

Pancreatic Squamous Cell Carcinoma: A Rare Pancreatic Cancer with Poor Prognosis

Rumyana Smilevska^{1,2*} and José Antonio Onrubia³

¹Department of General and Digestive Surgery, University Hospital San Juan de Alicante, Spain

²Department of Surgery and Pathology, Faculty of Medicine, University Miguel Hernández, Spain

³Department of Pathology, University Hospital San Juan de Alicante, Spain

***Corresponding Author:** Rumyana Smilevska, Department of General and Digestive Surgery, University Hospital San Juan de Alicante, Spain.

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Abbreviations

CT: Computed Tomography; PET-CT: Positron Emission Tomography/Computed Tomography; ORL: Otorhinolaryngology

Squamous cell carcinoma is a very rare type of exocrine pancreatic malignancy given that the pancreas lacks squamous cells and represents a 0,2% of all pancreatic cancers [1]. It was first described in 1949 by Lowry, *et al* [2].

The pathogenesis remains uncertain and there are different hypothesis including squamous metaplasia with malignant transformation, squamous transformation in adenocarcinoma, malignant transformation of a primitive pluripotent cell, and the tumour collision theory. Squamous metaplasia is described in histology of chronic pancreatitis and the presence of biliary or pancreatic stents [3]. There are less than 50 case reports found in the literature and a recent population based study report of 217 cases of primary pancreatic squamous cell carcinoma diagnosed between 2000 and 2010 in the United States from the Surveillance, Epidemiology, and End Results cancer registries [1].

Mean age of diagnosis is 62 - 68 years old and higher incidence rates in males are observed. There are no particular risk factors related to the squamous cell carcinoma, although the detection of squamous metaplasia in chronic inflammation suggests increased risk. The symptoms are identical with those of the ductal adenocarcinoma depending on the location and include anorexia, weight loss, abdominal or back pain, painless jaundice, melena. All parts of the pancreas are affected with predominance of the pancreatic head [1]. There are no specific radiological features, nevertheless, due to the hypervascularity of the squamous cell pancreas carcinoma, an enhancement of the tumour is seen on the CT with intravenous contrast and a tumour blush pattern on angiography [4,5].

The histology shows intercellular bridges and focal keratin pearl formation in the tumor cells. Immunohistochemistry confirms positive expression of cytokeratin 5/6 and p63. Nine mutated genes are identified in pancreatic squamous cancer tissue: C7orf70, DNHD1, KPRP, MDM4, MUC6, OR51Q1, PTPRD, TCF4, and TET2 [6]. The squamous carcinoma has a higher rate of poor differentiation compared to the adenocarcinoma [1].

There is no established optimal treatment strategy. The surgical treatment consists of a Whipple's procedure for the malignancies in the head and distal pancreatectomy is performed for the lesions that are located in the body and tail of the pancreas. The corner stone of the treatment is surgery followed by chemotherapy and few cases report also radiotherapy [7].

An overall survival of 6 - 16 months is reported [8]. In the cases where the curative resection is not possible, the median survival is 3 months [8,9]. The prognosis is reported to be poorer than the adenocarcinoma of the pancreas for overall, curative and palliative survival rates [1]. Given the poor differentiation and the rare primary of squamous cell carcinoma in the pancreas, a study with CT scans of head, neck and thorax PET-CT, oesophagogastroduodenoscopy, colonoscopy, and an ORL exam to exclude metastatic disease of other squamous primary site is recommended.

Conflict of Interest

No conflict of interest.

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