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An unusual presentation of accessory spleen mimicking a pancreatic neoplastic lesion in a splenectomized patient: A case report

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Abstract

Accessory spleen is a relatively common occurrence. However, an intrapancreatic accessory spleen can get hypertrophied following splenectomy and rarely mimic a pancreatic neoplasm leading to misdiagnosis. Here we present a 64-year-old male who had undergone left radical nephrectomy and splenectomy for renal cell carcinoma 14 years back, presenting with upper abdominal discomfort. He was found to have a mass in the pancreatic tail on imaging, suggesting an intrapancreatic neoplastic lesion. After a multidisciplinary team decision based on contrast-enhanced computed tomography and magnetic resonance imaging, he underwent an uncomplicated distal pancreatectomy, and the histology revealed an intrapancreatic accessory spleen. Contrast-enhanced computed tomography, magnetic resonance imaging, and positron emission tomography alone is not specific enough to confidently differentiate an accessory spleen preoperatively. Nuclear scintigraphy fused with contrast-enhanced computed tomography provides more specific and better anatomically localized evidence. Ultrasound-guided fine needle sampling showing lymphocytes with subsets of histiocytes, plasma cells, and immunohistochemistry showing CD8 (cluster of differentiation) positivity can be used to guide the definitive diagnosis. Differentiating an accessory spleen from a pancreatic neoplasm may be challenging preoperatively. Accessory spleen needs to be considered in the differential diagnosis of upper abdominal masses especially in patients who have undergone splenectomy.

Keywords

Intrapancreatic accessory spleen, accessory spleen, splenectomy, pancreatic neoplasm, case report

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Introduction

Accessory spleen is a condition where healthy splenic tissue lie separated from the main body of the spleen. It is formed during embryological development of fetus due to failure of mesenchymal remnants to fuse with the main splenic mass.[1] It is a relatively common condition where around 10%–30% of population may have accessory spleens according to an autopsy study.[1] Most common anatomical locations of occurrence are splenic hilum and pancreatic tail followed by greater omentum, stomach and related to other parts of the gastrointestinal tract.[1] Rarely, it is found in the pelvis or even in the retroperitoneal space.[2,3] Accessory spleens may be multiple as well.[4]

Usually, accessory spleen is an asymptomatic benign condition which is detected incidentally during imaging and does not require any surgical treatment. However, following splenectomy, accessory spleens may undergo compensatory hypertrophy and cause symptoms. Furthermore, when accessory spleen lies within the pancreas, it may mimic an intrapancreatic neoplasm. Therefore, having accurate preoperative differential diagnosis and scientifically excluding them is imperative to avoid unnecessary surgical morbidity. Herein, we report an accessory spleen in relation to the pancreatic tail which was misinterpreted as a pancreatic neoplasm preoperatively.

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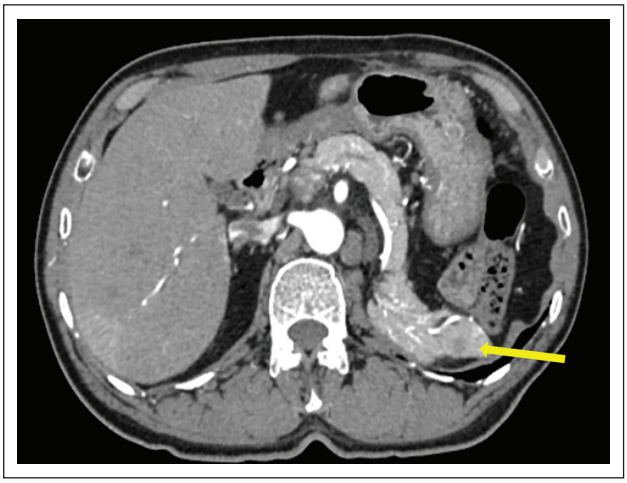

Figure 1. Computed tomography showing a contrast enhancing focal lesion (arrow) in the region of the pancreatic tail.

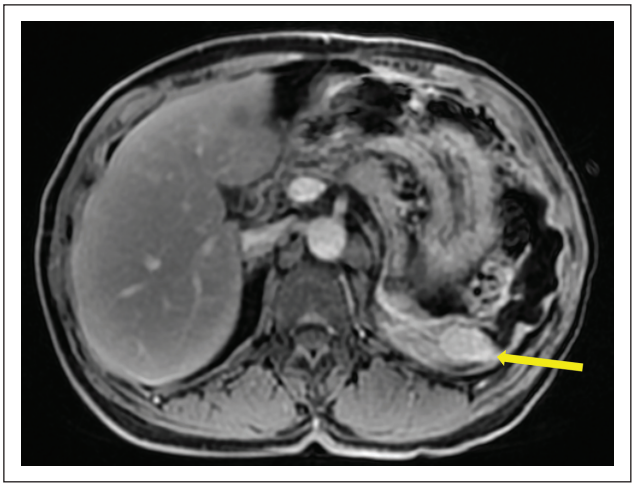

Figure 2. Magnetic resonance image of the upper abdomen that was reported as a lobulated malpositioned pancreatic tail simulating a mass.

Case presentation

A 64-year-old male who had undergone left-sided radical nephrectomy and splenectomy 14 years ago for renal cell carcinoma (RCC) with defaulted surgical follow-up, presented to our surgical clinic with vague upper abdominal pain and discomfort for 4 months. He underwent radical excision of RCC, and adjuvant therapy was not given. He was on levetiracetam regular dose for epilepsy with a good symptomatic control and otherwise, his past medical history was unremarkable. His family history was insignificant. His height was 168 cm and weight was 72 kg with a body mass index of 25.5 kgm^{-2}. Rest of his examination including the abdomen was unremarkable.

He underwent a contrast-enhanced computed tomography (CECT) which showed a contrast enhancing focal lesion in the region of the pancreatic tail with another contrast enhancing smaller nodule in anterior peripancreatic tissue (Figure 1). The radiological diagnosis was a primary or secondary malignant pancreatic neoplasm with an enlarged lymph node in the peripancreatic area. In view of further delineation, he underwent a magnetic resonance imaging (MRI) of the upper abdomen which reported a lobulated malpositioned pancreatic tail simulating a mass (Figure 2). Smaller lesion seen separately was suggested to be lymph node or splenunculus. There was no evidence to suggest a recurrence over the left renal bed and there was no liver metastasis.

The clinical and radiological findings were discussed at the multidisciplinary team meeting that included radiologists and hepatopancreatobiliary surgeons. Due to its hyperenhancing nature, pancreatic neuroendocrine tumor (NET) and metastasis from previous known malignancy were considered in the differential diagnoses. However, metastatic RCC was considered less likely because of the long lag of 14 years and because the renal malignancy was staged pT2N0M0. Consequently, a distal pancreatic neoplasm with a small supra pancreatic lymph node was deemed the likely diagnosis, and surgery was planned.

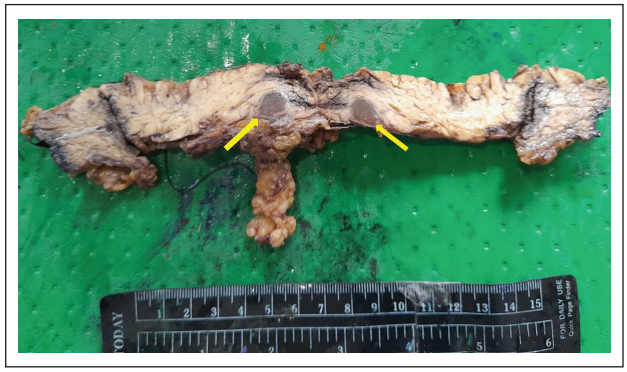

Figure 3. A cut open macroscopic specimen showing the pancreatic tail mass.

An open surgical approach was considered. Intraoperatively, splenic bed was empty suggestive of previous splenectomy. Distal pancreas was mobilized from retroperitoneal tissues and from the adhesions at the posterior wall of the stomach. Pancreatic tail with the mass was separated from pancreatic body and sent for histology which revealed vascular sinuses lined by endothelial cells resembling the red pulp and lymphoid aggregates with secondary follicles resembling the white pulp (Figures 3 and 4). Pancreatic parenchyma and duct system were histologically normal. Peripancreatic smaller nodule was the condensed fibrofatty tissue. Therefore, the histological conclusion was an intrapancreatic accessory spleen.

The postoperative period was rather uneventful and he was discharged on day 5 after surgery. Postoperatively, he developed thrombocytosis possibly indicating that the intrapancreatic accessory spleen was the only functioning splenic tissue in this patient.

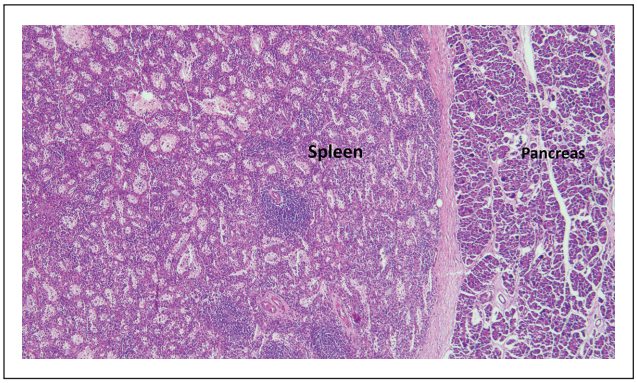

Figure 4. Image showing the histological appearance of the accessory spleen within the pancreatic tissue.

Discussion

The prevalence of intrapancreatic accessory spleens is reported to be 11%–17% of all accessory spleens located in the pancreatic tail region. Therefore, the estimated prevalence of intrapancreatic accessory spleens is 1.1%–3.4%, as accessory spleens are reported in around 10%–20% of individuals.[5] Since accessory spleen is a relatively common benign occurrence which does not require any surgical intervention, preoperative accurate diagnosis can prevent unnecessary surgery. Mortele et al.[6] reported the typical CECT findings of accessory spleen in non-splenectomized individuals. The anteroposterior diameter varied from 4 to 29 mm with a median of 11.9 mm. Around 78.3% of them were round in shape, 15% were ovoid, and 6.7% were triangular with well-defined margins. However, in patients who have undergone splenectomy, the accessory spleen undergoes compensatory hypertrophy to supplement the physiological demand, rendering these parameters unreliable. Homogeneous enhancement was also a characteristic feature of accessory spleens, differentiating them from most neoplasms. Most accessory spleens had same echogenic enhancement to main spleen on contrast-enhanced images, but almost one-third of accessory spleens were hypodense. Therefore, contrast enhancement is a nonreliable feature for differentiation. Therefore, high clinical suspicion along with more specific investigations are necessary for an accurate preoperative diagnosis.

More specific radiological diagnosis of ectopic splenic tissue can be achieved by nuclear scintigraphy using technetium-99 m-labeled Sulfur colloid.[7] However, low resolution of this imaging modality makes it difficult to delineate the exact anatomical location. Combined CECT imaging along with nuclear studies were used increasingly to diagnose accessory spleen accurately during preoperative workup.[7] This relatively low-cost investigation can prevent patients undergoing unnecessary major surgery by solving the dilemma which could not be achieved by CECT, MRI, or positron emission tomography (PET) in isolation.

Further confirmation of the diagnosis can be achieved using endoscopic ultrasound (EUS)-guided fine needle aspiration (FNA) especially in the context of intrapancreatic lesions.[8] FNA showing lymphocytes with subsets of histiocytes and plasma cells have been reported in accessory spleens.[8] Furthermore, immunohistochemistry showing CD8 positivity due to the presence of splenic sinus endothelial cells have been reported to aid in the diagnosis of accessory spleen.[9] In our resource-limited setting, advanced imaging such as PET scan was not performed as they are reserved for patients with proven malignancies due to the resource constraints.[9] EUS-guided FNA could not be performed due to nonavailability. Therefore, we proceeded according to the multidisciplinary team decision, which was based on CECT and MRI and favored a diagnosis of a pancreatic neoplasm.

This case report describes the challenges faced in a resource-limited setting to differentiate an intrapancreatic accessory spleen from a pancreatic neoplasm. This is specifically difficult in a patient who has already undergone a splenectomy for a previous malignancy where the suspicion for a recurrence is high. Accessory spleens within the pancreas may mimic NETs. NETs may secrete one or more products associated with a clinical syndrome (functional) or may be nonsecretory (nonfunctional).[10] Biochemical analysis will be helpful to confirm the presence of a functional NET however, it will not be possible to exclude nonsecreting tumors. Therefore, due to the resource constraints and nonavailability of investigations in the state sector, biochemical assessment was not performed.

Conclusion

The challenges faced in a resource-limited setting to differentiate an intrapancreatic accessory spleen from a pancreatic neoplasm were described in this report. Differentiation is specifically difficult in a patient who has already undergone a splenectomy for a previous malignancy where the suspicion for a recurrence is high. Accessory spleen can enlarge after splenectomy due to compensatory hypertrophy and should be considered a differential diagnosis in patients who present with upper abdominal masses. In our patient, CECT and MRI could not accurately diagnose an intrapancreatic accessory spleen. In the context of readily available resources, nuclear scintigraphy and EUS-guided FNA could have been used for confirmation preoperatively, avoiding unnecessary major surgery and related morbidity.

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None.

Authors contributions

Authors P.P., M.N., D.B., and U.J. contributed to the collection of information and writing of the article. Authors M.N. and U.J. contributed to writing and final approval of the article. All authors read and approved the final version of the article.

Availability of data and material

All data generated or analyzed during this study are included in this published article.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient for anonymized information and accompanying images to be published in this article. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

Not applicable.

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References

1. Guo W, Han W, Liu J, et al. Intrapancreatic accessory spleen: a case report and review of the literature. *World J Gastroenterol* 2009; 15(9): 1141–1143.
2. Taskin MI, Baser BG, Adali E, et al. Accessory spleen in the pelvis: a case report. *Int J Surg Case Rep* 2015; 12: 23–25.
3. Palumbo V, Mannino M, Teodoro M, et al. An extremely rare case of an oversized accessory spleen: case report and review of the literature. *BMC Surg* 2019; 19(1): 45.
4. Wang W, Li W, Sun Y, et al. Intra-gastric ectopic splenic tissue. *J Gastrointest Surg* 2016; 20: 218–220.
5. Li B-Q, Xu X-Q and Guo J-C. Intrapancreatic accessory spleen: a diagnostic dilemma. *HPB* 2018; 20(11): 1004–1011.
6. Mortelé KJ, Mortele B and Silverman SG. CT features of the accessory spleen. *AJR Am J Roentgenol* 2004; 183(6): 1653–1657.
7. d'Amico A, Cofalik A, Przeorek C, et al. Role of nuclear medicine imaging in differential diagnosis of accessory spleens in patients after splenectomy. *Pol J Radiol* 2012; 77(1): 68–71.
8. Schreiner AM, Mansoor A, Faigel DO, et al. Intrapancreatic accessory spleen: mimic of pancreatic endocrine tumor diagnosed by endoscopic ultrasound-guided fine-needle aspiration biopsy. *Diagn Cytopathol* 2008; 36(4): 262–265.
9. Jayarajah U and Abeygunasekera AM. Cancer services in Sri Lanka: current status and future directions. *J Egypt Natl Cancer Inst* 2021; 33(1): 13.
10. Aluri V and Dillon JS. Biochemical testing in neuroendocrine tumors. *Endocrinol Metab Clin North Am* 2017; 46(3): 669–677.