CASE REPORT

Carcinoma of the Ampulla of Vater Associated with Annular Pancreas

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Abstract Carcinoma of the ampulla of Vater associated with annular pancreas is a rare entity. Only seven cases have been reported so far in the English literature. We herein report one such case in a 42-year-lady who presented with progressively increasing jaundice and pain in the upper abdomen for past 2 months. A magnetic resonance cholangiopancreatography (MRCP) and MRI revealed an annular pancreas with an ampullary mass and a dilated CBD. An upper gastrointestinal endoscopic biopsy from the ampullary mass revealed a moderately differentiated adenocarcinoma. She was diagnosed as having annular pancreas with periampullary cancer that was subsequently confirmed at laparotomy. Histological examination confirmed a complete annular pancreas with a 2 cm × 2 cm moderately differentiated adenocarcinoma of the ampulla of Vater. The management, however, remains as in any case of periampullary malignancy. This highlights the importance that obstructive jaundice in an adult patient presenting with annular pancreas may be associated with a coexisting periampullary malignancy.

Keywords Ampulla of Vater · Anatomy · Periampullary cancer · Annular pancreas · Obstructive jaundice

Introduction

Annular pancreas (AP) is an uncommon congenital anomaly. A prevalence of 5–15 cases per 100,000 adults has been reported in an autopsy series [1]. AP is due to malrotation of the pancreatic ventral bud during embryological development. Popular theories explaining the pathogenesis include adhesion of the right ventral bud to the anterior duodenal wall and failure of the left ventral bud to involute [2]. AP is usually asymptomatic but is also known to present at times with duodenal obstruction, pancreatitis, peptic ulceration, and rarely obstructive jaundice with coexistent periampullary cancer [3].

We herein report a case of an annular pancreas complicated by carcinoma of the papilla of Vater.

Case Report

A 42-year-old lady presented with progressively increasing jaundice and pain in the upper abdomen for past 2 months. Her serum bilirubin was 3.1 mg/dl (post stenting), and the rest of her hemogram, chest X-ray, pulmonary function tests, and 2D Echo were normal. A magnetic resonance cholangiopancreatography (MRCP) and MRI revealed an annular pancreas with ampullary mass and a dilated CBD (Fig. 1) that is completely resectable. A complete ring of pancreatic tissue was seen to be encircling the second part of the duodenum upon laparotomy. There was no evidence of abdominal lymphadenopathy, liver deposits, or ascites. An endoscopic biopsy from the ampullary mass revealed a moderately differentiated adenocarcinoma. She underwent a successful standard Whipple's pancreaticoduodenectomy. Histopathological examination revealed a 2 cm × 2 cm moderately differentiated adenocarcinoma of the ampulla of Vater. Seven of 15 lymph nodes dissected had metastases. The

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Fig. 1 Magnetic resonance cholangiopancreatography (MRCP) image showing a complete ring of the pancreatic duct (*black arrow*) of the annular pancreas and ampullary mass (*white arrowhead*)

patient was given 6 cycles of gemcitabine with cisplatin-based adjuvant chemotherapy. She died of metastatic cancer 4 years after surgery.

Discussion

AP was first described by Tiedemann in 1818 and was termed “annular pancreas” in 1862 by Ecker. The first successful surgery for an annular pancreas, a gastrojejunostomy, was performed in 1905 by Vidal [3].

A complete ring of pancreatic tissue around the duodenum is not required for the radiologic diagnosis of AP; incomplete annulus has been reported in more than one third of cases by Sandrasegaran et al. [1]. The best noninvasive technique of assessing ductal anatomy is secretin-enhanced MRCP. Posterolateral extension of pancreatic tissue to the duodenum or the finding of a crocodile jaw appearance of the pancreatic head raises the possibility of AP [1].

Jaundice is an uncommon presentation and may not be a direct result of the AP. It may result from pancreatitis, choledocholithiasis, and alcoholic liver disease besides a malignancy of the ampulla of Vater, duodenum, pancreas, or even bile duct [4]. It has been hypothesized that chronic obstruction due to AP leads to continuous exfoliation of cells in the region of the ampulla and rapid mucosal turnover probably leads to dysplastic changes and metaplasia in that region [5].

A review of the 106 cases by Yogi et al. [6] found nine (8.5 %) having a pancreaticobiliary malignancy (five ampullary carcinomas, two pancreatic carcinomas, and two cholangiocarcinomas). Previously, only seven cases of ampullary carcinoma with AP have been reported in the English literature [3].

Conclusion

The possibility of a coexistent underlying malignancy of the pancreaticobiliary system should always be considered in a patient with AP presenting with obstructive jaundice. The approach, however, remains the same as in any other patient with periampullary cancer.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no competing interests.

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