LETTER TO EDITOR

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Intrahepatic Biliary Cystadenoma with Common Bile Duct Obstruction and Gallstone Formation

Gruczolak torbielowaty wewnątrzwątrobowych dróg żółciowych przebiegający z niedrożnością przewodu żółciowego i kamicą

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Biliary cystadenoma (BCA) is a rare condition. There have been less than 150 cases reported in the English literature, mostly in middle-aged women and not more than 10 cases in men [1]. The majority of cystadenomas arises from the intrahepatic duct and are usually asymptomatic [3]. Extension of an intrahepatic BCA into the common bile duct is rare [4], as is intrahepatic BCA causing jaundice from bile duct obstruction [5]. We present here a case of obstructive jaundice in a young woman caused by a multilocular BCA, arising in the left biliary tree with a pedunculated polypoid protrusion in the common hepatic and the common bile duct causing intraluminal obstruction.

Case History

A 24 year old female patient was admitted with jaundice. She had been complaining of upper abdominal pain, tiredness and pruritus. On admission she was in good physical condition, and physical examination revealed no abnormality. Blood examination showed normal blood count and LDH but an elevated ASAT of 260 (normal ≤ 35) u/l, alkaline phophatase of 724 (normal 80–275) u/l and bilirubin of 135 (normal 4–22) μmol/l. Screening for Ebstein Barr virus, Cytomegalovirus, HAV, HBV, HBC, CEA, alfa-fetoprotein and salmonella were all negative. A liver biopsy showed morphology consistent with large duct obstruction. Ultrasonography of abdomen revealed dilated intra- and extrahepatic bile ducts and a multilocular cyst measuring 35 × 25 mm in the fourth hepatic segment and endoscopic retrograde cholangiography (ERC) showed a polypoid configuration in the common hepatic duct. Computed tomography (CT) confirmed the presence of a cystic process in the left liver lobe. Ultrasonically-guided fine needle biopsy of the cyst wall was performed and morphologically fibrous tissue with inflammatory changes was found. Cyst puncture and intracystic injection of contrast medium did not reveal any communication to the biliary tree. Cytology of cyst fluid was benign on cytological examination.

Operation was performed because of recurrence of the cyst after aspiration. At laparotomy a multilocular cyst measuring 4 cm in diameter was found in the anterior part of the fourth liver segment (Fig. 1). Choledochotomy with extension into the common hepatic duct was done and a polypoid process in the common bile duct, extending into the common hepatic duct and attached by a stalk to the left biliary tree was seen. The common bile duct in addition contained two very small pigmented concrements. The tumour bearing part of the fourth liver segment was resected and the polypoid process in the common bile duct was removed. Choledochotomy was closed over a T-tube. Postoperative course was uneventful and the T-tube was removed. The clinical control after one year showed no evidence of recurrence.

Gross Pathology

The intraluminal part of the tumour measured 1.5 × 0.75 × 0.75 cm, appeared with a smooth surface and a cystic cut surface. The cystic process

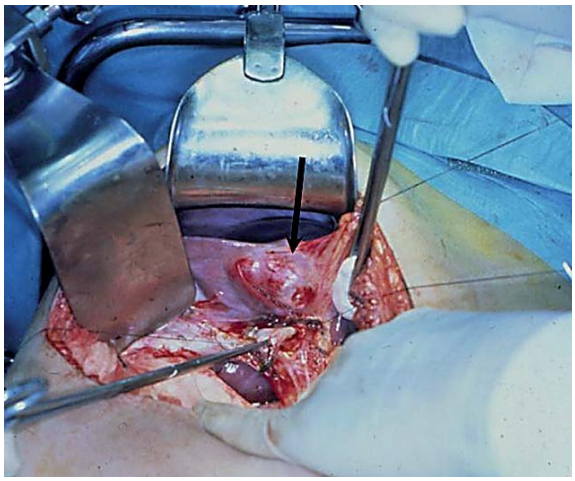

Fig. 1. Intraoperative appearance of the cystic tumour (arrow) in the fourth liver segment

Ryc. 1. Śródoperacyjny obraz guza torbielowatego (strzałka) w IV segmencie wątroby

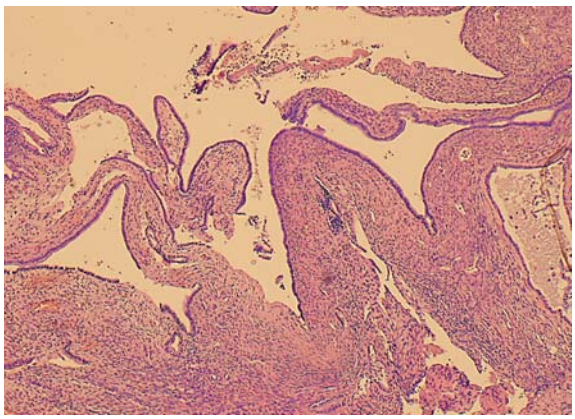

Fig. 2. H&E section of tumour showing multiple cysts lined by a uniform one-layered epithelium resting on a cellular stroma (Original amplification ×40)

Ryc. 2. Mikroskopowy obraz wielotorbielowatego guza wyścielonego jednorzędowym nabłonkiem pokrywającym podścielisko (barw. HE, pow. 40×)

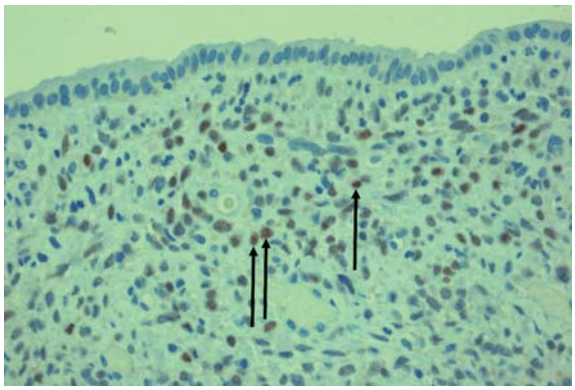

Fig. 3. Cystwall with a cellular stroma revealing estrogen positive nuclei (arrows) (Original amplification ×200)

Ryc. 3. Ściana torbieli wraz z podścieliskiem ukazująca dodatnią reakcję estrogenową jąder komórkowych (strzałki) (pow. 200×)

from the liver consisted of a multilocular cystic specimen partly covered by peritoneum and was well demarcated from the surrounding liver tissue.

Light Microscopy

Numerous sections were taken from the tumour and stained with H&E. In addition deparaffinized sections from two areas were stained conventionally for estrogen receptor (Immunotech, Clone: 1D5, Dilution: 1 : 100). The tumour consisted of multiple cysts outlined by a cuboidal epithelium without dysplasia. The cells were resting on a fibrovascular stroma which in many areas appeared cellular with an ovarian like stroma (Fig. 2 and 3). Most of the nuclei were positive for estrogen receptor (Fig. 3).

Discussion

Biliary cystadenomas (BCA) are rare. BCA are mostly intrahepatic and extrahepatic location is rare [6]. To our knowledge there have only been 4 reports of luminal bile duct obstruction causing jaundice from intrahepatic BCA, 3 of which were due to protruding polypoid masses [4, 5, 7] and 1 because of intracystic haemorrhage [8]. The exact tissue of origin of cystadenomas in the biliary tree is unknown. Tumour may arise from ectopic rests of embryonal gallbladder or may be a consequence of an acquired condition due to obstruction of congenitally aberrant bile ducts [9]. Because of the morphological similarity to ovarian stroma some have suggested development from ovarian tissue [2]. Based on the fact that estrogenic receptors have been found in the stroma of cystadenomas, estrogen has been suggested as a stimulus to the development and growth of cystadenoma. Furthermore the female predominance suggests that estrogen could be of importance [10].

Histologically cystadenomas are divided into two groups, depending on the presence or absence of mesenchymal stroma. Those with mesenchymal stroma are found exclusively in women [9]. Cystadenomas can undergo malignant change (cystadenocarcinoma, sarcoma) [9]. The presenting symptoms are discomfort or pain in the upper right abdominal quadrant or in the epigastrium. Abdominal mass is an occasional finding. Other unspecific symptoms include anorexia, tiredness, common nausea, vomiting and weight loss [1, 4]. Symptoms of bile duct occlusion may occur either due to intraluminal obstruction of extrahepatic ducts or compression of bile ducts due to growth of the cyst [4]. Cystadenomas can be completely

symptomless and found incidentally during operation for other reasons [2]. There is a long list of differential diagnosis. The most important are hamartomas, solitary congenital cysts, polycystic disease, haematomas, traumatic cysts, liver abscess, necrotic neoplasm, metastases and Caroli´s disease [3]. The most important diagnostic tools are abdominal ultrasonography and CT scan, which usually show multilocular cysts [5]. The presence of calcifications in the cyst wall or septae may suggest malignancy.

In the past BCA has been treated by aspiration, marsupialization, or partial excision. All these procedures have a high recurrence rate. Complete resection is currently the treatment of choice as it carries the lowest recurrence rate [3].

References

[1] Kazama S, Hiramatsu T, Kuriyama S, Kuriki K, Kobayashi R, Takabayashi N, Furukawa K, Kosukegawa M, Nakajima H, Hara K: Giant Intrahepatic Biliary Cystadenoma in a Male: a Case Report, Immunohistopathological Analysis, and Review of the Literature. Dig Dis Sci 2005, 50, 7, 1384–1389.

[2] Lei S, Domenico DR, Howard JM: Intrahepatic Biliary Cystadenoma With Intracystic Gallstone Formation. HPB Surg 1994, 7, 3, 241–247.

[3] Florman SS, Slakey DP: Giant Biliary Cystadenoma: Case Report and Literature Review. Am Surg 2001, 67, 8, 727–732.

[4] Sutton, CD, White SA, Berry DP, Dennison AR: Intrahepatic Biliary Cystadenoma Causing Luminal Common Bile Duct Obstruction. Dig Surg 2000, 17, 3, 297–299.

[5] Preetha M, Chung AY, Lim-Tan SK, Lim DT, Thng CH: Intrahepatic Biliary Cystadenoma Presenting With Obstructive Jaundice. Asian J Surg 2004, 27, 3, 243–245.

[6] Davies W, Chow M, Nagorney D: Extrahepatic Biliary Cystadenomas and Cystadenocarcinoma. Report of Seven Cases and Review of the Literature. Ann Surg 1995, 222, 5, 619–625.

[7] Beretta E, De Franchis R, Staudacher C, Faravelli A, Primignani M, Vecchi M, Conti E, Di Carlo V: Biliary Cystadenoma: an Uncommon Cause of Recurrent Cholestatic Jaundice. Am J Gastroenterol 1986, 81, 2, 138–140.

[8] Taketomi A, Tamada R, Takenaka K, Kawano R, Maeda T, Sugimachi K: A Case of Biliary Cystadenoma With Obstructive Jaundice. Oncol Rep 1998, 5, 4, 833–835.

[9] Wheeler DA, Edmondson HA: Cystadenoma With Mesenchymal Stroma (CMS) in the Liver and Bile Ducts. A Clinicopathologic Study of 17 Cases, 4 With Malignant Change. Cancer 15, 9, 1985, 56, 6, 1434–1445.

[10] Akwari OE, Tucker A, Seigler HF, Itani KM: Hepatobiliary Cystadenoma With Mesenchymal Stroma. Ann Surg 1990, 211, 1, 18–27.

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