Primary Squamous Cell Carcinoma of the Pancreas: A Report of Two Cases and Review of the Literature

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Abstract

Primary squamous cell carcinoma (SCC) of the pancreas is extremely rare. We herein report two such cases. Aside from various morphological characteristics, the clinical presentation of pancreatic SCC is the same as that of adenocarcinoma. The treatment is controversial, and the prognosis is poor.

Key words: squamous cell carcinoma, pancreas, etiopathogenesis, chemotherapy


Introduction

Pancreatic adenocarcinoma accounts for most cases of non-endocrine pancreatic cancer. Nevertheless, squamous cell carcinoma (SCC) of the pancreas is rare, representing only 0.5% of cases among all pancreatic neoplasms (1).

We herein report two cases of primary pure SCC of the pancreas diagnosed and treated at the Department of Oncology at Habib Bourguiba Hospital south of Tunisia.

Case Reports

Case 1

A 48-year-old man was referred with a 3-month history of epigastric pain exhibiting radiation to the back. A physical examination revealed mild tenderness over the epigastrium with moderately abundant ascites. The serum lipase level was normal (8 U/L, reference: 4-60 U/L); however, the carbohydrate antigen (CA) 19-9 level was elevated (980 U/mL, reference: 2-40 U/mL). Abdominal ultrasonography showed a 4-cm hypoechoic tumor in the pancreatic head with intraperitoneal effusion. The patient underwent computed tomography (CT) of the abdomen for a further evaluation, which showed a 4.6 cm low-density mass centered at the head of the pancreas involving more than 50% of the superior mesenteric artery with near occlusion of the superior mesenteric veins. A CT-guided biopsy of the pancreatic mass was performed and histological examination revealed malignant tumor proliferation with clusters of atypical squamous cells leading to a diagnosis of SCC of the pancreas (Fig. 1). CT scans of the head, neck and chest, an otorhinolaryngological assessment and an endoscopic examination of the gastrointestinal tract performed to identify other primary tumors were normal. Therefore, a primitive pancreatic origin was assumed. The patient was treated with chemotherapy using 5-fluorouracil (800 mg/m² over 24 hours, days 1-5) and cisplatin (80 mg/m², day 1). His clinical course was marked by an increase in the tumor size on ultrasonography after the first cycle of treatment. The chemotherapy regimen was stopped, and palliative radiotherapy at a dose of 30 Gy was planned; however, the patient died after approximately nine months of follow-up.

Case 2

A 42-year-old man was admitted to our hospital for treatment of vomiting and peri umbilical pain. A physical examination revealed epigastric tenderness. Routine laboratory data, including the lipase and amylase serum levels, were unremarkable, and tumor markers were within the normal ranges. Abdominal ultrasonography showed a 4-cm hypoechoic corporeal-caudal mass in the

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Abdominal CT confirmed the presence of a corporeo-caudal hypodense lesion in the pancreas measuring 4×3.7×3.1 cm (Fig. 2). The mass had invaded the splenic vessels, with no evidence of metastatic disease. The patient underwent spleno-corporeo-caudal pancreatectomy with pancreatic stump sutures, and a histopathological study showed moderately differentiated keratinizing SCC of the pancreatic body, measuring 6 cm, with no evidence of glandular formation in any of tissue sections of the tumor (Fig. 3). The lesion extended towards the peripancreatic fat, with negative finding for lymph node metastasis. The omentum and spleen were normal. According to the tumor-node-metastasis (TNM) classification, the tumor was diagnosed as T3N0M0. Pre-and postoperative investigations for other possible primary foci of SCC including CT scans of the head, neck and chest, an otorhinolaryngological assessment and endoscopic examination of the gastrointestinal tract were negative. The patient subsequently received chemotherapy according to the FUFOL regiment (5-fluorouracil, folinic acid). He is currently alive in complete remission after a follow-up period of 26 months.

**Discussion**

SCC of the pancreas is exceedingly rare, the first case of which was described by Lowry in 1949 (1). In a review of 6,688 cases of exocrine pancreatic cancer identified from various cancer registries between 1950 and 1985, the reported incidence of SCC varied from 0.5 to 5% (2). In contrast, the incidence was 0.5% in a series of 5,075 patients reported by Baylor and Berg (3). At Habib Bourguiba Hospital in Sfax, a total of 158 cases of exocrine pancreatic cancer were treated between 2000 and 2012, among which only two cases of SCC were identified.

In view of its rarity, SCC of the pancreas is presumed to be the result of metastasis from another primary site until proven otherwise (4). In our two cases, we concluded that the diagnosis was primary SCC of the pancreas after eliminating other possibilities. Indeed, the results of an otorhinolaryngological assessment, endoscopic examination of the gastrointestinal tract and CT of the head and chest were normal.

Several theories have been proposed to explain the development of SCC of the pancreas. First, the disease may result from the malignant transformation of squamous metaplasia secondary to chronic inflammation, such as that involving chronic pancreatitis (2). Second, the lesion may originate from mixed adenosquamous carcinoma in which the glandular components have disappeared (5). Third, the tumor may drift from a biopotential primitive capable of differentiating into either glandular adenocarcinoma or SCC (6) originate from pre-existing adenocarcinoma with squamous metaplasia (5). In the present two cases, a histopathological study showed pure SCC without an adenomatous component or metaplasia. Moreover, our patients had experienced no previous episodes of pancreatitis.

The clinical presentation of SCC is similar to that of adenocarcinoma in our experience, in accordance with data reported in the literature (7).

In our first case, the tumor was judged to be inoperable due to invasion of the superior mesenteric artery. In contrast,
the second patient underwent curative resection, which is the best therapeutic option for appropriate indications. Among several chemotherapeutic options, including the combination of cisplatin and 5-fluorouracil or vinblastine (8), we used the combination of 5-fluorouracil and cisplatin in our first patient; however, his clinical course was marked by locoregional progression of the tumor. We applied the FUFOL regimen in our second case, with a very good outcome. One report described a better response to chemo radiotherapy regimens based on gemcitabine (9), and Ravry documented an objective response with a striking symptomatic improvement following treatment with bleomycin in one of two patients with the squamous cell variety of pancreatic cancer (10). However, no standard chemotherapy regimen has been established.

SCC of the pancreas has a poor prognosis. In fact, Brown et al. demonstrated a median survival of seven months (range: 6-16 months) for patients who undergo curative resection. Our second patient, who was treated with curative surgery and the FUFOL regimen, is currently alive in complete remission after 26 months of follow-up. For patients who do not undergo curative resection, the median survival is three months (range: 0.25-9 months) (9), which is in agreement with the findings in our first case, in which the patient died after a follow-up period of nine months.

SCC of the pancreas is rare, and aside from various morphological characteristics; the clinical presentation is the same as that for adenocarcinoma. Curative surgical treatment should be performed in cases of tumor spread, if the patient’s general condition permits. Otherwise, chemotherapy may help to improve the patient’s status or achieve an objective response in exceptional circumstances.

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References