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Case Report

Retroperitoneal germ cell tumor mimicking neoplasm of ampulla of Vater

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ABSTRACT

A 20-year-old man previously healthy, presented to the emergency department for evaluation of obstructive jaundice, backache and weight loss. Abdominal ultrasonography demonstrated a dilated common bile duct (up to 2 cm), increased gallbladder dimensions (10.3 x 4.9 cm) with biliary sludge. Computed tomography of the abdomen showed a retroperitoneal tumor with 18 x 14 cm dimensions, and numerous large retroperitoneal lymph nodes. The liver demonstrated a bilateral intrahepatic bile duct dilatation. An endoscopic retrograde cholangiography was performed. The study revealed an ulcerative neoplasm of ampulla of Vater, and the cholangiogram obtained images of dilated extra-hepatic and intra-hepatic bile ducts with a large stenosis in the distal third of the common bile duct. After biopsies were analyzed, the definite diagnosis was a retroperitoneal extragonadal germ cell tumor (subtype: embryonal carcinoma) with infiltrative invasion to duodenal wall. A testicular ultrasonography ruled out a testicular tumor. Endoscopic stenting was necessary for decompress the unresectable malignant bile duct obstruction. Patient was referred for cisplatinum based chemotherapy.

Keywords: Extragonadal germ cell tumor, Extrinsic bile duct strictures, Retroperitoneal germ cell tumor, Neoplasms of ampulla of Vater

INTRODUCTION

Bile duct stricture is a fixed narrowing of a focal segment of the bile duct that results in proximal biliary dilatation and clinical features of obstructive jaundice.[1] A wide spectrum of hepatobiliary and pancreatic diseases, both benign and malignant, can result in the development of biliary strictures.[1-3] Differentiating between a malignant and benign stricture is of paramount importance.[1,2]

Malignant bile duct strictures may be secondary to primary bile duct, pancreatic or duodenal neoplasms such as cholangiocarcinoma, pancreatic adenocarcinoma and ampullar and periamullary carcinomas. Extrinsic compression or invasion by periportal or peripancreatic lymphadenopathy, metastatic disease to bile ducts, gallbladder carcinoma may on occasion cause malignant stenosis.[1,2]

The imaging techniques to evaluate patients with obstructive jaundice are abdominal ultrasound, computed tomography, optical coherence tomography, and magnetic resonance cholangiopancreatography.[1,2] Endoscopic retrograde cholangiopancreatography (ERCP) is the most widely used endoscopic procedure in evaluating bile duct strictures.[2]

Other endoscopic technologies include endoscopic ultrasonography and intraductal ultrasonography. Cholangioscopy, which can be done either percutaneously or perorally, enables direct visualization of the bile duct strictures using a cholangioscope.[1,2]

ERCP with brush cytology and/or endobiliary forceps biopsy is undoubtedly the initial and most widely used method for the evaluation of bile duct strictures endoscopically.[2] ERCP provides means for biliary drainage and ablative therapy.[1,2]

Endoscopc stenting is the decompressive procedure of choice for unresectable malignant bile duct obstruction.[3] Endoscopic decompression offers lower overall cost, shorter hospitalization, and lower morbidity when compared with biliary-enteric surgical bypass.[3,4]

Here, we report an extremely rare case of a retroperitoneal extragonadal germ cell tumor presenting as obstructive jaundice and mimicking neoplasm of ampulla of Vater.

CASE REPORT

A 20-year-old man, previously healthy, presented to the emergency department for evaluation of obstructive jaundice. His medical history and family history were unremarkable.

He started in the previous 4 months with backache, that initially was relieved with over-the-counter analgesics. One month thereafter, he started with constitutional symptoms, remarkably he lost 15 kg in two months. He reported pale stools, dark urine and pruritus. The physical examination resulted in a patient with generalized jaundice, with lumbar and abdominal pain.

Initial laboratory data showed increased total leukocyte count (14.5 /µl), increased neutrophils (89.9%), normocytic anemia (Hemoglobin 8.3 g/dl; hematocrit 25.8%), and normal platelet count (350, 000). Coagulation tests: prothrombin time of 20.3 sec, international normalized ratio (INR) of 1.46, and a partial thromboplastin time of 30.7 sec.

Biochemical analysis revealed: increased serum glucose (240 g/dl), and pre-renal azotemia (creatinine of 2.19 mg/dl; ureic nitrogen of 81.3 mg/dl) and normal electrolyte count. The liver function tests showed: total bilirubin of 4.38 mg/dL, direct bilirubin of 2.73 mg/dl, alanine aminotransferase of 419 U/l, aspartate aminotransferase of 1515 U/l, gamma-glutamyl transferase of 144 U/l, and alkaline phosphatase of 546 U/l.

The values for the tumor markers were carcinoembryonic antigen (CEA; 30.7 U/mL), CA 19-9 (82.5 U/mL), alpha-fetoprotein (AFP; 1542 U/mL), beta-human chorionic gonadotropin (327 U/ml), and lactic deshidrogenase 4851 U/l. Immunology tests revealed negative serologies for B and C hepatitis viruses, and HIV tests were non-reactive.

Abdominal ultrasonography demonstrated a dilated common bile duct (up to 2 cm), increased gallbladder dimensions (10.3 x 4.9 cm) with biliary sludge and normal wall thickness (0.3 cm).

Computed tomography of the abdomen and thorax showed a retroperitoneal tumor with 18 x 14 cm dimensions, and numerous large retroperitoneal lymph nodes.

The liver demonstrated a bilateral intrahepatic bile duct dilatation. A large right pleural effusion was also found (Figure 1).

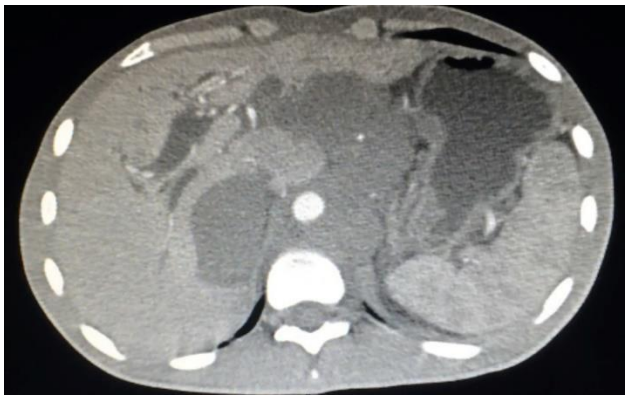

Figure 1: Abdominal computed tomography findings. A retroperitoneal tumor with 18 x 14 cm dimensions and bile duct dilation.

Due to the dilation of bile ducts he was taken to the endoscopy unit and an Endoscopic Retrograde Cholangiopancreatography (ERCP) was performed.

The study revealed an ulcerative neoplasm of ampulla of Vater, and multiple biopsies were taken (Figure 2).

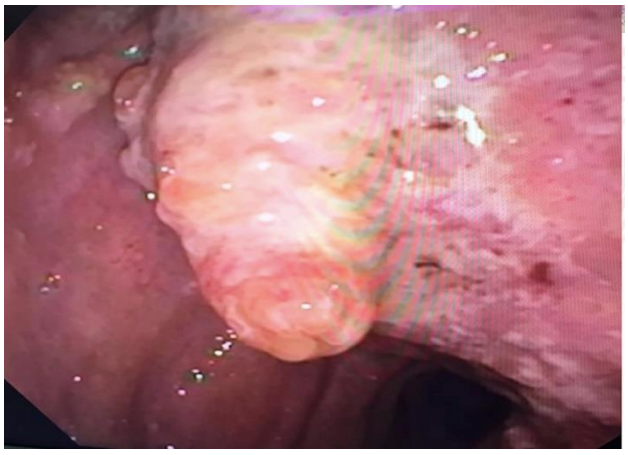

Figure 2: Endoscopy (ERCP) revealed neoplasm of ampulla of Vater.

After cannulation, the cholangiogram obtained images of dilated extra-hepatic and intra-hepatic bile ducts (up to 2.0 cm) with a large stenosis in the distal third of the common bile duct (Figure 3). A biliary stent was placed in order to decompress the bile ducts.

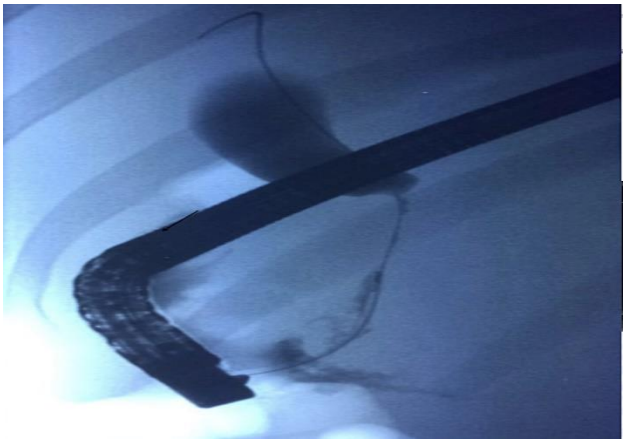

Figure 3: Cholangiogram findings. Stenosis in the lower third of common bile duct with proximal dilatation.

Histological examination (with hematoxilin and eosin) revealed the presence of malignant carcinoma cells. The mucosa presented ulcerative lesions, with acute and chronic inflammation. Lymphatic invasion was observed (Figure 4). Immunohistochemical examination revealed that the tumor was positive for CD30 (Figure 4).

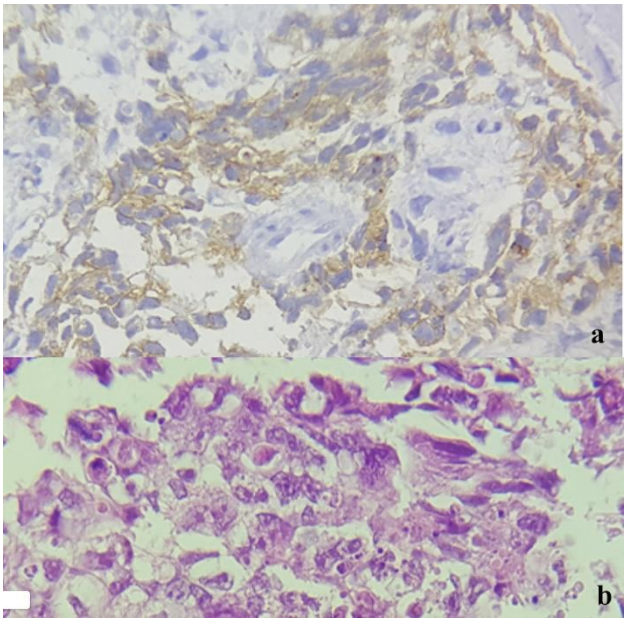

Figure 4: Histopathological and immunohistochemical sections of ampulla of Vater biopsy samples. a) tumor was positive for CD30 b) Malignant cells with carcinoma phenotype (HE, x20).

A testicular ultrasonography was performed and there was no evidence of a testicular tumor.

The definite diagnosis was an extragonadal retroperitoneal germ cell tumor (subtype: embryonal carcinoma) with invasion to duodenal wall. According with the International Germ Cell Cancer Collaborative Group (IGCCCG), the patients was classified as intermediate risk.[5] The patient was sent to chemotherapy (cisplatin-based), but unfortunately, he died a few weeks later.

DISCUSSION

Many disease processes, arising from primary or metastatic disease in intrahepatic, extrahepatic, or hilar locations, can lead to malignant biliary strictures.[3]

Identification of an ampullary mass, papillary bulging, irregular asymmetric luminal narrowing of the distal common bile duct, and diffuse proximal intra and extrahepatic biliary dilatation are signs of malignant ampullary obstruction.[1] Obstruction can ultimately lead to jaundice, pruritus, secondary biliary cirrhosis, cholangitis, coagulopathy, and weight loss trough malabsorption.[3]

Ampullary carcinoma is defined as carcinoma arising in the ampullar complex distal to the confluence of the pancreatic duct and common bile duct. Malignant tumors arising within 2 cm of the major duodenal papilla can be classified as periampullary carcinomas and include carcinoma of the ampulla of Vater, distal common bile duct, head of the pancreas and duodenum.[1] Biliary metastases are very rare and may cause strictures mimicking cholangiocarcinoma. They are commonly from primary cancers of the lung, gallbladder, breast and colon.[1,2]

Methods of tissue sampling during ERCP include intraductal bile aspiration cytology, cytopathologic analysis of retrieved plastic biliary stents, fine-needle aspiration cytology, brush cytology, and endobiliary forceps biopsy.[2,3] Combining cholangiographic impression with cytology and/or endobiliary forceps biopsy may offer sensitivity up to 85% and specificity up to 100%.[2]

The endoscopic goal is for adequate biliary drainage to palliate obstructive symptoms but also to limit the number of interventions in the patients' remaining life.[3] Although endoscopic treatment is the procedure of choice to biliary decompression, complications can arise from the procedure (perforation, bleeding, pancreatitis) or can be stent specific (occlusion, migration or infection).[3,4]

Extragondal germ cell tumors represent only 2-5% of adult germ cell malignancies. They mostly manifest in the mid axis of the body, the most common primary site is the anterior mediastinum (50-70%) followed by the retroperitoneum (30-40%).[6-8] Other localizations such as the pineal gland, the prostate, the urinary bladder, the liver and the sacrococcygeal region have been described occasionally.[8] To the best of the author´s knowledge this is the first case report of a patient with a retroperitoneal germ cell tumor presenting with obstructive jaundice, mimicking a tumor of ampulla of Vater.

These tumors histologically contain the same components as their gonadal counterparts, but no evidence of a primary malignancy is present in the testes by testicular high-resolution ultrasonography or physical examination.[6,7] So, these patients should have testis image (ultrasound) to rule out a testis primary.[6] They tend to be more aggressive than testicular germ cell tumors.[6,7]

Testicular and extragonadal germ cell tumors also share similar serologic features such as secretion of the tumor markers alpha-fetoprotein and beta-human chorionic gonadotropin.[7] Extragonadal germ cell tumors most likely originate from a urogenital misplacement of gonadal cells during embryogenesis.[7-9] A malignant transformation of these cells, depending on the influence of their microenvironment, leads to EGCTs with different histological features.[8] The distinctive genetic abnormality that is found in both tumors is an isochromosome i(12p).[6,7]

Retroperitoneal germs cell tumors represent 10% of all malignant primary retroperitoneal tumors.[6] The genesis of retroperitoneal germ cell tumors is still under debate.[8-10] A retroperitoneal location could represent a true extragonadal germ cell tumor or a tumor with primary origin in the testis that has metastasized and has turned into scarring tissue locally, the so called "burned out tumor".[8] If a true extragonadal tumor is suspected, it can occurs in the retroperitoneal parenchymatous organs (e.g., the pancreas, or kidneys) or as primary process of the retroperitoneum.[8,9] These tumors tend to grow slowly and produce few symptoms, so the majority are found bulky at presentation.[7] It has been reported that most patients with extragonadal non-seminomatous germ cell tumor have advanced disease at the time of presentation.[7,11]

Patients with retroperitoneal germ cell tumors may complain about abdominal pain (29%) or back pain (14%), weight loss (9%), fever (8%) and vein thrombosis (9%).[7,8]

The appropriate imaging methods to evaluate retroperitoneal germ cell tumors are ultrasound, computed tomography and magnetic resonance imaging. Imaging findings should be confirmed histologically.[8] The diagnostic protocol should include the germ cell-specific markers alpha-fetoprotein (AFP), human chorionic gonadotropin (β-HCG) and lactate dehydrogenase (LDH) to make a correct classification of the patient according to the IGCCCG.[7,8] Patients with a retroperitoneal non-seminomatous germ cell tumor are classified as having a good, intermediate or poor prognosis according to the IGCCCG criteria.[5,8]

For gonadal germ cell tumors vast recommendations for a stage-adapted therapy have been published, but for the treatment of extragonadal germ cell tumors only a limited number of appropriate evidence based tools exists.[5,8] The treatment of extragonadal germ cell tumors follows the same principle as those for gonadal tumors.[7,8] The therapy results of primary retroperitoneal germs cell tumors are similar to those of metastatic gonadal tumors.[8] However, survival of patients with extragonadal germ cell tumor, especially those with a non-seminomatous component, remains inferior compared with that of patients with advanced testicular cancer.[11] The therapeutical multimodal approach consist of cisplatinum-based chemotherapy and surgical resection.[5,8]

CONCLUSION

This is the first case report of a patient with a retroperitoneal extragonadal germ cell tumor (embryonal carcinoma) infiltrating duodenum and mimicking neoplasm of ampulla of Vater. Endoscopic stenting was necessary for decompress the unresectable malignant bile duct obstruction. Although a very rare entity, retroperitoneal extragonadal germ cell tumor should be included in the differential diagnosis of patients presenting with malignant extrinsic compression of the common bile duct.

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