Primary Pure Squamous Cell Carcinoma of the Duodenum: a Case Report

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ABSTRACT

Primary pure squamous cell carcinoma of the duodenum is a very rare type of duodenal neoplasm and is more likely to be presented as a metastatic tumor. The literature offers little information on this subject and includes very few articles and case reports. Laboratory tests, CT and ultrasound examinations, x-rays and immunohistochemical markers assisted us in making this rare diagnosis of primary squamous cell carcinoma of the duodenum in a 47 year old female patient, who presented with weight loss and melena. The 8 cm duodenal tumor with pancreas invasion was resected by a cephalic duodenopancreatectomy. The pathology examination revealed a primary duodenal squamous cell carcinoma moderately differentiated (G2), invasive in the head of the pancreas, with keratinization, stage II B (pT4N0MxL0V0R0). Positive outcome after surgery was highlighted, no recurrence being registered at the 6 month CT scan follow-up.

Key words: squamous cell carcinoma – duodenopancreatectomy – duodenal primary tumor.

INTRODUCTION

Generally, malignant tumors of the small intestine are rare (most of them being adenocarcinomas), but primary carcinomas are very rare neoplasms. Only 0.1–1.3% of all gastrointestinal tract neoplasms are primary carcinomas of the small intestine [1, 2]. The duodenum is the most frequent location, registering more than half of all the small intestinal carcinomas [3, 4]. The jejunum is a more frequent location than the ileum [5, 6]. Small intestinal primary squamous cell carcinomas are exceptionally rare with only a few case reports in the literature [7, 8]. Fewer than ten cases of the duodenum primary squamous cell carcinoma have been reported [9]. Small intestinal squamous cell carcinomas are more likely to be metastatic from other sites [10]. In this report, we describe an unusual case of a pure squamous cell carcinoma developed in the duodenum of a 47-year-old woman.

CASE PRESENTATION

A 47-year old woman was complaining of weight loss (5 pounds in 3 weeks), melena, pain in the right upper quadrant and right interscapular region. A computer tomography (CT) scan performed in the past revealed a 8 cm large duodenal tumor. The patient was directed to our hospital for further investigations. There was no evidence of any relevant disease in the family history.

At physical examination, she was pale and no mass was found at abdominal palpation. Laboratory tests revealed anemia (Hb= 9.3 g/dl; Ht= 29.8%), leukocytosis (WBC= 20,000/mm3). The serum level of tumoral markers was normal: CEA=1.8 ng/mL, CA 19-9=1.0 U/mL. An abdominal ultrasound was performed, and stenosis of the duodenum produced by the tumor was detected. To complete the investigations, we performed a barium swallow exploration that confirmed the duodenal stenosis (Fig. 1). An upper digestive endoscopy showed a semi-circumferential vegetative tumor in D2 and extending to D3. The chest X ray showed no tumors of the lungs. The Papanicolaou test was negative as well as breast examination, which excluded the possibility of a metastatic tumor from distant sites.

Another CT scan was performed, evidencing a 8 cm duodenal tumor extending from D2 to D3, with invasion of the head of the pancreas.
the head of the pancreas and a possible invasion of the right kidney and the inferior vena cava; the common bile duct was dilated up to 10 mm (Fig. 2). Around the tumor, the CT scan revealed 5 lymph nodes, with dimensions up to 15 mm. The liver was normal, with no focal lesions and no ascites.

We decided to perform a palliative surgery, but intraoperatively we discovered a duodenal mass (about 8/8 cm) that invaded the head of the pancreas without invasion of the right kidney, the inferior vena cava or the superior mesenteric vein and artery. Around the tumor there were a few inflammatory lymph nodes with the dimensions of 12-15 mm. A cephalic duodenopancreatectomy (Whipple procedure) was performed (Figs. 3, 4). The postoperative recovery of the patient was quick and uneventful.

The histological examination showed a primary duodenal squamous cell carcinoma moderately differentiated (G2), invasive in the head of the pancreas; pT4N0MxL0V0R0 stage II B. Keratinization was also observed. To confirm the diagnosis, additional immunohistochemical staining analyses of the duodenal lesion were performed: CK 7 (-), CK 20 (-), CA 19.9 (-), and CK 5/6 (+). Because of the high specificity of these immunohistochemical markers for squamous type, we used all of them and found that over 90% of the tumoral cells were CK 5/6 positive (Fig. 5).

**DISCUSSION**

Generally, a tumor located in the periampullary region with a diameter of 8 cm is presumed to be non-resectable and our initial pre-operative intention was to palliate the symptoms. However, the intra-operative evaluation enabled us to perform a R0 resection of the tumor through a Whipple intervention. All other sites of potential source of squamous carcinoma metastases were excluded.

In the medical literature there is little information and few cases are presented with this diagnosis. This is due to the rarity

![Fig. 1. Barium swallowing showing duodenal stenosis and widening of the duodenal frame.](image1)

![Fig. 2. CT scan. a) Inhomogeneous duodenal mass (sagittal section); b) duodenal tumor (contrast CT - transversal section); c) duodenal tumor (native CT - transversal section).](image2)
Primary pure squamous cell carcinoma of the duodenum

of this type of primary carcinoma. In his paper, Terada et al mentioned that only two cases of duodenum squamous cell carcinoma had been reported [11]. Regarding the location, the majority of duodenal carcinomas develop in the D2 segment, near the ampulla, the periampullary sites being exposed to the pancreatic juice and bile, presumed as mitogens [11].

Immunohistochemistry is useful in reaching the correct diagnosis and over the past decade, the expression levels of CK7, CK20 have been widely used to distinguish pulmonary from gastrointestinal carcinomas [12].

The pathogenetic chain leading to this type of tumor in the duodenum is unknown. There are theories which suggest that the duodenal pluripotential stem cells are the initial origin for these tumors [13]. Other theories regarding the origin of squamos cell carcinoma in the duodenum have been issued: nests of ectopic squamous cells, proliferation of uncommitted mucosal basal cells into squamous cells, squamous metaplasia secondary to chronic mucosal damage [12].

CONCLUSION

Because of the low incidence of this type of tumor, clinicians should be aware and also take into consideration rare histological tumor types located in the duodenum. Usually this tumor type is metastatic in the duodenum, but there are also primary squamous carcinomas. Even large tumors of the duodenum or periampullary region are candidates for a curative resection (R0).

Consent. Written informed consent was given by the patient for publication of this case report and any accompanying images.

Conflicts of interest. The authors declare that they have no conflicting interests.

Authors’ contributions. FG and EM conceived the case report, and wrote the manuscript. EM made substantial contributions to the acquisition and interpretation of data and drafting of the manuscript. NH gave final approval of the version to be published. All authors read and approved the final manuscript.

REFERENCES