JOP. J Pancreas (Online) 2013 May 10; 14(3):261-263.

CASE REPORT

Intrapancreatic Accessory Spleen. A Case Report and Review of Literature

Jana Katuchova[1], Helena Baumohlova[2], Pavol Harbulak[1], Miloslava Stofcikova[3], Marian Svajdler[2], Andrej Repovsky[3], Jozef Radonak[1]

[1]First Department of Surgery and [2]Department of Patology, University Hospital and Safarik University. Kosice, Slovakia. [3]Diagnostic Center. Humenne, Slovakia

ABSTRACT

Context Accessory spleen is a congenital focus of healthy splenic tissue that is separated from the main body of spleen. Although an accessory spleen usually appears as an isolated asymptomatic abnormality, it may have clinical significance in some situations. **Case report** We report the case of 53-year-old woman with a 2-year history of upper abdominal discomfort after meals and weight loss. The pathologic lesion was diagnosed by the abdominal sonography and the magnetic resonance tomography in the pancreatic tail. The patient was operated with suspicion of a solid pseudopapillary neoplasm or a nonfunctioning islet cell tumor. Histopathological examination found an intrapancreatic accessory spleen, which is a congenital abnormality consisting of normal splenic tissue in ectopic sites. **Conclusion** We present possibilities of differential diagnosis of this entity.

INTRODUCTION

Accessory spleen is a congenital abnormality consisting of normal splenic tissue in ectopic sites that is found in approximately 10-15% of the general population [1]. The most common location of an accessory spleen is the splenic hilum. Despite that the second most common site of the accessory spleen is the tail of the pancreas, the intrapancreatic accessory spleen is not always recognized preoperatively. Radiologically, intrapancreatic accessory spleen appears to be a well-defined, solitary and hypervascular lesion. The differential diagnosis includes well-differentiated adenocarcinoma, mucinous cystic neoplasm, neuroendocrine neoplasm, solid pseudopapillary tumor or metastatic tumor to the pancreas [2].

We report the case of 53-year-old woman with a mass in the pancreatic tail, diagnosed by the abdominal sonography and the magnetic resonance tomography (MR). The patient was operated with suspicion of a solid pseudopapillary neoplasm or a nonfunctioning islet cell tumor. Histopathological examination describes intrapancreatic accessory spleen.

Received October 9th, 2012 – Accepted Februaty 4th, 2013

Key words Diagnosis, Differential; Pancreas; Pancreatic Neoplasms; Spleen

Correspondence Jana Katuchova
First Department of Surgery; University Hospital and Safarik University; Tr. SNP 1; Kosice, 04011; Slovakia
Phone: +421-55.640.3896; Fax: +421-55.640.3808
E-mail: jana.katuchova@upjs.sk

CASE REPORT

A 53-year-old woman was admitted to a local hospital with a 2-year history of upper abdominal discomfort after meals and weight loss. Physical examination and laboratory data including peripheral blood counts, blood sugar and liver function test were all unremarkable. Tumor markers, including CA 19-9, CA 125, carcino-embryonic antigen (CEA) and alpha-fetoprotein were within the normal range. Abdominal sonography was performed, and a well-circumscribed tumor 2.1x2.3 cm with a low echogenicity was noted in the pancreatic tail. MR confirmed a 1.7x2.1 cm mass in the pancreatic tail (Figure 1). A nonfunctional islet tumor or a solid pseudopapillary neoplasm of pancreatic tail was suspected on June 6th, 2012. Although no definitive diagnosis was established preoperatively, the possibility of malignancy could not be ruled out. Consequently, a laparotomic exploration was arranged to treat the lesion. A distal spleen preserving hemipancreatectomy was performed (Figure 2). During histopathological examination the mass was established as intrapancreatic accessory spleen (Figure 3). The tumor size was 2.1x1.7x1.0 cm. Within a macroscopic examination, accessory spleen was surrounded by a fibrotic capsule that separates the spleen from the adjacent pancreatic parenchyma. In the histologic analysis intrapancreatic accessory spleen was composed of red and white pulp that is similar to that of the normal spleen (Figure 3). The red pulp was made from numerous vascular sinuses. Between these sinuses the lymphoid follicles and the cells of the

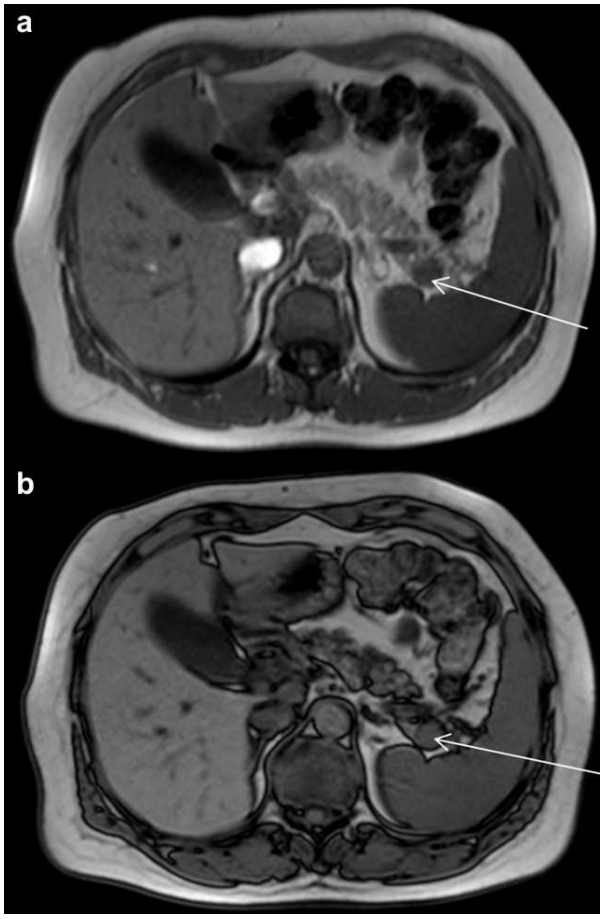

Figure 1. Magnetic resonance imaging of intrapancreatic accessory spleen (arrows). T2W (**a.**) and T1W (**b.**) images show a hyperintense lesion in the tail of the pancreas.

reticuloendothelial system lie, which constitute the white pulp altogether.

Six moths after the operations the patient is doing well without digestive problem.

DISCUSSION

Accessory spleen is a relatively common congenital defect, found in 10-30% of patients at autopsy, and is due to the fusion failure of splenic anlage, which is located in the dorsal mesogastrium to fuse. Most of the cases are located near the splenic hilum and the tail of the pancreas. They are normally asymptomatic, and operative intervention is not indicated. Although an accessory spleen usually appears as an isolated asymptomatic abnormality, it may have clinical significance in some situations [3, 4, 5].

Peripheral blood counts, blood sugar, liver function test and tumor markers tests (CA 19-9, CA 125, CEA, alpha-fetoprotein) are indicated for patients with upper gastrointestinal symptoms in the diagnostic process. The diagnostic imaging methods include abdominal sonography, computed tomography (CT), MR and endoscopic ultrasound (EUS) with a larger number of pancreatic lesions [5, 6, 7]. The most important differential diagnosis of intrapancreatic accessory spleen is the asymptomatic pancreatic neuroendocrine neoplasms. Both intrapancreatic accessory spleens and pancreatic neuroendocrine neoplasms are typically hyperenhancing on contrast enhanced CT [8]. Pancreatic neuroendocrine neoplasms are typically hypervascular, particularly on the arterial phase. On the other hand, when the mass within the tail of pancreas is small, the pattern of contrast enhancement may be difficult to analyse on CT. An alternative but uncommon differential diagnostic is metastasis to the pancreas, especially metastases from renal cell carcinomas. The hypervascular appearance of metastases from renal cell carcinomas has a similar appearance to intrapancreatic accessory spleen. Clinical history of the renal cell carcinoma helps to make a correct diagnosis. The splenic artery aneurysm may also stimulate intrapancreatic accessory spleen [8, 9]. Yamaoka *et al*. referred about an adenocarcinoma in intrapancreatic accessory spleen. The authors wrote about an 8 cm well-demarcated tumor, which was heterogeneously enhanced during the early vascular phase [10]. When an accessory spleen is located in the pancreas, intrapancreatic accessory spleen is located within 3 cm of the distal tail of the pancreas. If a solid pancreatic tail mass is located more close to the body of the pancreas, it is unlikely to represent intrapancreatic accessory spleen, and more likely to represent other mass such as the pancreatic neuroendocrine neoplasm. It may mimic an islet cell tumor, pseudopapillary neoplasm or well vascularised metastases [11].

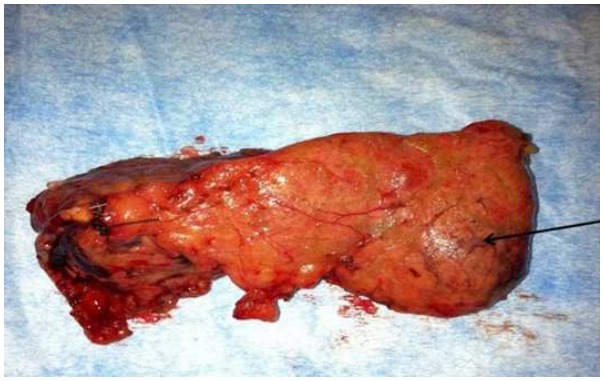

Figure 2. Spleen preserving distal hemipancreatectomy with intrapancreatic accessory spleen in the tail of pancreas (arrow).

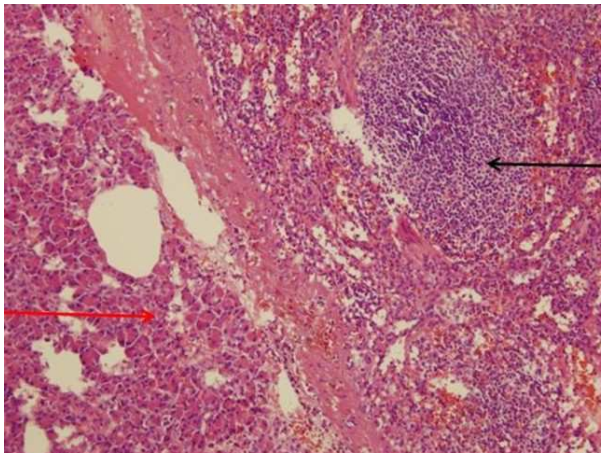

Figure 3. Histopathological detection of the intrapancreatic accessory spleen (black arrow) in the tail of the pancreas (red arrow).

During differential diagnosis for the evaluation of pancreatic lesions, endoscopic ultrasound-guided fine-needle aspiration biopsy is a very sensitive test [12, 13]. Maldi *et al*. found that endoscopic ultrasound-guided fine-needle aspiration biopsy is a safe and sensitive test that allows an accurate diagnosis of intrapancreatic accessory spleen, ruling out neuroendocrine tumor [12].

Epidermoid cyst in intrapancreatic accessory spleen is exceedingly rare. Usually these patients underwent a surgical resection because they were diagnosed to have pancreatic tumor, which were possibly malignant cystic tumors. If epidermoid cysts of accessory spleen can be diagnosed accurately, and are small and asymptomatic, surgery may not be necessary [14, 15]. On the other hand when a malignancy in pancreatic lesion or in intrapancreatic accessory spleen cannot be ruled out, surgical intervention is strongly recommended [10].

Gomi *et al*. reported about the present of splenopancreatic abnormalities in non trisomy 13 subjects and in trisomy 13. They found only five cases of intrapancreatic accessory spleen and two cases of fusion of the pancreatic tail and splenic hilus and/or accessory spleen among 1,060 non trisomy 13 subjects. In comparison, all 21 trisomy subjects had intrapancreatic accessory spleen and/or of fusion of the pancreatic tail and splenic hilus and/or accessory spleen. The authors illustrated the close relationship between occurrence of intrapancreatic splenic tissue and/or fusion of the pancreatic tail and splenic hilus and/or accessory spleen and trisomy 13. Gomi et al. also showed usefulness of this relationship in differentiating trisomy 13 from other malformation syndromes, even on macroscopic examination [16].

In our case of the patient with abdominal discomfort and loss of weight, we could not safely exclude malignancy. Therefore the patient underwent spleen preserving distal hemipancreatectomy.

CONCLUSION

In conclusion, when an asymptomatic intra-pancreatic mass is detected, the possibility of accessory spleen should be considered. The use of CT, MRI and the endoscopic ultrasound-guided fine-needle aspiration biopsy may detect intrapancreatic accessory spleen, and one can avoid unnecessary surgery. Surgical intervention is strongly recommended for pancreatic lesions or intrapancreatic accessory spleens when a malignancy cannot be eliminate.

Acknowledgments This paper was supported by the Centre of Excellence for Electromagnetic Fields in Medicine (CEEPM) project 26220120067

Conflict of interest There is no commercial association that might pose a conflict of interest. The customary listing of sources of support and institutional affiliations at the end of a manuscript is proper and does not imply a conflict of interest.

References

1. Wadham BM, Adams PB, Johnson MA. Incidence and locations of accessory spleens. N Engl J Med 1981;304(18):1111.

2. Spencer LA, Spizarny DL, Williams TR. Imaging features of intrapancreatic accessory spleen. Br J Radiol 2010;83(992):668-673.

3. Guo W, Liu J, Li JS et al. Intrapancreatic accessory spleen: A case report and review of the literature. World J Gastroenterol 2009;15(9):1141-1143.

4. Schwartz TL, Sterkel BB, Meyer-Rochow GY et al. Accessory spleen masquerading as a pancreatic neoplasm. Am J Surg 2009;197(6):e61-63.

5. Matthaei H, Schmelzle M, Braunstein S et al. Pancreatic incidentalomas: a growing clinical challenge exemplified by an intrapancreatic accessory spleen. Wien Klin Wochenschr 2011;123(5-6):186-188.

6. Park JS, Kim WJ, Jeong YG et al. A case of intrapancreatic accessory spleen mistaken as a pancreatic mass due to different enhancin pattern from normal spleen. Korean J Gastroenterol 2011;58(6):357-360.

7. Zeman M, Zembala-Nozynska E, Sczasny J et al. Intrapancreatic accessory spleen imitating a pancreatic neoplasm. Pol Przegl Chir 2011;83(10):568-570.

8. Kawamoto S, Johnson PT, Hall H et al. Intrapancreatic accessory spleen: CT appearance and differential diagnosis. Abdom Imaging 2012;37(5):812-827.

9. Yamaoka R, Nishihira T, Shimada T et al. Adenocarcinoma in an intrapancreatic accessory spleen: Report of a Case. Surg Today 2011;41(11):1552.1555.

10. Sothilingam K., Leedahl T, Krieger S et al. Intrapancreatic accessory spleen: A case report and review of the literature. Int J Surg Case Rep 2011; 2(6):128-130.

11. Maldi E, Carucci P, Pacchioni D et al. Endoscopic ultrasound fine-needle aspiration in the diagnosis of intrapanreatic accessory spleen. Dig Endosc 2011;23(3):270.

12. Schreiner AM, Mansor 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.

13. Rodriguez E, Netto G, Li QK. Intrapancreatic accessory spleen: A case report and review of literature. Diagn Cytopatholl (in press), http://onlinelibrary.wiley.com/doi./10.1002/dc22813/.

14. Itano O, Chiba N, Wada T et al. Laparoscopic resection of an epidermoid cyst originating from an intrapancreatic accessory spleen: report of a case. Surg Today 2010;40(1):72-75.

15. Hu S, Zhu L, Song Q. et al. Epidermoid cyst in intrapancreatic accessory spleen: computed tomography findings and clinical manifestation. Abdom Imaging 2012;37(5):828-833.

16. Gomi K., Sato Y., Tanaka M. et al. Specificity of splenopancreatic field abnormality in trisomy 13 syndrome: Macroscopic and histological analysis in 21autopsy cases. Pathol Int 2009; 59(3): 147–151.