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Case Report

Primary Leiomyosarcoma of the Pancreas associated with Portal Hypertension

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Abstract

Introduction: Primary Pancreatic Leiomyosarcoma is a rare tumour. It probably originates from the smooth muscle of the pancreatic ducts or the small pancreatic vessels. Given its rarity and the scarce published cases, only few data are available regarding its epidemiological characteristics, evolution and therapeutic strategies. Common elements of the previously published cases were poor prognosis and aggressive course with early haematogenous metastasis.

Case Report: A 61-year-old man presented with subacute abdominal pain and weight loss. Abdominal computerized tomography showed a large solid mass that encompasses the body and head of the pancreas in addition to multiple solid nodular lesions in the liver compatible with metastases.

Pathology Report: Primary pancreatic leiomyosarcoma, immunologically-stained positive for caldesmon, smooth muscle actin. Chemotherapy was given with initial improvement. The patient survived for 27 months.

Conclusion: The rarity and the ominous course of primary pancreatic leiomyosarcoma impose great medical challenge. It also lacks specific clinical and imaging characteristics and evidence-based treatment strategy. The immunohistochemically diagnosis proves to be essential, due to the important differential diagnosis. Its ominous prognosis corresponds to its aggressive course and early haematogenous metastases.

Keywords: *Primary Leiomyosarcoma of the Pancreas; Rare Tumors of the Pancreas; Pancreas*

Abbreviations

AP: Pathological Anatomy; VEDA: High Digestive Videoendoscopy; IGV 1: Isolated Gastric Varices; HPT: Portal Hypertension; AFP: Alpha Fetus Protein; CEA: Carcinoembryonic Antigen; FMO: Multiorgan Failure; IHQ: Immunohistochemistry; AML: Smooth Muscle Actin; AME: Specific Muscle Actin; EBV: Epstein-Barr Virus; CT: Computed Tomography

Introduction

Primary leiomyosarcoma (PML) of the pancreas is a rare tumor. Its origin is probably the smooth muscle of the ducts and the wall of the small pancreatic vessels. To date, between 52 and 64 cases have been published in the English literature [1,2]. Given its infrequency, there are few data that accurately describe its epidemiological characteristics, its evolution and the therapeutic strategies. A common element is the poor prognosis and aggressive behavior with early metastases via the hematogenous route.

The objective of this publication is to present a case of rare pancreatic injury with a review of the literature.

Clinical Case

A 61-year-old male patient consulted in February 2014 with recent onset abdominal pain and weight loss. He ingested alcohol of 70 g/d for 20 years, with prolonged exposure to pesticides. He had no relevant family history. On physical examination, hepatomegaly 3 courses along the costal margin, with a hard elastic consistency and an irregular border, and mucous skin jaundice. The rest of the exam is normal. Given the recent onset of symptoms and a history of alcoholism, a VEDA was performed, observing a single tortuous bluish cord in the gastric fundus compatible with an isolated fundic varix (IGV 1) of the Sarin classification (Figure 1).

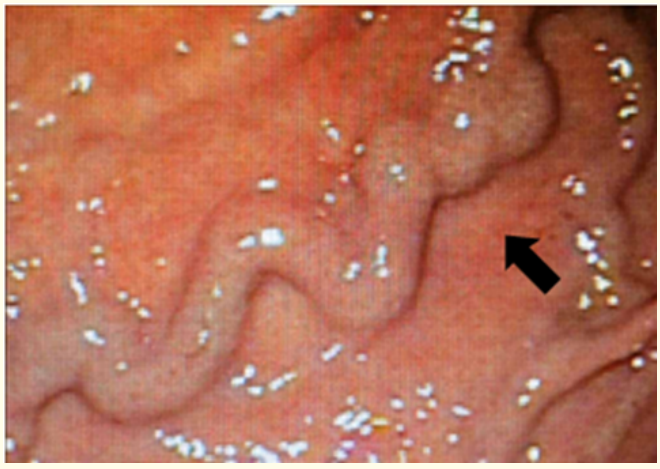

Figure 1: *Endoscopic finding of an isolated fundic varix.*

Biochemical studies are requested. The results are: Htco 30%, platelets: 156000, GOT 124 UI/ml, GPT 250 UI/ml, FAL: 770 UI/ml, GGT: 400 UI/ml, BT: 6 g/dl, BD: 3.4 g/dl, amylase: 78 IU/L and albumin 3.5 g/l. Abdominal ultrasound revealed hepatomegaly with irregular borders and the presence of multiple hypoechoic solid nodules. In the pancreas, an irregular hypoechoic formation measuring 5 x 5 cm was observed and the spleen was normal. An MRI of the abdomen was requested, where a voluminous solid mass effect was reported involving the body and head of the pancreas, measuring 10 x 8 cm and extending to the peripancreatic fatty tissue. In addition, multiple solid nodular liver lesions with peripheral contrast enhancement and rapid lavage in the late venous phase were observed. The pancreatic mass caused the obstruction of the splenoportal axis, invading the splenic vein (Figure 2 and 3).

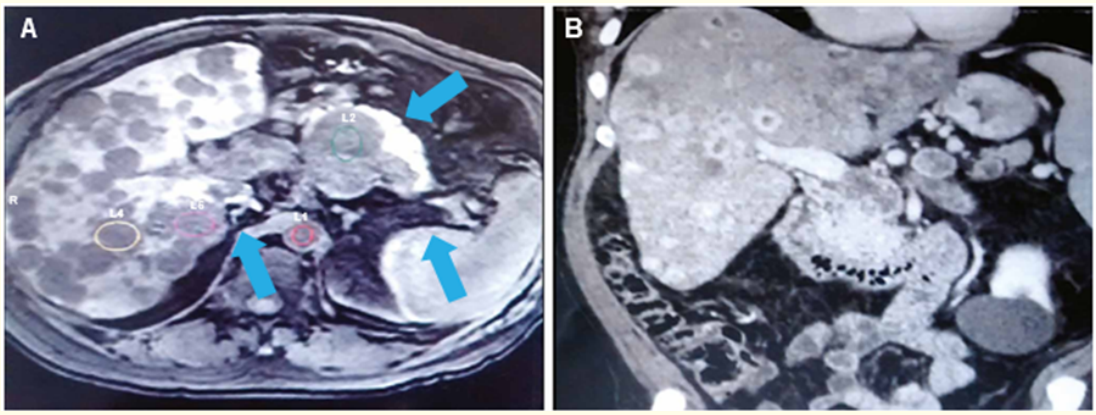

Figure 2 and 3: *MRI: A) A compromise of the pancreatic cephalic portion and multiple liver lesions is observed. B) A thrombosis of the splenoportal axis can be seen.*

Up to this point, the clinical picture has been interpreted as an oncological disease with a strong suspicion of being primary of the pancreas, with involvement of the liver and the splenoportal axis generating regional HPT. The markers Tumor CA 19-9, AFP, CEA and also 5-hydroxyindoleacetic acid were normal. A CT-guided pancreatic biopsy was performed. AP: primary leiomyosarcoma of the pancreas with caldesmon and desmin (+).

He started treatment with doxorubicin and dacarbazine (15 cycles) with good general condition, asymptomatic, anicetheric, and weight gain. A control CT scan was performed 15 months after diagnosis, where a slight improvement was observed in relation to previous liver lesions as well as pancreatic involvement (Figure 4 and 5). In addition, liver puncture was performed in order to histologically reevaluate them (Figure 6). After 27 months of diagnosis, he presented hemorrhagic ascites, dying with a picture of sepsis and MOF.

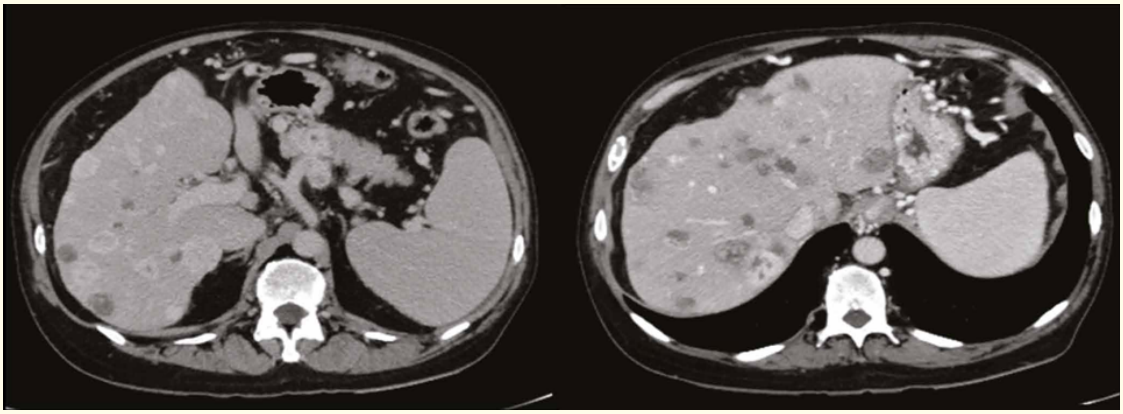

Figure 4 and 5: *Decrease in the size of both the pancreatic lesion and the liver metastases.*

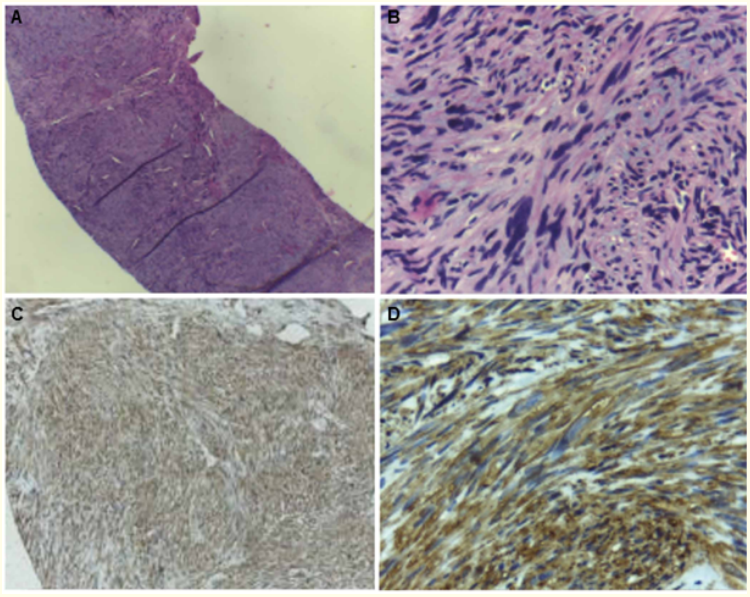

Figure 6: *Pathology and IHC of liver metastases. A) H&E 100x: a fascicular pattern of distribution of tumor fused cells is observed with small slits corresponding to their vascular network. B) H&E 400x: spindle cell tumor cells can be seen, some pleomorphic, with nuclei in the shape of a "cigar", hyperchromatic, with variable atypia, granular cytoplasm and fascicular distribution. C and D) Actin marking. Membrane and cytoplasmic staining is observed in tumor cells.*

Discussion

Pancreatic LMS is a rare mesenchymal tumor that originates in smooth muscle tissue present in the structures of the pancreas (ducts and vascular walls) [3]. Baylor., *et al*. demonstrated the presence of only five leiomyosarcomas out of 5057 malignant pancreatic tumors examined [4]. Despite its infrequency, it is the most frequent form of primary sarcomas of the pancreas [5].

The average age of presentation is 53 years, with the same distribution between men and women [6]. Clinically, it manifests with non-specific symptoms such as abdominal pain and weight loss. They constitute voluminous masses with cystic degeneration, necrosis and hemorrhage, affecting the entire gland equally [7].

With res- Regarding imaging diagnosis, it does not present specific morphological characteristics, which can lead to difficulties in differentiating it, even from pancreatic pseudocysts [8]. They are tumors with aggressive behavior. A review showed the existence of distant metastases in up to 25% of patients at the time of diagnosis and in up to 19% with involvement of neighboring organs [9]. They spread mainly by the hematogenous route, being infrequent lymphatic involvement [10]. It is important highlight the ability to simulate other mesenchymal and non-mesenchymal neoplasms. This fact requires an exhaustive IHC and even molecular study [10]. In this regard, they are characterized by expressing at least 2 smooth muscle markers (AML and SMA, caldesmon, calponin and myosin of smooth muscle) [11]. In relation to their evolution, presents an average survival of 48 months, with the impossibility of radical surgical resection being the main factor of poor prognosis [12]. Differential diagnosis should be made with sarcomatoid spindle cell carcinomas, associated smooth muscle tumors infection with EBV and other spindle cell or epithelial sarcomas including GIST (CD 117 +) [12].

Conclusion

Primary pancreatic LMS represents a rare neoplasm with nonspecific clinical and imaging features. Immunohistochemical diagnosis turns out to be essential due to the important differential diagnoses. It is a neoplasm with a poor prognosis due to its aggressive behavior with early hematogenous metastases. Finally, it is important to highlight the endoscopic diagnosis of isolated fundic varices. As we know, their presence in non-cirrhotic patients suggests the possible existence of regional HPT. In this sense, its finding should motivate the study of the splenoportal axis in search of venous thrombosis, mainly. In our patient, this finding turned out to be the initial event for the final diagnosis of primary pancreatic leiomyosarcoma.

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