

症 例

Carcinoma of the Peripapillary Portion of the Duodenum without Jaundice

by

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The presence of obstructive jaundice is usually helpful in the diagnosis of a tumor at the peripapillary portion of the duodenum, but I experienced a case of the peripapillary carcinoma without jaundice. The purpose of this paper is to report this case and to discuss about the classification of the duodenal carcinoma.

Case Report

A 63-year-old man was admitted to the Saiseikai Suita Hospital with a 2-month history of nausea, loss of appetite and heartburn. At first the patient had fever ranging from 38° to 39° C, but the temperature had dropped to normal a month before admission. He had no history of jaundice, vomiting, abdominal pain or tarry stools. On examination there was no evidence of jaundice. Abdomen was flat and no tumor was felt. The gallbladder was not palpable. An X-ray film of the chest was normal. Roentgenologic examination of the stomach and duodenum revealed an obstructing lesion at the junction of the second and third part of the duodenum. Hypotonic duodenography disclosed an irregular filling defect (Fig. 1), which had the appearance of a peripapillary carcinoma of the duodenum.

Laboratory studies revealed ; R. B. C. 3,410,000, hemoglobin 7.7g/dl, hematocrit 28%, W. B. C. 12,100, serum amylase 16

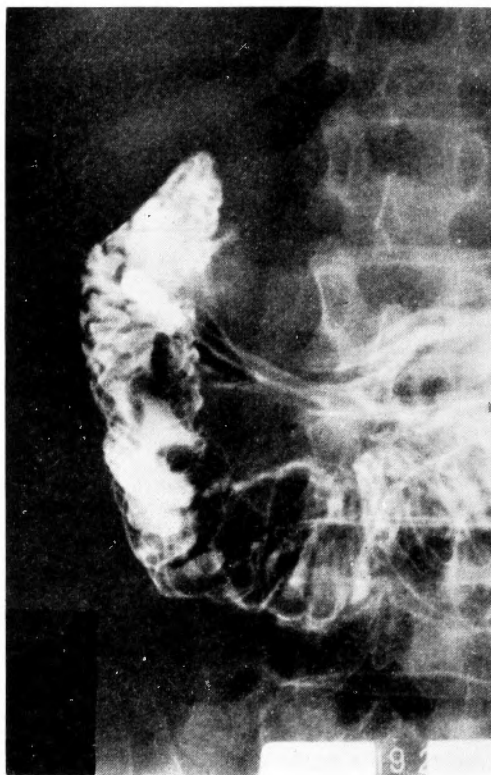


Fig. 1. Hypotonic duodenography shows an irregular filling defect at the junction of the second and third part of the duodenum.

Key words : Classification, Duodenal carcinoma, Jaundice, Peripapillary carcinoma

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units, urine amylase 16 units, Meulengracht 8, total serum bilirubin 1.2mg per cent, thymol turbidity tests 0.7, GOT 49, GPT 43, alkaline phosphatase 60 Bodansky units, LDH 170, BUN 15mg per cent, blood glucose 76 mg per 100ml, urinalysis negative.

At operation an egg-sized mass was found in the second part of the duodenum. The common bile duct was dilated to a maximum diameter of 1.5cm. Neither enlargement of the gallbladder nor dilatation of the pancreatic duct was seen. The liver appeared healthy. Radical pancreatoduodenectomy was carried out. Anastomosis was performed on CHILD'S method. The excised specimen showed a fungating tumor around the papilla (Fig. 2). An inserted probe shows the common bile duct leading to the papilla. Papilla was located approximately in the center of the tumor. The pancreatic duct entered the duodenum separately and was not involved. Microscopically the tumor was a moderately well-differentiated adenocarcinoma (Fig. 3). In the original specimen transition between the normal duodenal epithelium and the tumor cells was noted, and the tumor was considered to have originated from the duodenal epithelium. Direct invasion of the carcinoma into the head of the pancreas was observed but the common bile duct was not invaded (Fig. 4). There was no evidence of spread to the regional lymph nodes. The postoperative course was uneventful. The patient was discharged a month after operation. He appeared well two and a half years after operation.

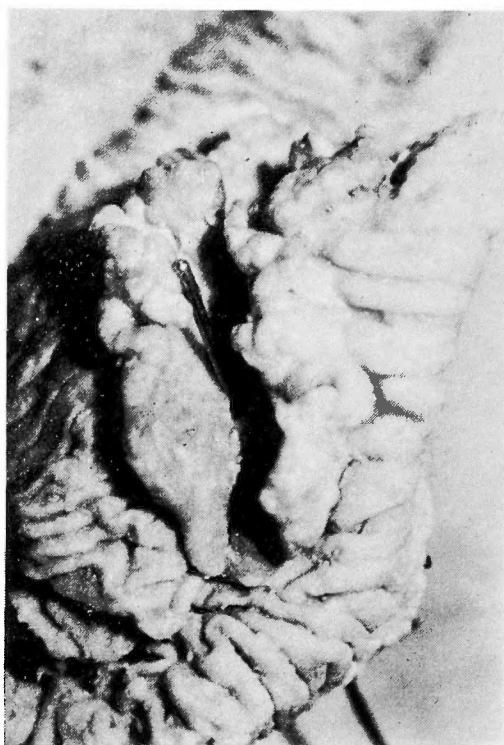


Fig. 2. The excised specimen shows a fungating tumor around the papilla. A probe is inserted into the common bile duct.

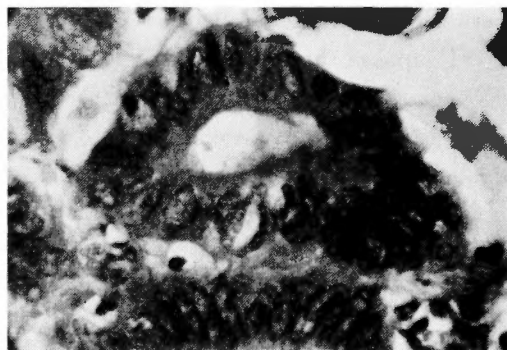


Fig. 3. The tumor is a moderately well-differentiated adenocarcinoma (X400).

Discussion

When a tumor of the duodenum is located near the papilla of VATER, the patient may develop progressive jaundice. If the tumor is of peripapillary site, 99% of the patients are said to have jaundice^{1),2)}. Some authors³⁻⁷⁾ reported that jaundice was intermittent in the



Fig. 4. The common bile duct is not invaded by the tumor cells (X200).

case of peripapillary carcinoma. According to MONGE⁶⁾, jaundice fluctuated in half the patients of 65 with carcinoma of the papilla, but in only 2 of the 28 with carcinoma of the pancreatic head.

In the case of the patient presented here, there was no jaundice and the icterus index was normal, although the level of the alkaline phosphatase was elevated and the common bile duct was dilated. HOFFMAN⁸⁾ stated that the jaundice may entirely disappear or be intermittent for the following reasons: (1) a soft, friable tumor in this location may ulcerate and slough away, accompanied by a temporary or lasting relief of the jaundice; (2) increased intrabiliary pressure may force the bile through the obstructed papilla (BRILL); (3) the subsidence of papillary edema may release the obstruction and afford relief (MEYER and ROSENBERG). On the other hand, MONGE⁶⁾ stated that variation in the jaundice was not

related to ulceration and sloughing of the surface of carcinoma of the papilla, but may perhaps have been due to the soft, polypoid, and somewhat mobile nature of these tumors.

I suppose that the patient was not jaundiced because there was no infiltration to the common bile duct. It is said that carcinoma of the stomach will stop short at the pylorus and carcinoma of the caecum will stop at the ileocaecal valve. Similarly it is supposed that duodenal carcinoma stopped at the papilla of VATER, but the common bile duct was dilated presumably due to a transient papillary edema. That the tumor had originated from the duodenal mucosa is evident pathologically for the following reasons: (1) macroscopically the tumor protruded from duodenal mucosa into the duodenal lumen, (2) microscopically transition from the normal duodenal epithelium to the tumor cells was noted, (3) the common bile duct was not infiltrated by the tumor cells. But clinically this case should not be included in duodenal carcinoma for the present as indicated in my previous paper⁹⁾.

Most of the peripapillary lesions arise in the biliary or pancreatic ducts rather than from the duodenal mucosa. But in the literature many peripapillary carcinomas have been included in the duodenal neoplasms because of the difficulty in deciding the true site of the origin of these carcinomas. This accounts for the high percentage of peripapillary lesions in the older literature. I believe that the peripapillary lesions should be in a separate category. The terms "peripapillary" and "periampullary" are occasionally used indis-

criminally, as though they were synonymous words indicating the same anatomic structure. I think the term "peripapillary" should indicate those tumors which invade macroscopically the papilla of VATER while "periampullary" should indicate those tumors which spread submucosally without macroscopical changes in the duodenal mucosa. As to "periampullary", there is a far less possibility of a tumor originating from the duodenal mucosa.

The difference between periampullary carcinoma and pancreatic carcinoma is sometimes difficult. But WARREN⁷⁾ and SUZUKI¹⁰⁾ emphasize the importance of division of these two cancers because of the difference of prognosis. Carcinoma of the pancreas has a much worse prognosis than that of the ampulla.

However, the problem still remains as to how we should classify a far-advanced carcinoma which may have come to include the papilla of VATER or the pancreas. I hope an appropriate classification be made by some authoritative organization.

Relatively little is known about the anatomy of the ampulla of VATER. MIYAZAKI¹¹⁾ expressed a doubt upon the existence of the ampulla on the basis of his experiences in the intraoperative cholangiography (362 cases) and necropsy (11 cases). I wish to agree with him from my experiences of pancreaticoduodenectomy.

From the above-mentioned evidence, the classification of duodenal carcinoma as "supra-ampullary", "periampullary" and "infraampullary" is not recommended. I propose to classify them simply "suprapapillary" and "infrapapillary" as RESNIK¹²⁾, since embryologically the former derived from the foregut while the latter from the yolk sack (midgut).

Summary

A case of peripapillary cancer without jaundice is presented. I supposed that the patient had no jaundice because there had been no infiltration to the common bile duct, and the common bile duct was dilated due to a transient papillary edema.

At present the classification of the duodenal carcinoma is ambiguous. I am of opinion that peripapillary lesions should be in a separate category, although there remains some question as to how we should classify a large extensive carcinoma which may have come to include the papilla of VATER or the pancreas. It is expected that an authoritative organization decides the classification. Untill then I wish to emphasize that peripapillary carcinoma should be excluded from the duodenal cancer.

According to MIYAZAKI¹¹⁾, the existence of the ampulla of VATER is doubtful. Therefore it is not recommended to use the word "ampulla of VATER". I propose to classify the duodenal cancer simply as "suprapapillary" and "infrapapillary".

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和文抄録

乳頭部癌の無黄疸症例

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十二指腸乳頭部周囲に発育した癌でありながら、黄疸を来たさなかった1例を報告した。文献には、乳頭部癌を十二指腸癌に含めているのが多く見られるが、乳頭部癌で十二指腸粘膜より発生するのは少なく、乳

頭部は除くべきだと思われる。そして十二指腸癌は、乳頭上部癌と乳頭下部癌とに分類し、乳頭部癌については、膨大部癌、膵頭部癌等と共に別の分類法を考えるべきだと思われる。