CASE REPORTS

18-YEAR OLD PATIENT WITH EXTRAHEPATIC BILIARY DUCT CARCINOID – CASE REPORT

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Most patients with obstructive jaundice caused by an etiology other than choledocholithiasis are at risk of adenocarcinoma development. Other types of tumors are less common, although more benign. The presented study described management in case of liver hilar tumors with special regards to common hepatic duct carcinoids.

Key words: biliary duct tumors, choledocholithiasis, neuroendocrine tumors, mechanical jaundice

Carcinoid tumors, according to the new definition proposed by WHO in 2000, are classified as neuroendocrine tumors. They are derived from endocrine cells. Carcinoids account for 50% of all gastrointestinal neuroendocrine tumors (1). The above-mentioned are located in the small bowel – 41.8%, rectum – 27.4%, lungs – 25.3%, and only 0.1-0.2% are observed in the extrahepatic biliary ducts (2, 3). Due to the large number of pancreatic tumors and biliary duct carcinoma, carcinoid diagnosis is rarely considered during differential diagnostics of mechanical jaundice (4). However, it is essential when planning proper treatment, and determining prognosis. Unfortunately, diagnosis is rarely recognized on the basis of preoperative examinations, and proper diagnosis is established intraoperatively, or after histopathological evaluation.

Due to the patients’ young age and coexistence of an additional pancreas in the biliary ducts, the presented case is atypical, and according to our knowledge a similar case has not been described, thus far. The authors believe that a short description considering the management of rare biliary duct tumors will bring knowledge concerning the tumor.

CASE REPORT

An 18-year old male patient – P.K. (history number 24607/2012) was admitted to the Department of General Surgery, Western Hospital in Grodzisk Mazowiecki, June 2012, with diagnosis of liver hilar tumor. In May, 2012 the patient was hospitalized at the Department of Internal Diseases, due to jaundice and itching of the skin, without other symptoms. Abdominal ultrasound, computed tomography (fig. 1) and cholangio-MR (fig. 2) were performed confirming the diagnosis of liver cancer located below the right hepatic duct, 32x23 mm in size, common hepatic duct dilatation – 18 mm, enlarged gall-bladder without deposits, small cyst in the 7/8 liver segment, and an additional spleen. The patient was subjected
to ERCP with the implantation of two stents. The examination showed distal stenosis (3 cm) of the common hepatic duct, its morphology corresponding to the compression of a lesion from the outside (fig. 3). On admission, the patient was in good general condition, without symptoms. Family history was negative.

Laboratory results were as follows: WBC – 6.4x10^3/µl; RBC – 5.16x10^6/µl; MCHC – 35.0 g/dl; PLT – 255x10^3/µl; INR 1.02; APTT 26.6 s; prothrombin time – 3.4 s; index – 98.5%; complete bilirubin – 1.2 mg/dl; incomplete bilirubin – 0.3 mg/dl; Na – 138 mmol/l; K – 4.63 mmol/l; urea – 17 mg/dl; creatinine – 1.00 mg/dl; AST – 24 U/l; ALT – 41 U/l; CRP – 0.5 mg/l; neoplastic markers: CA 19-9-28.55; CEA 3.4 ng/ml; AFP – 0.74 IU/ml.

The patient was qualified for surgery with initial diagnosis of hepatic hilar tumor. The intraoperative examination revealed the presence of gall-bladder distention. At the site of the biliary duct a solid infiltration, 3-4 cm in diameter, covering the common hepatic duct (fig. 4) was observed. After puncture and gall-bladder content evacuation the patient underwent cholecystectomy. After hepatoduodenal ligament vascular preparation, the common biliary duct was isolated and severed. The prostheses were removed. The pe-
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The peripheral segment was ligated. The biliary ducts were prepared. Macroscopically, the common hepatic duct was normal, located 1 cm below the biliary outflow. The biliary ducts and tumor were excised followed by Roux-Y loop anastomosis, isolated PDS 4/0 sutures were used. A separate incision was required for biliary drainage. No anastomotic splinting was performed.

The early postoperative period was uneventful, postoperative wound healing was normal. Six days after surgery the biliary drain was removed. The patient was discharged from the hospital on the 11-th postoperative day with recommendation of surgical and oncological control.

The histopathological result (no 100448464) was as follows: well-differentiated neuroendocrine tumor – NET G1 [(carcinoid), CD 56 (+); chromogranin (+); synoptophysine (+) in neoplastic cells Ki-3%], infiltrating the gall-bladder wall from the outside; four lymph nodes with follicular hyperplasia and sinus histiocytosis (fig. 5, 6). The histopathological sample showed the presence of normotypic pancreatic parenchyma (fig. 7). Macroscopically, one observed a necrotic gall-bladder (12 cm) with a clear, solid tumor, 3.3x2.5x2.3 cm in size, involving the neck and part of the body of the gall-bladder.

**DISCUSSION**

Carcinoids are tumors, which are derived from neuroendocrine cells. Currently, they are diagnosed in 3 per 100 000 patients. This might be associated with progress in imaging diagnostics, considering the fact that in case of pancreatic autopsy examinations, neuroendocrine tumors are diagnosed in 10% of patients (5). Typically, they are diagnosed in patients after fifty, more often in women (6). Therefore, the presented 18-year old male patient without a positive family history is, according to our knowledge, one of the youngest patients with such a tumor location.

Typical location of the carcinoid is the appendix, small bowel, rectum, and lungs. The described location of the tumor in the extrahepatic biliary ducts is atypical. This is associated with the fact that carcinoids are derived from Kulchitsky’s cells, which are scarce in the biliary ducts (7). The first patient with such a location was described by Pilz in 1961 (8), and our literature review showed some 80 similar cases (9, 10). It should be noted that the histopathological examination revealed the existence of a small (5 mm), ectopic pancreas in the vicinity of the hepatic duct. This location is atypical, and thus far, only less than 10 such cases were mentioned (11). It was not associated with the carcinoid, although considering the presence of an ectopic pancreas, the pos-
sibility of carcinoid development should be considered.

In most cases the major symptom of extrahepatic biliary duct carcinoid is jaundice, although the tumor was diagnosed in selected cases during diagnostics of right upper quadrant or back pain. Isolated cases of carcinoid diagnosis were also described during gastrointestinal bleeding diagnostics (12), acute pancreatitis (13), and in several cases, incidentalomas were observed. Other unspecific symptoms accompanying carcinoid diagnosis include nausea, vomiting, weight loss, and general weakness. 10% of patients are diagnosed with carcinoid syndrome (14), however, it has not yet been described in case of biliary duct carcinoids. Our study patient only complained of jaundice with itching of the skin.

Diagnostic imaging included the ultrasound examination, which showed biliary duct expansion, and computed tomography, which enabled to visualize the presence of a hilar tumor. The MR of the biliary ducts provided further details concerning the size and location of the tumor. In doubtful cases endoscopic ultrasonography might prove helpful, which may be combined with a biopsy. In clinical practice, the above-mentioned is poorly accessible and has no influence on the therapeutic plan. Endoscopic retrograde cholangiopancreatography (ERCP) is of great value offering therapeutic options, such as stenting and possibility of a biopsy. The latter was performed showing significant stenosis of the common hepatic duct, followed by biliary duct prosthesis implantation. The biopsy was not performed, due to the possibility of external compression of the lesion. Unfortunately, preoperative histopathological diagnosis is rare (<15%) (15), which is associated with difficult access and submucous location of the tumor.

Cholangiocarcinoma (90%) is the most common cause of extrahepatic biliary duct stenosis, considering the location, or infiltration originating from the pancreatic head (16). In such cases prognosis is extremely unfavorable. Since non-neoplastic stenoses are relatively rare, and obviously of iatrogenic cause, it is now believed that symptomatic strictures of the biliary ducts should be treated as adenocarcinoma, until proven otherwise. Most carcinoid lesions are characterized by slow growth with relatively rare metastases. The treatment of choice consists in the radical resection, although due to their biology, small lesions (<2 cm) might be subjected to local resections. Cases of complete recovery were described after carcinoid enucleation. Additionally, in case of patients with large lesions and metastases, palliative surgical procedures prolong survival. The presence of neurofibromatosis type I, which is observed in 25% of patients with carcinoids might facilitate proper diagnosis (17).

It was also found that in comparison to biliary duct carcinoma, carcinoids are diagnosed in younger patients (50-60 years), with a longer period between the onset of symptoms and diagnosis. Biochemical diagnostics include chromogranin A (CgA), serotonin, and 5-HIAA. One may also perform octreotide scintigraphy, in order to diagnose possible metastatic lesions (10). Unfortunately, their value is limited to patients with hormonally active tumors, and those with somatostatin receptors. The „spoke wheel” appearance observed on computed tomography might prove helpful, which often accompanies desmoplastic reactions (18). In case of our patient, on the basis of preoperative examinations we were unable to establish the final diagnosis, and thus, surgical exploration was initiated associated with radical resection.

When planning treatment of carcinoids, therapy should be based on the recommendations of the “Task Force on Neuroendocrine tumors” (19). Recommendations elaborated by European and American Societies are also available (20, 21, 22). They mostly concern typical location tumors, although some might be applied in case of our patient. During the postoperative period adjuvant therapy should be considered. Cold and hot (conjugated with radioisotopes) somatostatin analogs are becoming more commonly used, as far as somatostatin receptors have been diagnosed. There was no benefit from the use of radiotherapy, however, in case of adjuvant therapy, chemotherapy proved useful. The efficacy of interferon alpha was also demonstrated. It is worth noting that in case of carcinoids patients benefit from cytoreductive surgery, even if total tumor resection is not possible. A five-year observation period is recommended after surgery.

Existing literature data is insufficient to precisely determine prognosis in case of our patient. It seems however, that prognosis in case of patients with diagnosed extrahepatic biliary duct carcinoids is good (10, 13). In case
of non-invasive neuroendocrine tumors subjected to radical resections, recovery is assessed at 80-100% (23). At the same time it should be remembered that this is a high-risk group, where 20% of patients will be diagnosed with yet another neoplastic lesion (based on the analysis of 13 000 cases from the NET neoplastic database).

In conclusion, extrahepatic biliary duct carcinoids are rarely observed. In case of a relatively small tumor size, they may cause symptoms of jaundice, which allows their earlier diagnosis and better prognosis. Radical surgery is the method of choice or palliative management with adjuvant therapy in selected cases.

REFERENCES


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