Granular Cell Tumor of the Common Bile Duct : A Case Report

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Abstract

Granular cell tumors (GCTs) are uncommon soft tissue tumors, usually presenting in the skin and subcutaneous tissue tongue and oral cavity. We present a case report of granular cell tumor of the common bile duct involving both extra-and intrapancreatic portions. The histogenesis appears to be related to Schwann cells, similar to granular cell tumors of other sites, as evidenced by histologic and immunohistochemical findings. Review of the English literature concerning biliary tract GCTs revealed a high occurrence in African-American females in their third decade. By-pass operation to correct the biliary tract obstruction may be appropriate, if the nature of the tumor can be obtained from intraoperative diagnosis by frozen section.

Key word : Granular Cell Tumor, Common Bile Duct

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Granular cell tumors (GCTs), usually benign lesions, are relatively rare in occurrence, and the common sites are the skin subcutaneous tissue of the extremities and chest wall, tongue and oral cavity, while the less common sites include the breasts, larynx, stomach, vulva, anogenital region and intestine(1). GCT of the biliary tract is rare, and in the world literature, it most commonly occurs in relatively young African-American females in their third decade(2).

CASE REPORT

A 40-year-old Thai female presented in April 1999 with a 1-month history of jaundice pruritis and pale-colored stool. Ultrasonogram revealed dilation of both hepatic ducts. She underwent

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endoscopic retrograde cholangiopancreatography (ERCP), which showed marked stricture of the middle portion of the common bile duct causing marked dilation of the proximal bile duct (Fig. 1). Ultrafast CT scan of the abdomen showed mild dilation of intrahepatic bile ducts. Biochemical analyses of blood disclosed a 3 fold elevation of liver transaminases, bilirubin of 8.7 mg/dl (normal 0.2-1.2) and alkaline phosphatase of 1,068 IU/L (normal 27-86).

Clinically, adenocarcinoma of the common bile duct was diagnosed, so the Whipple's procedure was performed. After the operation, the patient was clinically improved and then discharged. The 1-month follow-up revealed no abnormal sign and symptom, and all abnormal biochemical findings had returned to normal limit.

Grossly, the operative specimen consisted of an en bloc specimen of duodenum with attaching head of the pancreas and common bile duct, and a separated gallbladder including cystic duct. There was a mural thickening of the common bile duct by an ill-defined encircling white-tan mass measuring 1.5 cm in thickness and 2.2 cm in length (Fig. 2A). The mass was located in both intra-and

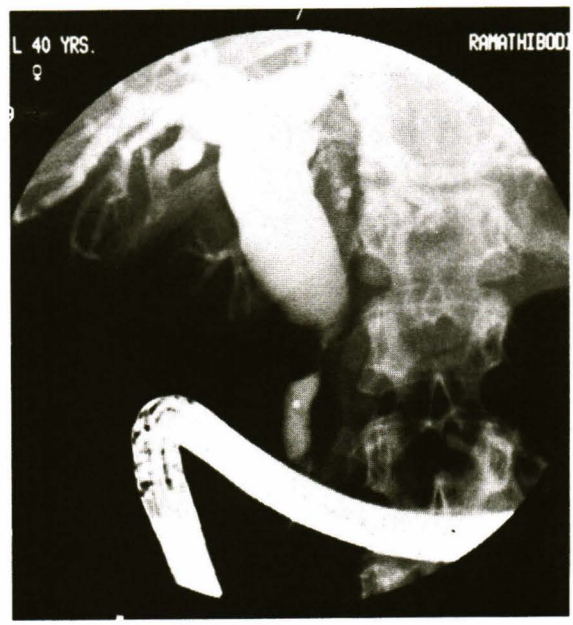

Fig. 1. ERCP showing marked stricture of the middle portion of common bile duct.

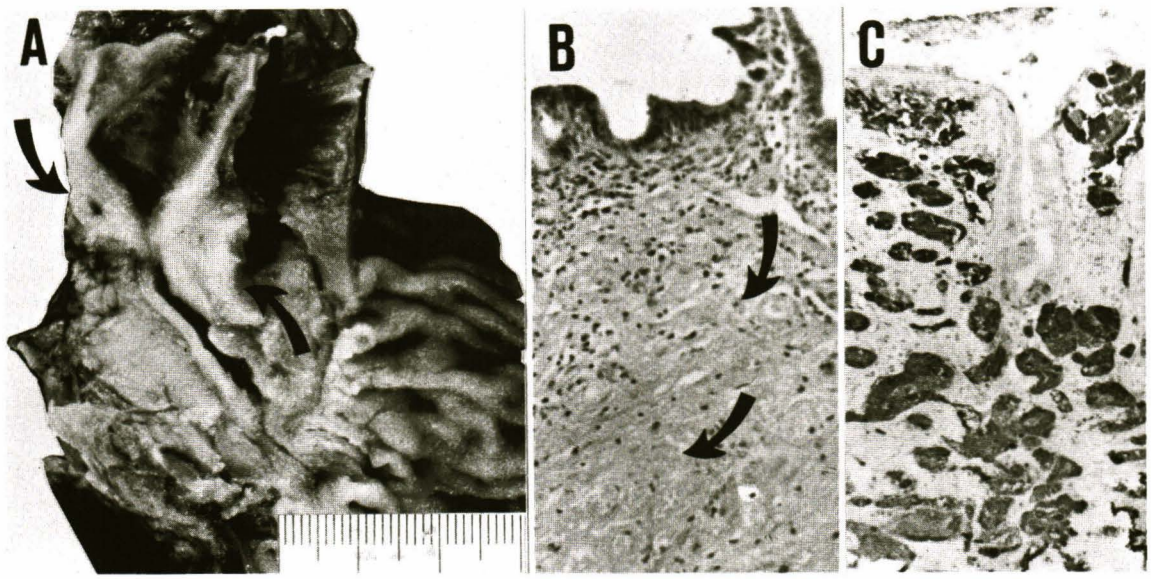

Fig. 2
A. Gross specimen showing localized intramural mass (arrows) encircling common bile duct.
B. Section of the tumor showing large irregular cells with pale eosinophilic granular cytoplasm (arrows). H&E, X150.
C. Strongly positive reactivity to S-100 protein in the cytoplasm of the tumor cells. S-100, X150.

extrapancreatic parts of the common bile duct, and 1.5 cm. from the surgical margin. The proximal common bile duct was dilated to a maximum diameter of 1 cm. The gallbladder and cystic duct were within normal limit.

Histologically, the low-power examination of the longitudinal section from the thickening part of the common bile duct disclosed the area of fibrosis with scattered large tumor cells. The bile duct mucosa appeared normal with focal ulceration. The higher power examination revealed slightly irregular tumor cells with abundant pale eosinophilic granular cytoplasms and relatively small round to oval vesicular nuclei without any significant atypism (Fig. 2B).

The further investigations included periodic acid-Schiff stain after diastase (D-PAS) showing highly positive reaction of the cytoplasmic granules, which were negative to von Kossa's stain for calcium. Immunohistochemically, the granular cells showed strong immunoreactivity to S-100 protein (DAKO, Fig. 2C) and negative immunoreactivity to lysozyme (DAKO) and CD 68 (Immunotech) markers for histiocytes.

DISCUSSION

GCT was originally named granular cell myoblastoma by Abrikossoff in 1926[3]. Muscle in origin was an early concept but it is no longer regarded as a likely possibility, and for this reason, the non-committed term granular cell tumor is preferred[4]. GCT of the biliary tree is relatively rare, compared to same tumors at other sites as previously mentioned. However, GCT is one of the most frequent benign nonepithelial tumors of the extrahepatic biliary ducts[5]. At this site, both epithelial and nonepithelial benign tumors are one-tenth as common as extrahepatic bile duct carcinomas[6]. The first case report of biliary tract GCT was presented by Coggins in 1952,[3] thus far, there are 58 case reports in the world literature[2]. The GCT occurrence includes intra-and extrahepatic biliary system,[7] but common bile duct is the most frequent site of involvement, followed by cystic duct and common hepatic duct[2].

Our patient's sex and age are comparable to those in the literature review. That is, GCTs involving the biliary tract are predominantly found in relatively young (3rd decade) African-American females[2]. Clinical manifestation of jaundice and radiologic examinations by ultrasonography and endoscopic retrograde cholangiopancreatography showing dilated proximal bile ducts are typical of tumors involving the lower biliary tract requiring surgery[1].

On gross examination, GCT of the common bile duct can be classified into two distinct patterns. The less frequent is a diffuse encasement of a large part of the bile duct by dense fibrous tissue. This process frequently extends into the cystic and hepatic ducts. The more common, as seen in our case, is a localized fusiform mass involving the common duct in an encircling fashion, ranging in size from 1.0 to 3.5 cm[8]. The mass has been described as firm, rubbery to hard, gritty and even so hard as to be mistaken for a calculus[9]. About 15 per cent of GCTs at other sites are multifocal, but only rarely are they multifocal within the extrahepatic biliary tracts[6]. From one literature review of the intraoperative diagnosis by frozen section, none of the diffuse type could be diagnosed correctly, contrary to the localized type[8].

Histologically, the GCT of the biliary tree resembles those in the other sites. The typical features are nests or sheets of large irregular granular eosinophilic cells with small centrally located vesicular nuclei. Large granules may be seen in the hematoxylin and eosin sections, however, D-PAS staining usually highlights the presence of the granules[1]. Fibrosis may be prominent in long-standing cases[1,2]. Exceedingly rare malignant changes are reported, representing only 1 to 2 per cent of all GCTs. Clues to the diagnosis of malignancy are large tumor size (> 4-5 cm), numerous mitoses (>2/10 high power fields), necrosis, and distant metastasis, thus far, no malignant change or recurrence has been demonstrated in biliary tract GCT[1,4]. Differential diagnoses of GCT include fibroxanthoma, malakoplakia, reactive granular cell changes in association with trauma, and rhabdomyoma. The marked reactivity of tumor cells to S-100 protein indicate tumor cells originating from or differentiating towards Schwann cells[1,3]. While the negative stainings to lysozyme and CD68 markers for histiocyte rule out the diagnoses of fibroxanthoma and malakoplakia. Moreover, the absence of intracytoplasmic Michaelis-Gutmann body, confirmed by negative reactivity to von Kossa's stain for calcium, is helpful in exclusion of malakoplakia[10,11]. Absence of previous surgery or trauma in our case excluded reactive granular cell changes[12] and the lack of strap or tadpole

cells with absence of cross striation also excluded rhabdomyoma[4].

Treatment of biliary tract GCT consists of complete surgical excision with biliary tract reconstruction[2]. Hepaticojejunostomy or choledochojejunostomy may be performed, depending on the site of involvement. In the literature, there are four reported cases of Whipple's procedure performed in GCT of the intrapancreatic and/or distal common bile duct similar to our case[2,8]. A simpler procedure such as by-pass operation may be an alternative choice for surgeons if the nature of the tumor is known from the intraoperative frozen section.

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แกรนูลาร์ เซลล์ ทูเมอร์ ของท่อน้ำดีรวม : รายงานผู้ป่วย 1 ราย

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แกรนูลาร์ เซลล์ ทูเมอร์ (Granular cell tumor) เป็นเนื้องอกที่พบได้น้อย มักเกิดบริเวณผิวหนัง ชั้นใต้ผิวหนัง ลิ้น และช่องปาก รายงานผู้ป่วยนี้นำเสนอแกรนูลาร์ เซลล์ ทูเมอร์ ของท่อน้ำดีรวมทั้งส่วนที่อยู่ภายนอกและภายในตับอ่อน เชื่อว่าต้นกำเนิดของเนื้องอกชนิดนี้คือ เซลล์หุ้มปลายประสาท (เซลล์ชวานน์) โดยมีหลักฐานสนับสนุนคือ ลักษณะของเนื้อเยื่อทางกล้องจุลทรรศน์และผลการย้อมพิเศษรวมถึงปฏิกิริยาภูมิคุ้มกันของเนื้อเยื่อ จากการศึกษาพบว่า กรานูลาร์ เซลล์ ทูเมอร์ ของท่อน้ำดี พบได้บ่อยในหญิงชาวอาฟริกัน-อเมริกัน ที่มีอายุอยู่ในช่วง 30-40 ปี และหากทราบผลการวินิจฉัยเนื้องอกนี้โดยวิธีโฟรเซนเซ็คชั่น (frozen section) ในระหว่างการผ่าตัด อาจให้การรักษาโดยวิธีเบี่ยงทางเดินน้ำดี (by-pass operation) ก็น่าจะพอเพียง

คำสำคัญ : แกรนูลาร์ เซลล์ ทูเมอร์, ท่อน้ำดีรวม

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