Granular cell tumors (GCT) are rare benign tumors. Less than 1% of GCTs involve the extrahepatic biliary tree. Most researches favour a Schwann cell origin. Patient, caucasion, female, 31 year old presented with 4 month history of painless jaundice and pruritus. US and CT revealed dilatation of intrahepatic biliary tree and surgery was performed. Firm tumor mass was found above the conjunction of cystic duct and common hepatic duct (CHD) that caused obstruction and gallblader empyema. The patient underwent radical surgical procedure because Klatskin肿瘤 was clinically suspected. Pathohystology and immunohistochemistry confirmed granular cell tumor. Eight years after surgery the patient is wellbeing without symptoms. To our knowledge 69 cases of GCT of the extrahepatic biliary tree have been reported and none of the acute acalculous cholecystitis cases accompanied by GCT of CHD. Granular cell tumors are rarely diagnosed preoperatively. Wide resection offers the best chance for cure.

Key words: granular cell tumor, acute acalculous cholecystitis, biliary duct, jaundice

INTRODUCTION

Granular cell tumor (GCT) initially was described by Abrikossoff, in 1926. He suggested it could be of muscular origin and coined the term "myoblastic myomata". However, there are some reviews in which this tumor was described earlier. Some authors suggested a neurogenic origin. Most researchers favour a Schwann cell origin based on the histological, electron microscopic and immunohistochemical findings. The current literature supports the use of the noncommittal term, granular cell tumor. These rare tumors are nonmetastasizing and have a predilection in the dermal and subcutaneous tissues, especially within the oral cavity, chest wall and extremities, but may present at any location. Less than 1% of GCTs involve the extra-hepatic biliary tract. According to the literature this is the 70th published case.
Control cholangiography (10th operative day) through transintestinal drains was normal. (figure 3)

Eight years after initial surgical treatment patient is without signs of illness.

DISSCUSSION

A majority of the GCT reported cases involve females (91%) and Afroamericans (65%) with mean age of 34.7 years. The most common sites of the GCTs are dermis tongue, and subcutaneous tissues of the extremities and chest wall. Less common localizations include breast, larynx, gastrointestinal tract and vulva.

The first GCT of the extrahepatic biliary tree was reported in 1952 by Coggins and was found during autopsy. Boekhorst et al and Karakozis et al describe 53 and 57 cases respectively till 2000. To our knowledge 69 cases have been reported till now. Frequently reported anatomic sites in the biliary tree involved: common bile duct (28%), cystic duct (28%), common hepatic duct (24%), intrapancreatic bile duct (13%) and gall bladder (7%).

Five cases of multifocal GCTs of the biliary tract have been reported. Concomitant extrabiliary involvement was noticed in five cases only.

The patients present with abdominal pain, obstructive jaundice or both. The differential diagnosis of any obstructive lesion in the region of the hepatic duct confluence includes cholangiocarcinoma, sclerosing cholangitis or the more common benign biliary tumors, such as papillomas or adenomas, and should also include a biliary granular cell tumor. Report in the last decade demonstrated increasing de novo presentation of acute acalculous cholecystitis (ACC) in the absence of acute illness or traumatic injury.

In literature, ACC combined with GCT of hepatic bile duct was not reported till now except as complication of invasive procedures (ERCP, PTC, stenting). In this patient, intraoperative cholangiography was done and ACC could be explained by tumor spreading on cysticus that had relatively high conjuction and by occlusion of art. cystice that caused ischaemia. Karakuzis et al noted that the cystic duct tumors presented as reccurrent biliary colic and cholecystitis.

Granular cell tumors are rarely diagnosed preoperatively. In the report by Lack et al., only 3 of 110 GCST were diagnosed preoperatively. Intraoperative frozen section have been employed to establish exact diagnosis.

Macroscopically these granular cell tumors appear yellow-white and are usually less than 3 cm. The tumor is unencapsulated and consists of closely packed polyhedral cells with centrally placed uniform round nuclei, granular eosinophilic cytoplasm and absent mitotic activity. The histologic, electron microscopic, and immunohistochimical findings in these patients clearly indicate a nonepithelial origin with features suggestive of neuroectodermal origin. The coarse granules are strongly positive for PAS and the tumour is immuno-reactive to S-100, neuron-specific enolase, vimentin, cathepsin B and inhibin.

GCT is generally considered a benign tumor, but it can be malignant. That is very rare and has only been described in the skin. Until now, malignant, granular cell tumor has not been described in the biliary system.

Surgery remains the treatment of choice for this type of tumor locating in the extrahepatic biliary tract. Complete excision of the tumour with biliary reconstruction offers a cure. Hepaticojejunostomy is the most frequent and best choice. For tumours originated from the gallbladder or cystic duct, simple cholecystectomy is sufficient.
ple procedure is recommended where the intra-pancreatic portion of the bile duct is involved, although simple bypass or diversion of the neoplasm have also been reported for tumours in this location1,4.

Treatment of biliary granular cell tumor with stents, either percutaneous or endoscopic, other than for temporary biliary decompression is inappropriate in this curable disease.

In summary, GCT often present with initial symptoms of abdominal pain, jaundice or both. They clinically mimic cholangiocarcinoma or sclerosing cholangitis in a relatively young group of female patients. ACC could be a rare complication of GCT localized on common hepatic duct as in this patient. Wide resection with free margins is a curative treatment choice.

SUMMARY

Granular cell tumors (GCT) are rare benign tumors. On extrhepatitic bile ducts they are diagnosed at a median of 1% of all malignancies. They are quite often not diagnosed preoperatively.

Treatment of biliary granular cell tumor with stents, either percutaneous or endoscopic, other than for temporary biliary decompression is inappropriate in this curable disease.

In summary, GCT often present with initial symptoms of abdominal pain, jaundice or both. They clinically mimic cholangiocarcinoma or sclerosing cholangitis in a relatively young group of female patients. ACC could be a rare complication of GCT localized on common hepatic duct as in this patient. Wide resection with free margins is a curative treatment choice.

REFERENCES