JUSTE**

From the Universidade Federal de Juiz de Fora, Juiz de Fora, MG, Brasil

### Correspondence:

Cleber Soares Júnior,  
e-mail: cleberdoc@ig.com.br

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### INTRODUCTION

**N**on-Hodgkin's lymphoma (NHL) involving the pancreas is less common but well documented and this organ is the primary site of it<sup>10,11</sup>. The role of surgical resection in curing this disease is poorly defined. The majority of patients with pancreatic lymphoma presents a mass in the head of the gland<sup>13</sup>, and is often described as a large mass with or without associated lymphadenopathy. Because pancreatic carcinoma has such a dismal outlook, many patients do not undergo biopsy and the failure doing this diagnosis eliminates the chance of cure<sup>1,12</sup>. It is present here a case of pancreatic lymphoma in which cholangitis was its main sign.

### CASE REPORT

A 41 year-old male was examined with a two month history of weight loss and jaundice. Patient signs and symptoms consisted in intense jaundice, fever and pain in the right side (Charcot's triade). There were also pruritus, faecal acholia and choluria. Weight loss was referred as to 8 kg in two months.

The patient did not record any other disease, neither alcoholism nor smoking. There was no history of pancreatitis, diabetes, biliary lithiasis or cancer. Laboratory results are showed total and conjugated bilirubin of 5.56 mg/dl and 4.77 mg/dl respectively. Alkaline phosphatase was 1500 U/l (normal range: 27 – 100 U/l) and  $\gamma$ -glutamyl transferase was 450 (normal range: 5–27 U/L). Prothrombin activity was 46%. Viral markers, HbsAg, Anti-HBc and Anti-HCV were negative. Abdominal ultrasound showed intra and extrahepatic dilatation of biliary tree and a mass in the head of pancreas. Abdominal tomography scan showed a heterogeneous 6 cm diameter lesion in the head of

pancreas, intense dilatation of biliary tree, encasement of mesenteric superior and portal veins (Figure1). There were neither ascitis nor cholelithiasis, no metastasis was found. Gastroduodenoscopy depicted esophagitis, an important extrinsic compression of duodenum with deformation of second portion.



**FIGURE 1** – Abdominal computed tomography scan showing an hipodense tumor involving hepatic and mesenteric vessels

The institutional protocol do not use preoperative biliary decompression and so, it was not done. Angiography also was not chosen. Diagnosis was defined as pancreatic neoplasm in the head and the surgery was expedited. The patient was submitted to a large subcostal wall incision. After opening the peritoneal cavity, visual inspection and palpation of abdominal and pelvic cavities was the initial aim. There were no distant metastases, but portal vein involvement, encasement of hepatic artery and superior mesenteric vessels were encountered. Actually the tumour extended beyond the normal limits of pancreatic resection and there was fixity to the retroperitoneum. It was not identified omental and peritoneal seedings, and a lot of compromised lymph nodes were obvious in the perihepatic location. All these intraoperative findings contraindicate head pancreatic resection (Whipple pancreaticoduodenectomy).

The patient was submitted to a palliative procedure that consisted in a biliodigestive anastomosis (side-to-side hepaticojejunal anastomosis) associated to a

gastrojejunal bypass. Gallbladder was already removed. There was no intraoperative haemorrhage, no need for transfusion. Penrose drain was used as a protocol. Some biopsies were done. Operative time was about four hours; cefazolin was used as a prophylactic antibiotic.

There were some complications on postoperative course mainly because of gastrointestinal bleeding that started six days after surgery. Endoscopic evaluation was necessary to control haemorrhage and was done twice with good results. Transfusion also was necessary, and the patient needed three units of globules. Hospital discharge occurred after 11 days.

Histopathology examination of intraoperative biopsies showed pancreatic lymphoma. Patient was evaluated by the oncology service and started a combination CHOP chemotherapy: Cyclofosfamide, Adriablastina, Vincristina (Oncovin) and prednisone (100mg/dia). He is now alive and free of symptoms after 60 months of treatment.

## DISCUSSION

Extranodal non-Hodgkin's lymphoma accounts for approximately 25% of cases. In most series of primary extranodal lymphomas, the gastrointestinal tract is the most common site, accounting for 30% to 40%, the most common sites for solitary extranodal lymphoma were stomach, skin, and brain. Survival correlated with histologic subtype. Primary non-Hodgkin's lymphoma of the pancreas is a rare disease and accounts for less than 1% of extranodal non-Hodgkin's lymphomas and 0.3% of all pancreatic tumors<sup>2</sup>. Its diagnosis is difficult without histological examination, and clinical and imaging findings are not pathognomonic.

Patients with gastrointestinal lymphoma may present with nausea and weight loss (50%) due to malabsorption (this does not constitute a B symptom in the conventional sense), abdominal pain (83%), a palpable mass (58%). Acute pancreatitis is extremely rare 3, 5, 9, although elevation of the serum amylase may be seen in 57% of cases. Despite the large size of tumors, jaundice is present in only 33% of patients<sup>2</sup>, and cholangitis is extremely rare. Disease can sometimes be differentiated from adenocarcinoma by computed tomography scan and confirmed by fine needle aspiration. Other preoperative differential diagnosis includes gastric carcinoma, choledocholithiasis, other unspecified abdominal neoplasm. Surgical intervention may be necessary to confirm the diagnosis<sup>8</sup>.

Clinical and imaging findings usually are not specific, imaging studies often cannot differentiate pancreatic lymphoma from other pancreatic tumors, although a large well-circumscribed, homogeneous tumoral mass in the pancreas should raise the suspicion of lymphoma. In patients with primary pancreatic lymphoma, no marked pancreatic ductal dilatation or other alteration of Wirsung's duct is present even with ductal invasion; lymph node involvement below the level of the renal veins, as

another finding, should suggest the diagnosis. In patients with diffuse infiltration of the pancreatic gland without clinical signs of pancreatitis, the radiologist should be alert to the possibility of pancreatic lymphoma<sup>4,7</sup>.

It must be kept in mind that primary extranodal presentations should be investigated and treated like their nodal counterparts, in practice, the same guidelines are applied to the management of the two types.

Pancreatic lymphoma does not require surgical staging or a palliative Whipple's procedure before chemotherapy or radiation therapy. A better prognosis with nonsurgical treatment is additional impetus to search for secondary signs of primary pancreatic lymphoma<sup>10,11</sup>. On the other hand there are experience in the literature that suggests that surgical resection may play a beneficial role in the treatment of localized pancreatic lymphoma<sup>6</sup>. Koniaris relate a review of the pancreatic lymphoma in the English language literature where fifty-eight of these cases represented stage I or II lymphoma, which was treated without surgical resection with a 46% cure rate. Fifteen patients who had surgical resection for localized disease have been reported with a 94% cure rate<sup>6</sup>.

It is important to emphasize that intraoperative biopsies were fundamental for the diagnosis and prognosis. If treated as a patient with adenocarcinoma, probably the patient would not have this uneventful recovery.

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