Ectopic spleen and liver hemangioma mimicking metastatic pancreatic neuroendocrine tumor

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ABSTRACT

Pancreatic tumors comprise benign lesion and malignant lesion, most importantly pancreatic adenocarcinoma, acinar cell carcinoma, neuroendocrine carcinoma or metastasis. Surgical resection provides the only chance for cure for malignant pancreatic tumors. In some cases, surgical resection is performed because a malignant lesion is suspected, however, histopathological examinations eventually reveal a benign lesion. Here, we report the case of a 49-year-old woman, who was initially diagnosed with a neuroendocrine tumor of the pancreas with metastasis to the liver. The patient underwent distal pancreatectomy and atypical liver resection. Surprisingly, however, histopathological examination revealed an intrapancreatic accessory spleen (IPAS) of the pancreatic tail as well as liver hemangioma. This unique case report highlights the impact of extensive preoperative examinations to differentiate benign and malignant pancreatic lesions and, possibly, prevent patients from unnecessary surgery.

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1. Introduction

Neuroendocrine carcinoma of the pancreas (panNET) represent a rare tumor entity which amount for only 2% of all pancreatic neoplasms [1,2]. panNETs mainly develop in the pancreatic head and about 15–53% of patients suffering from panNETs become symptomatic due to hormone secretion such as insulin or gastrin [3]. The majority of all panNETs show malignant characteristics since panNETs often metastasize to the liver [1]. Importantly, in 80% of all newly diagnosed panNETs multiple liver and/or extrahepatic metastases are detected [4,5].

For tumor located in the pancreatic head a pancreaticoduodenectomy is performed in most cases. In contrast, for tumors located in the pancreatic tail a distal pancreatectomy is the surgical procedure of choice. Although a surgical approach is considered whenever a malignant, resectable and non-metastasized tumor is suspected, histopathological examinations ultimately reveal a benign diagnosis in 5–21% upon pancreaticoduodenectomies and 11% upon distal pancreatectomies [6–9].

2. Case report

We report on a 49-year-old woman with benign kidney cysts. No additional preconditions were known and the patient was not under any long-term medication. The patient underwent a follow-up MRI because of kidney cysts in 03/2014. Here, a tumor in the pancreatic tail was identified which initially showed characteristic of a benign lesion. In a subsequent MRI performed in 10/2014 the lesion had increased in size and now presented with some radiological features of a malignant tumor (Fig. 1). Therefore the patient was admitted to our hospital for further examination. The patient was asymptomatic and routine laboratory analysis did not show any pathology, including normal CA 19–9. To further characterize the pancreatic lesion a computed tomography (CT) of the abdomen was performed. Here, the lesion showed radiological characteristics of a neuroendocrine tumor (Fig. 2). Moreover, the CT scan revealed a contrast-enhanced lesion of the liver for which malignancy could not be excluded (Fig. 3).

In a subsequently performed endosonography the pancreas showed no pathological signs and the pancreatic lesion could not be identified excluding the possibility to obtain a biopsy. To further characterize the pancreatic tumor a Dotatate PET-CT was performed which revealed strong Tracer-uptake of the pancreatic lesion, strongly suggesting a neuroendocrine tumor of the pancreatic tail (Fig. 4). Subsequently, the patient underwent spleen-preserving distal pancreatectomy and atypical liver resection. Intraoperatively, the tumor presented as a soft lesion of dark
Interestingly, intraoperatively performed histopathological examination did not show malignant features of the pancreatic tumor, which was the reason why the spleen was preserved.

The postoperative course of the patient was uneventful. Surprisingly, the histopathological analysis revealed the pancreatic lesion being an intrapancreatic accessory spleen (Fig. 5) and the liver lesion being a hemangioma.

3. Discussion

Accessory spleens originate from a congenital failure of splenic bud fusion during embryogenesis [10]. This abnormality is detected in about 10–30% in autopsies. In most cases accessory spleens are located at the splenic hilum. Interestingly, CT scans reveal asymptomatic accessory spleens in 16% of patients undergoing CT scans of the abdomen [11,12]. Although, intrapancreatic accessory spleens can mimic intrapancreatic tumors, such as neuroendocrine tumors.
Fig. 4. Dotatate PET-CT showing strong Tracer uptake in the suspect lesion of the pancreatic tail.

Fig. 5. Histopathology of the resected pancreatic tissue: HE (A) staining suggesting an intrapancreatic accessory spleen, B: immunohistopathological CD20 (B) and CD3 (C) staining confirming the benign diagnose of an accessory spleen of the pancreatic tail.

Interestingly it has been reported that IPAS can mimic panNET in nuclear imaging, such as Dotatate PET-CT, due to somatostatine-positivity of intrasplenic lymphocytes [15]. Non-invasive differentiation between IPAS and panNET can be provided by using 99mTc-sulphur-colloid or 99mTc-tagged heat-damaged RBC Scintigraphy, due to selective phagocytosis by reticulum-endothelial cells in the liver and spleen [16,17]. Furthermore the suspected diagnosis of panNET was hardened by CT-imaging, which showed a contrast-enhanced lesion of the liver, for which malignancy could not be ruled out, which finally lead to atypical liver resection. Postoperatively the histopathological analysis revealed a liver hemangioma. Interestingly, it has been [13], up to date only a few cases of non-cystic intrapancreatic accessory spleen have been reported in the literature.

In cases of panNETs presenting with synchronous liver metastases surgery is usually considered in cases when complete surgical resection of the primary tumor and the metastatic lesions appears feasible. In contrast, the diagnosis of asymptomatic IPAS does not require a surgical treatment [1,2]. This fact underlines the crucial necessity to provide adequate preoperative diagnostics to prevent patients from unnecessary surgery.

In the presented case a metastatic panNET was suspected preoperatively based on the MRI and CT scan as well as on a Dotatate PET-CT, which lead to unnecessary distal pancreatectomy.
reported that sclerosed hemangioma can mimic liver metastasis of gastrointestinal cancers [14].

In conclusion, we consider IPAS a relevant differential diagnosis in patients with suspected panNET, which should be taken into account before surgery is provided.

Conflicts of interest
All authors declare no conflict of interest.

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Not applicable.

Author contribution
C.C.E., M.K. and D.H.-B. designed the study. C.C.E and J.L. performed the research and wrote the manuscript. T.F.B. provided pathological analysis. S.A.S. provided radiological analysis. All authors read and approved the manuscript.

Consent
Not applicable.

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