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Case Report
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Primary Squamous Cell Carcinoma of the Pancreas

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Abstract

Context: Squamous cell carcinoma of the pancreas is extremely rare, and has a very poor prognosis. Only radical surgery, when possible, can provide prolonged survival.

Case: This report concerns a patient treated for a squamous adenocarcinoma of the distal pancreas with locoregional extension, revealed by an atypical symptomatology; the imaging revealed a mass of the distal pancreas with signs of locoregional extension. We performed a en bloc resection. The patient received adjuvant therapy with cisplatin and gemcitabine. Multiple metastases of the liver in a febrile context were discovered during the seventh postoperative month.

Conclusion: Surgical resection remains the more affective approach to deal with Squamous cell carcinoma.

Keywords: Squamous cell carcinoma; Radical surgery; Adjuvant therapy; Liver meatstasis

Introduction

Squamous Cell Carcinoma (SCC) is considered as an aggressive subtype of exocrine tumors of the pancreas. According to observations collected in the literature, mainly in the form of clinical case reports, the incidence of pure squamous carcinoma of the pancreas is 0.5% to 5% [1,2]. In the majority of the published cases, pain still dominates the clinical charts, followed by weight loss in more than half of the patients, and jaundice in ¼ of the patients. Lymph node invasion and liver metastases are frequently found at the time of diagnosis. The survival of patients in whom the tumor was resected is better than that in which the tumor was not resected.

Case Presentation

A 58 years old man with a history of chronic gastritis and gout under treatment was referred to us for management of a tumor of the distal pancreas with locoregional extension. The patient suffered from epigastric pain, subocclusive syndrome, asthenia, anorexia and a weight loss of 12 kg in 03 months.

The initial physical examination revealed a hard, immobile, painless, epigastric palpable mass. Gastrointestinal endoscopy showed chronic congestive gastritis with intestinal metaplasia. Imaging (ultrasound, CT scan and MRI) showed a distal pancreatic tumor with splenic vascular involvement. This tumor invaded the spleen and the colon. Additionally, there was a presence of some celiac lymph nodes. No hepatic, thoracic or osseous localization (Figure 1).

Pancreatic enzymes, as well as tumor markers, were in the normal range (ACE: 3.73 ng/ml, CA 19-9: 224.4 U/l), the rest of the blood test was normal. After careful consideration, and given the frequency of occlusive signs, we opted for a surgical option with the aim to perform an en bloc oncologic resection (R0). The surgical exploration has identified a large tumor in the distal pancreas, infiltrating the left mesocolon, the spleen and adhering to the large gastric curvature, as well as the presence of multiple mesenteric and celiac lymph nodes. There were no carcinosis, or liver metastasis. A monoblock resection, "caudal spleno-pancreatectomy" enlarged to the large gastric curvature, the left 1/3 of the transverse colon and the left colic angle and the adrenal were performed in addition to the cleaning of the celiac trunk and the vascular mesenteric (Figure 2A). The immediate post-operative care was kept simple; the patient was discharged on Day 10.

The pathology of the specimen showed a mass of the pancreas of 10 cm, with infiltration of the spleen and the left adrenal gland. However, the mass was in contact with the stomach and the left

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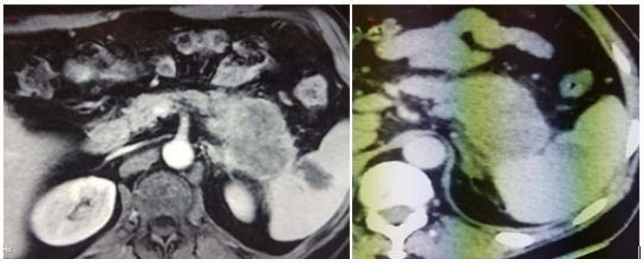

Figure 1: Pancreatic tumor with splenic vascular involvement. Tumor invaded the spleen and the transverse colon. Additionally celiac lymph nodes.

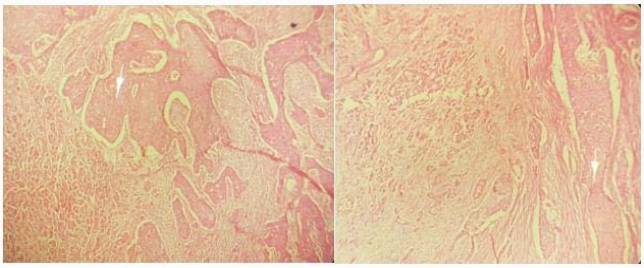

Figure 2A: Squamous Cell Carcinoma ET: x10.

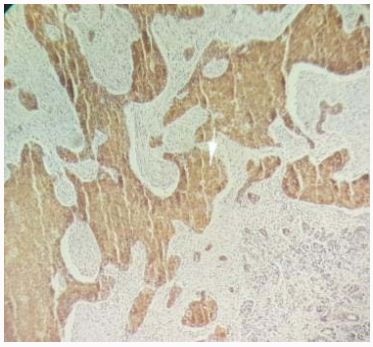

Figure 2B: CK19 positive.

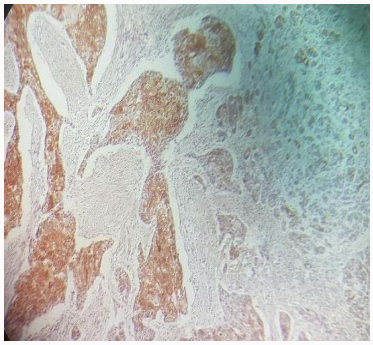

Figure 2C: Immunohistochemistry AC 34BE12 positive.

colon without tumor infiltration.

The microscopic examination carried out on the different cutting levels showed a pancreatic parenchyma seat of a malignant tumor proliferation of epithelial origin edifying clusters and large nuclei of tumor cells, of polyhedral form, with well limited eosinophilic cytoplasm, centered by a basophilic, hyperchromatic nucleus, seat of cytonuclear atypia with weak mitotic activity. The tumor stroma was

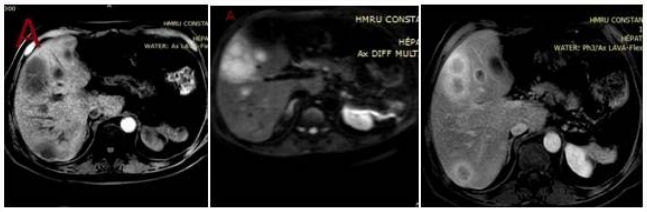

Figure 3: MRI: 7 month post-operative Liver metastasis.

fibro conjunctive of average abundance, and rich in inflammatory infiltrate diffuse lymphocytes. Furthermore, an immunohistochemy was positive for anti-34BE 12 antibodies and CK19 (Figure 2B and 2C).

The patient was referred to the oncology department for adjuvant therapy with cisplatin and gemcitabine. In the seventh postoperative month, the patient reported experiencing fever and fatigue and was admitted in an infectious disease department where an antibiotic therapy was established based on clinical manifestations and on data from a CT scan, which result perfusion disorders and nodular lesions of the liver which may be of infectious origin. After three days of hospitalization, the patient was transferred to our department. The patient was experiencing fatigue and a fever 39.5°C-40°C. Biology finds hyperleukocytosis with polynuclear >20000, thrombocytes >600000, PCR >242 mg/l, SV: 110 mm/120, the blood cultures were negative. The morphological assessment revealed a pancreatic compartment with poorly limited diffuse hypodense reorganization, no collections, and aortic lymph node 20 mm in diameter. MRI and CT scan showed a multiples liver metastasis (Figure 3). The patient was referred a second time to oncology for a new line of chemotherapy, which was not carried out due to the worsening of his clinical condition and the occurrence of death.

Discussion

Squamous cell carcinoma of the pancreas is a rare and aggressive tumor with a poor prognosis, accounting for approximately (0.5% to 5%) of all malignant tumors of the exocrine pancreas, and is characterized by the presence of at least 30% of scaly cells within the glandular elements of the pancreas [2,3].

There have been several hypotheses on the possible origins of squamous carcinoma, especially as no squamous epithelium is normally present in the pancreatic tissue. The histological transformation of pre-existing adenocarcinoma into squamous carcinoma is the most accepted theory. However, the existence of an ectopic squamous epithelium and squamous metaplasia of the pancreatic ductal epithelium are also used to explain the mechanism of occurrence of CSP [4,5]. Although the majority of adenosquamous carcinomas occur in the body or tail of the pancreas, they can still occur anywhere and their clinical manifestations are not specific and cannot be distinguished from those of other malignant tumors of the pancreas. Indeed, their clinical presentations and their evolution depend essentially on the location of the tumor and the invasion of adjacent structures [6].

Imaging of adenosquamous carcinoma of the pancreas is not specific, but the presence of a lobulated, infiltrating, and mainly cystic mass with raised edge and central necrosis. These characteristics are rarely observed in the case of pancreatic adenocarcinoma and can point towards this rare diagnosis [9-13]. Additional observations

such as portal vein thrombosis have also been described [14]. The rare incidence of adenosquamous carcinomas makes it difficult to establish a uniform therapeutic approach, but surgical resection remains the only therapeutic means offering the best chances of survival. Surgical resection is the only curative option, but is often limited by the presence of local and distant metastases [8]. Most adenosquamous carcinomas are considered to be sensitive to radiotherapy and chemotherapy; however, squamous cell carcinoma of the pancreas has a particularly poor response [15].

Chemotherapy includes 5-fluorouracil, gemcitabine, and platinum-based regimens, with reported median survival of only 3 months without curative resection [15]. The median survival is 7 months in patients treated with the protocol combining surgery, chemotherapy and radiotherapy. Tumor grade is associated with prolonged survival, patients with low or intermediate level tumors had better survival and quality of life compared to those with high grade tumors (16.2 *vs.* 3 months respectively) [1].

Conclusion

Squamous cell carcinoma of the pancreas is extremely rare. It is a misunderstood phenomenon with an unknown pathophysiology and a very poor prognosis. R0 resection, when possible, combined with chemotherapy and radiotherapy can provide a better survival rate.

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