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Carcinoid tumor of common bile duct: A rare presentation

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Abstract
Carcinoid tumors of common bile duct are extremely rare tumors. Clinically these tumors have indolent course and are typically managed by radical surgery. Here, we reported clinic- pathological features of carcinoid tumor in common bile duct in a 38-year young female, who presented with right upper abdominal pain of 1-month duration associated with multiple episodes of non-bilious vomiting and signs of icterus. Endoscopic retrograde cholangiopancreatography (ERCP) revealed lower common bile duct stricture, biopsy of which showed well differentiated carcinoid tumor. Patient was sent for radical surgery.

Keywords: carcinoid tumor, common bile DUCT, ERCP, well differentiated tumor, radical surgery

Introduction

Carcinoid tumors are a type of neuroendocrine tumor that are extremely rare in extrahepatic biliary duct. These tumors have a silent course with rare aggressiveness. Clinically these tumors are indistinguishable from other common bile duct tumor. Here, we reported clinic- pathological features of carcinoid tumor in common bile duct in a middle-aged female.

Material and Methods

Case Presentation

A 38-year young female, without any associated co-morbidities or illness, presented with history of pain abdomen for one month which was insidious in onset. Pain was moderate in intensity, not relieved by oral analgesics, occurring at right upper abdomen and radiating to back & right scapular region. It was associated with frequent, non-bilious, non-projectile vomiting along with yellowish discolouration of skin and sclera for the same duration. She had no bowel-bladder disturbances or history of upper or lower gastrointestinal bleeding. She was a multipara with two live children and was having regular, normal menstruation. Patient was non-smoker, non-alcoholic. She had a history of Koch's 15-years back, took anti-tubercular therapy for 6-months & was cured completely.

General physical examination showed dark yellow discolouration of sclera and skin (icterus); systemic examination was normal. Complete hemogram & renal function test of the patient were within normal limits, but liver enzymes (SGOT and SGPT) and bilirubin (both direct and indirect) were elevated. On ultrasonography abdomen, markedly enlarged gall bladder (13.1 × 4.4 × 5.1 cm) with impacted calculus in neck region, grossly dilated intra-hepatic biliary radicles (IHBR) & common bile duct and large amount of sludge at lower end of common bile duct were visualized. She underwent endoscopic retrograde cholangiopancreatography (ERCP), which revealed stricture in lower common bile duct, biopsy of which revealed well differentiated endocrine neoplasm suggestive of carcinoid. Patient was sent for radical surgery.

Discussion

First described in 1988, carcinoid tumors have distinct histological, chemical and biological characteristics along with varying clinical features depend on anatomical site. With approximately one-third cases metastasize to distal sites, disease free survival is 2 to 11 years [1]. Carcinoid tumors of common bile duct was first described in 1959 [2]. Which are account for 0.1-0.2% of all carcinoids of the gastrointestinal tract [3]. This extreme rare tumors arise from Kulchitsky cell, which are embryonal neural crest cells [3]. The most common anatomical location of extra hepatic carcinoid tumor is common bile duct [4]. Majority of the retrospective analysis concluded that it has a female preponderance, most commonly seen around 5th decades of life and have indolent clinical course [5]. Characteristics symptoms are obstructive jaundice and right upper abdomen pain due to mass effect mimicking cholangiocarcinoma, the commonest cancer of common bile duct. Approximately one-third reported cases had distant metastasis; liver, lymph nodes and pancreas being the metastatic sites [6]. Carcinoid and other neuroendocrine tumors of biliary tract and pancreas may be associated with Von-Hippel-Lindau (VHL) disease, although specific pathology remains unclear [7]. Surgical resection followed by histological review and immunohistochemical staining is the only options for definitive diagnosis, most CBD carcinoids being immune-reactive to chromogranin [3]. Aggressive surgical resection with negative margin remains the curative treatment and most important prognostic marker [6]. Adjuvant chemotherapy (5- fluorouracil and strepzocin) and radiotherapy were tried in different clinical trials without survival benefit [1]. Even removal of resectable single metastatic lesion like solitary liver metastasis also have

beneficial outcome [8]. Overall, the 10-year survival rate of carcinoid tumors of the extrahepatic bile ducts was 80% [4].

Conclusion

In conclusion, the extreme rarity of carcinoid tumor of the common bile duct causing similar symptoms like other common tumor of that site makes its pre-operative diagnosis very unlikely. However, whenever such diagnosis is made, primary tumors should be removed always and if deemed unresectable debulking should be attempted. More clinical experiments are needed to guide adjuvant therapy in incomplete resected cases with a demonstrable survival advantage.

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