Virchows Arch. Abt. A Path. Anat. 355, 209–212 (1972)

Case Report

On Primary Duodenal Carcinoma Developing from Polyposis

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Received October 27, 1971

Summary. This report deals with a duodenal adenocarcinoma that metastasized to the liver, causing the death of the 41-year-old patient. The carcinoma arose from an intestinal polyposis, the symptoms of which could have developed over a period of 19 years before the patient's death.

Introduction

Primary carcinoma of the duodenum was first described by Hamburger through an autopsy in 1745. It is one of the uncommon carcinomas of the alimentary tract. Its incidence in comparison to other intestinal carcinomas is quoted to be 0.3–4.0% by various investigators. The site most commonly affected in the small intestine is the duodenum (Marzoli and Meyer-Burgdorff, 1962).

Because primary duodenal carcinomas are so rare, it seems of value to report personal observations.

Case Report. Nineteen years ago a 41-year-old patient first complained of cramplike pains in the upper abdomen. The family history as well as his own past history revealed no pathological findings. A gastritis with hypoacidity and anaemia was diagnosed. His complaints recurred. Roentgenograms of the gastrointestinal tract, taken 9 years previously, showed a severe duodenitis of the whole duodenum and a bleeding duodenal ulcer. In September 1963 a Billroth's operation (B II) with retrocolic gastroenterostomy was performed, during which a $1 \times 1 \times 2$ cm jejunal polyp was removed. Microscopically a relapsing erosive gastritis and a benign adenomatous polyp were diagnosed. One year later another intestinal polyp was removed during a repeat gastric resection. For the first time an intestinal polyposis was diagnosed but surgical treatment of it proved impossible. Over the next years the pathological changes led to recurrent intestinal haemorrhage; tarry stools and chronic anaemia were often detected. In April 1970 a radiographic follow-up showed no evidence of pedunculated intestinal polyps. The barium test was considered as normal. Four months later the patient was severely cachectic and died with signs of circulatory failure.

Results

At autopsy numerous irregularly-shaped polyps, almost the size of cherries, were found projecting from the duodenal mucosa distal to the papilla of Vater. An ulcerated carcinoma $4.0 \times 5.5 \times 1.5$ cm was detected between the pedicles of two polyps. A well-defined border of the carcinoma could be identified (Fig. 1). The craterlike mucosal defect was situated near the orifice of the papilla. The mucosa of the papilla, however, was normal, soft and pliable and not affected by the cancer. *Microscopically*, a partly adenomatous, partly scirrhous carcinoma was seen at the base of a polyposis. The carcinoma spread partly over intact mucosa, partly into the submucosa (Figs. 2, 3). Numerous *metastases*, almost the

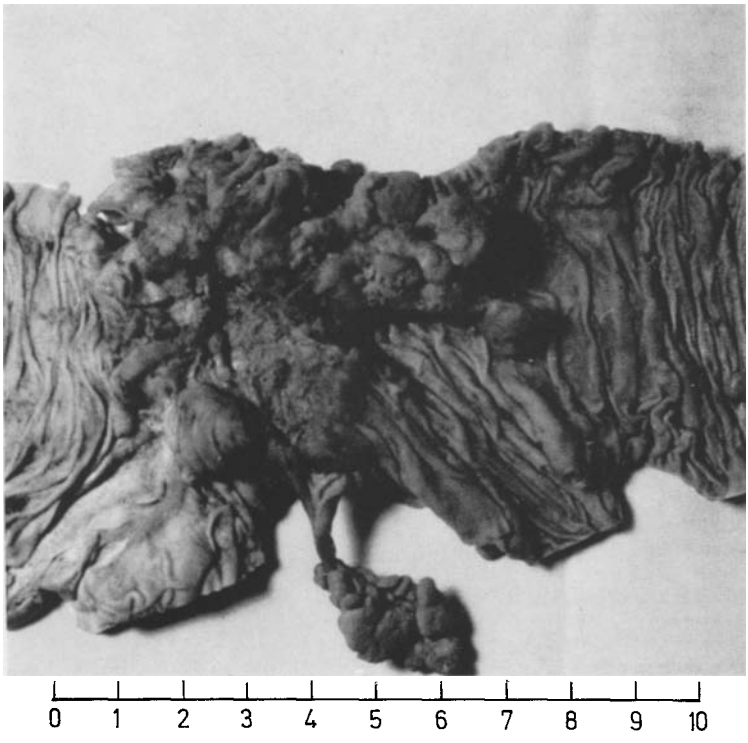

Fig. 1. Duodenal carcinoma developing from polyposis. ♂, 41 years

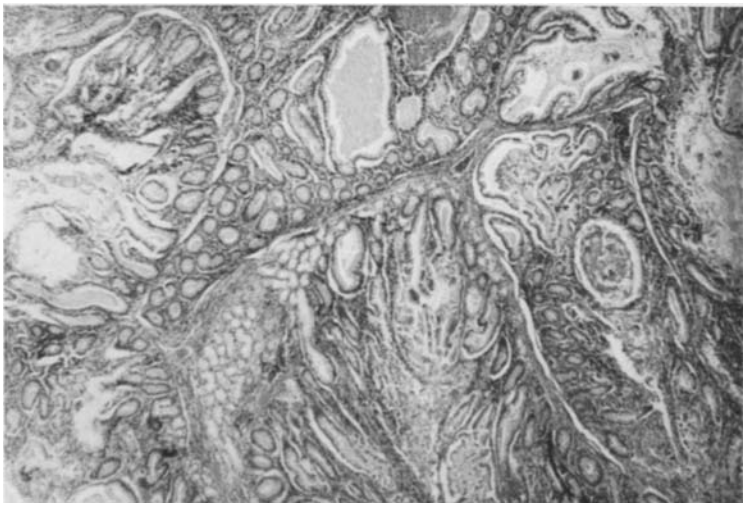

Fig. 2. Edge of an adeno-carcinoma developing from polyposis of the duodenum. (HE); enlargement ×25

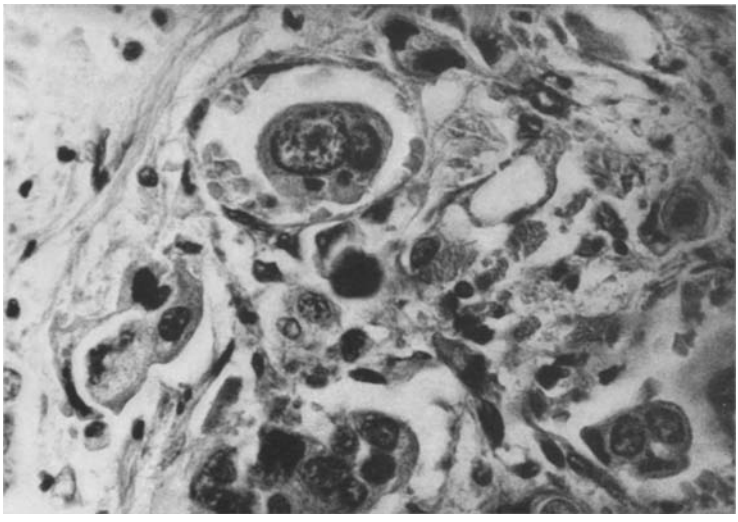

Fig. 3. Pleomorphic nuclei of glandular structures of a duodenal adenocarcinoma. (HE); enlargement ×400

size of plums, were discovered in the liver. The paraaortal lymph nodes revealed metastases. The hepatic jaundice was severe, with only a moderate dilatation of the choledochus.

Discussion

Of 438 cases of duodenal carcinoma, 59.2% affected the papilla, 18.3% the infrapapillary portion, and 22.5% the suprapapillary segment. Most commonly only one duodenal carcinoma develops. Clinical manifestations usually appear between forty and seventy years of age (Hentschel, 1968). According to Kramer's statistics and observations (1968), men were affected more frequently than women. The average age was 56.8 years. Therefore, our 41-year-old patient is comparatively young.

The clinical symptoms depending on where the carcinoma develops:

1. The infrapapillary duodenal carcinoma leads to frequent vomiting, the vomitus containing a mixture of food, bile and pancreatic juice. This is the main symptom of stenosis.

2. The suprapapillary carcinoma, if detected early, shows signs of stenosis like those with pyloric stenosis: hypermotility and gastric ectasia.

3. The peripapillary carcinoma at an early stage causes biliary obstruction with bile retention jaundice and biliary vomiting without colic.

There may be general symptoms such as recurring pains, especially in the epigastric region and a permanent sensation of tension and fullness. Acute or chronic haemorrhage and a trace of blood in the stools, as was often seen in our patient over a period of ten years, seem to be less frequent. Characteristic cramp-like recurring pains, which is a diagnostic complaint of duodenal carcinoma, were experienced by our patient over a period of 19 years. In a clinical workup ten

years previously these symptoms were diagnosed as caused by a bleeding duodenal ulcer with concomitant anaemia and severe duodenitis. In the course of a gastric resection three years later for a duodenal ulcer, a duodenal polyposis was discovered. This mucosal neoplasm produced over many years repeated intestinal haemorrhage and a chronic anaemia. Four months before the patient's death an induration was clearly palpated in the right upper abdominal quadrant. Cachexia progressed. Jaundice developed as a result of the peripapillary tumor and multiple metastases in the liver; the total serum bilirubin increased to 22 mg-%. We believe the clinical signs and symptoms as eve observed them, were characteristic and are in good accord with previous observations.

Microscopically, the most common type of carcinoma of the duodenum is the adenocarcinoma. It usually develops from malignant transformation of the crypts of Lieberkühn, but may also arise from duodenal glands. Occasionally scirrhous, solid, papillary, medullary or gelatinous types of the carcinoma are observed (Lehmann, 1956; Maurer, 1968). The symptoms rarely lead to a definite clinical diagnosis. In a history like that given above, detailed roentgenograms of the small intestine is the most important diagnosticprocedure, as numerous investigators emphasize (referring to Hasselbach and Witte, 1969). Further differential diagnostic procedures may be employed, e.g., the selective catheter arteriography of the superior mesenteric artery or the search for tumor cells in the duodenal juice. As in our case seven years before death, if an intestinal polyposis with a tendency to relapsing haemorrhage is diagnostically established, a malignant transformation should be held as possible even in younger patients an operation should be performed.

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