Mucinous biliary obstruction associated with a cholangiocarcinoma

Kolanjiyokansere bağlı musinöz biliyer obstrüksiyon

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SUMMARY: *A 60 year old who man had repeated episodes of bile duct obstruction and cholangitis caused by a solitary tumor in the left hepatic duct was presented. Characteristic features of the tumor were profuse secretion of mucus and choledochus dilatation distal to the tumor. The tumor initially appeared to be benign but showed malignant transformation within three years. He is surviving under our care following surgical left hepatic resection.*

Key words: Mucin, bile duct tumor

ÖZET: *Sol hepatik kanalda lokalize tümöre bağlı tekrarlayan obstrüksiyon ve kolanjit bulguları gösteren, 60 yaşında bir erkek hasta sunuldu. Tümörün özelliği, aşırı müsin üretmesiydi. Buna bağlı olarak tümöre distal koledok da genişlemişti. Önceleri benign gözüken tümör, 3 yıl içinde malign değişim gösterdi. Sol hepatik lobektomi uygulanan hasta, halen kontrolümüz altında yaşamını sürdürmektedir.*

Anahtar sözcükler: Musin, safra yolu tümörü

Bile duct carcinoma (cholangiocarcinoma) occurs with equal frequency in males and females. The averege age of presentation is 60 years. An increased incidance of this disease has been reported in patients with ulcerative colitis, sclerosing cholangitis, clonorchis sinensis infestation and in a variety of congenital dilatations of the bile duct (Caroli's disease, congenital hepatic fibrosis, polycystic disease) (1).

Grossly, bile duct carcinomas can be polypoid and superficial, but most are nodular or sclerosing, with deep penetration into the wall. Occasionally, they are multicentric and/or associated with carcinoma of the gallbladder. Microscopically, the large majority of bile duct malignancies are well-differentiated mucin-secreting adenocarcinomas (1, 2). Mucin production is one of the characteristics of these tumors, but usually only small amounts are produced. There are, however, some cases of cholangiocarcinoma producing large amounts of mucin. In these cases, mucin rather than the tumor itself plays an important role in the clinical course. We present a case of solitary adenocarcinoma of the left hepatic duct that posed two problems. Firstly, this neoplasm repeatedly caused obstruction of the common bile duct and cholangitis due to mucus secretion. Secondly, although appearing histologically benign at initial biopsy, histological examination after left hepatic lobectomy revealed adenocarcinoma.

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CASE REPORT

A 60 year old man was referred to our clinic in July 1993 complaining of recurrent episodes of abdominal pain, nausea, vomiting and jaundice accompanied by chills and fevers. The patient had attended another hospital six months prior to this admission with the same complaints but no cause was found for cholestasis and there was no pathological finding on laparotomy with only a drainage procedure being perfomed.

On physical examination, the patient was icteric. Abdominal examination did not revealed any significant finding. Laboratory data on admission were within normal limits except for a slight increase in transaminases (AST: 42 Ü, ALT: 73 Ü) and bilirubins (Bilirubin direct: % 0.7mg, B. indirect % 2.4 mg). Ultrasound and CT showed dilated extrahepatic and left intrahepatic bile ducts. Endoscopic retrograde cholangiogram revealed an

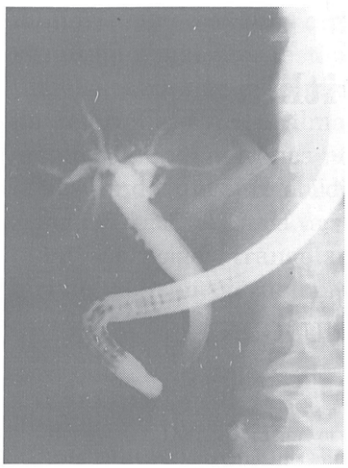

Figure 1. Doubtful filling defect in the left hepatic duct and intra-extrahepatic bile duct dilatation on ERCP

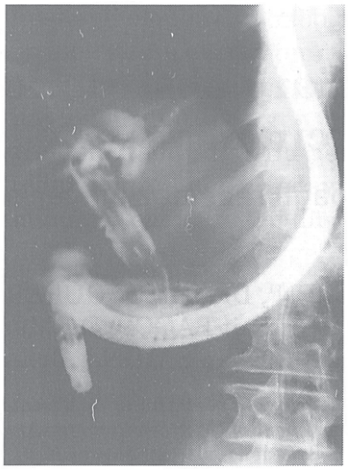

Figure 2. Amorphous filling defects in choledochus due to mucin on ERCP.

unclear filling defect at the left hepatic duct and intra-extrahepatic duct dilatation (Figure-1).

Papillotomy was performed. The choledochus was cleaned with a balloon catheter and a nasobiliary catheter was placed. Brush and bile cytology were negative.

The patient had been well until December 1994 but he was readmitted in June 1994 with obstructive jaundice. Ultrasound (US) revealed multiple stones in the gallbladder and drainage procedures were performed again. Cholecystectomy with choledochus exploration and operative cholangioscopy were perfomed at this time. But no distinct pathology could be found in the duct.

In June 1995, ERCP showed a large, smooth, amorphous filling defect within the common bile duct (Figure 2). Gelatinous mucus was removed by balloon catheter and a mucin secreting tumor was considered for the first time. A retrograde cholangioscopy was then performed at Yüksek İhtisas Hospital and papillomatous tumor about 1 cm in size was found in the left hepatic duct (Figure 3). Biopsies taken with standart forceps showed villous adenoma.

In March 1996, left hepatic lobectomy was performed and histological examination revealed adenocarcinoma and biliary cirrhosis (Figure 4).

The patient has been surviving symptom free six months after the last operation.

DISCUSSION

Mucin-producing adenocarcinomas of the bile duct are rare. In a series of cases reported by Nakajima et al (3), there was only one mucinous carcinoma among 102 consecutively examined cases of intrahepatic cholangiocarcinoma. On the other hand, there is different types of mucin producing cholangiocarcinoma with intraductal spreading, with most affected patients showing obstructive jaundice caused by mucinous compaction of the common bile duct. Here, we described a patient with mucinous cholangiocarcinoma which secreted mucin into the choledochus. Such patients may present with pain, obstructive jaundice or sepsis. Intermittent symptoms are due to obstruction.

Neoplastic obstruction of the common bile duct is a frequently encountered problem in gastroenterology and the mechanism of bile duct obstruction is generally related to direct invasion by the tumor. The duct may also be externally compressed by the tumor mass itself or by lymph nodes. Several additional mechanisms have been proposed for the intermittent jaundice often seen in ampullary lesions, which include a ball-valve effect, sloughing of cells from the tumor surface and invasion of the nerves of the wall of the bile duct. With all of these mechanisms, biliary obstruction relates to the tumor size, extent or location within the biliary system (4-6).

In this report, thick gelatinous mucin produced by a nonobstructing neoplasm was the mechanism of biliary obstruction. There have been very few reports of this type of biliary obstruction (4, 7). It is important to recognize that the obstructing mucus may be either proximal or distal to the neoplasm, leading to confusion regarding the location of the tumor.

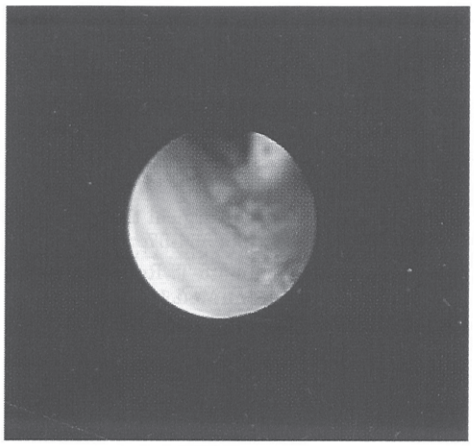

Figure 3. Papillomatous tumor in the left hepatic duct at cholangioscopy

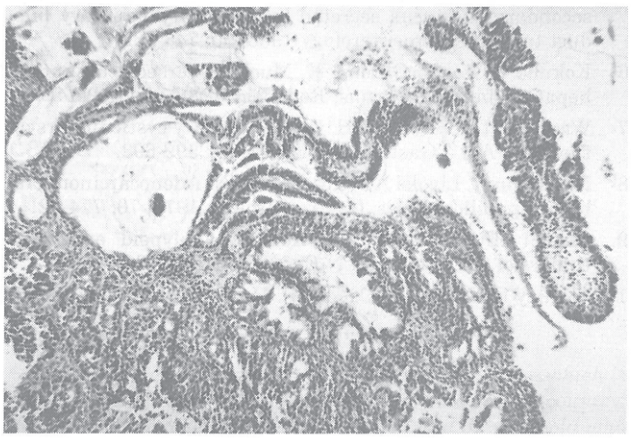

Figure 4. Histological appearence of adenocarcinoma in the operation specimen obtained after lobectomy.

Ultrasound and CT are initial diagnostic procedures with the most characteristic appearance of mucin hypersecreting biliary neoplasms on CT and US being marked dilatation of the intra-and extrahepatic bile ducts distal to the tumor. On cholangiograms, filling defects indicating retention of mucin (large, amorphous defects) in the dilated bile ducts are characteristic. In most cases, CT and US can not differentiate mucin from bile. However, echogenic spots in the dilated duct on US or high attenuation on CT are helpful in suggesting the retention of mucin. The presence of a tumor is another important finding, but they generally not large enough to visualize, as in our case. Cytological procedures (fine-needle aspiration, brushing or bile aspiration) yield diagnostic material in about 70 % of cases (8). Symptoms that arise from these tumors may be present for many years and the diagnosis can be missed. Caroli described a patient who had symptoms for 37 years (9, 10, 11, 12). Clinical symptoms for three years in the patient reported here suggest that this neoplasm may have originally been benign.

Carcinoma of the bile ducts carries a grave prognosis and surgery offers the only hope for such cases as well as the best form of palliation. Percutaneous or endoscopically placed tubal drainage of the biliary tract is becoming more common either for relief of obstruction preliminary to definitive surgery or as definitive treatment for unresectable tumors. In either case, there are hazards as well as benefits and there is no trial which has compared catheter drainage with surgical treatment. Infections introduced by the catheter, hemorrhage and perforation are serious complications. There is a role for these techniques, however, particularly in patients who are not candidates for surgery.

The overall five year survival rate is only 5 %. with death commonly occuring as a result of unrelieved ductal obstruction complicated by sepsis (12-15).

In summary, the case reported is an extremely rare instance of a patient in whom a small solitary neoplasm of the bile duct produced mechanical obstruction and cholangitis secondary to mucus secretion. Although initially appearing to be benign, adenocarcinoma was subsequently diagnosed. This case illustrates, however, that even with recurrence of symptoms, if adequate drainage is provided, the course can remain relatively uncomplicated for a long time.

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