



Intrapancreatic Accessory Spleen - Proper Diagnosis Prior the Surgery

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Abstract

The aim of this study was to summarize the diagnosis and treatment of Intrapancreatic Accessory Spleen (IPAS) and differentiating between IPAS from Pancreatic Neuroendocrine Tumor (PNET). False diagnosis of PNET is still associated with unnecessary surgery.

Keywords: Accessory spleen; Intrapancreatic accessory spleen; Neuroendocrine tumor; 68Ga-DOTANOC PET/CT; 99mTc-HDRBC scintigraphy

Introduction

The incidence rate of Pancreatic Neuroendocrine Neo-Plasms (PNET) or Pancreatic Neuroendocrine Tumors (PNET), both Functional (F-PNET/PNET) and Non-Functional (NF-PNET/PNET), is constantly increasing, and is currently approximately 0.32/100,000 people/year. PNETs account for approximately 30% of all Gastro-Entero-Pancreatic Neoplasms (GEP NENs) [1-6]. Approximately 35% of gastrointestinal NETs are placed in the pancreas presenting with or without clinical symptoms [7,8]. The development of new diagnostic methods observed in recent years has contributed changes in the detection of PNET, but diagnosis can still be challenging. Intrapancreatic Accessory Spleen (IPAS) is one of the lesions usually diagnosed occasionally and with diagnosis difficulty because they imitate neuroendocrine neoplasm. An alternative but uncommon differential diagnostic consideration is metastasis to the pancreas. An important pitfall to recognize is that non-neo-plastic hypervascular lesions, such as splenic artery aneurysm, may simulate IPAS [9].

Accessory Spleen (AS) is a frequent congenital abnormality (10% to 20%) that have separated from the main body of the spleen, as a result of the fusion failure of multiple splenic anlagen located in the dorsal mesogastrium during the fifth week of the embryonic life [10]. The location of AS varies, but includes splenic hilum (80%), the tail of the pancreas (IPAS), less commonly the greater omentum and the splenic ligament, wall of the ileum, mesentery, adnexa or scrotum [11]. The diagnosis of AS can be important in three clinical situations. First IPAS can mimic an adenopathy or an abdominal tumor with a possible pancreatic malignancy and most common preliminary diagnosis was underlined inactive neuroendocrine tumors. Second, an AS can occasionally become symptomatic in case of torsion, rupture or hemorrhage. Third, the surgeon must be warned of the existence of one or more accessory spleens when a total splenectomy is recommended like in some cases of hematologic disorders [12]. The wandering accessory spleen is an extremely rare appearance with few cases described in the literature and in which an accessory spleen with a long vascular pedicle is mobile in the abdomino-pelvic cavity [12-14].

Medical Examination

Autopsy studies informed that IPAS is rarely recognized radiologically. With recent improvements in imaging techniques IPAS is more commonly detected on imaging studies, but still not characteristic. In Ultrasound (US), Computer Tomography (CT) or Magnetic Resonance Imaging (MRI) usually present round or oval, well vascularized and well delineated from the pancreatic parenchyma [9]. However, computed tomography with contrast agent and conventional magnetic resonance are limited in this evaluation mainly in injuries smaller than 10 mm [15]. On abdominal US, IPAS is usually, homogeneous, hypoechoic mass with posterior enhancement. The presence of a vascular hilum entering the lesion is a diagnostic feature of AS with sensitivity of 90% [16]. On CT, IPAS appears as a well- circumscribed mass with an arciform pattern of enhancement

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due to varying flow rates of contrast through the red and white pulp. Whereas for Magnetic Resonance Imaging (MRI), IPAS is shown as a low signal intensity mass on T1-weighted images and a high-signal intensity mass on T2-weighted images [11]. Magnetic resonance combined with diffusion-weighted phase revealed to be a high accurate method in diagnosis and differentiation of intrapancreatic ectopic spleen and small solid pancreatic tumors. The intrapancreatic ectopic spleen normally is seen in magnetic resonance of hyperintense diffusion-weighted in T2 and hypointense weighted in T1 compared to normal pancreatic tissue [15]. The ^{68}Ga -DOTA-TOC PET/CT is a high specificity method for diagnosis of pancreatic neuroendocrine tumors because there is an important expression of receptors of somatostatin on lymphocytes and it can allow to distinguish between IPAS and PNET [17,18]. Other diagnostic modality, using functional imaging of splenic tissue such as $^{99\text{m}}\text{Tc}$ -HDRBC scintigraphy, ultrasound imaging following intravenous administration of a contrast agent or magnetic resonance imaging following administration of superparamagnetic contrast agent (SPIO- enhanced MRI) may be helpful. Scintigraphy with marked erythrocytes with technetium-99 is one of the most specific methods to diagnose intrapancreatic ectopic spleen. Marked erythrocytes are injected with radiopharmacy and more than 90% of the material is uptake by the splenic tissue, therefore contributing significantly to detect intrapancreatic splenic tissue and differentiate neuroendocrine pancreatic tumors and, mainly, avoid unnecessary surgical procedure [11].

Discussion

It is highly preferable if diagnosis of IPAS is non-invasive and allows to avoid unnecessary surgery and decrease possible patient morbidity and mortality.

Bing-Qi Li et al. [19] summarized the epidemiologic features, clinical manifestations, tumor biomarker abnormalities, imaging characteristics of IPAS based on a systemic review of 74 full articles including 105 patients. More than 50% patients with IPAS were diagnosed by postoperative pathology and underwent unnecessary operation. Because most IPASs are asymptomatic and do not pose clinical threat, the majority of IPASs do not require any medical treatment. Osher et al. [20] reports based of the 46 case reports of IPAS for which the imaging methodology employed is still in use 17 were correctly diagnosed as IPAS and were therefore not referred for surgery. On the other hand, 29 subjects were misdiagnosed with PNET and were operated on. All subjects with IPAS incorrectly diagnosed as PNET underwent distal pancreatectomy. Authors sum up that the IPAS patients had nonspecific presenting symptoms, similar and nearly equal sex distribution and typically manifest the 6th decade of life. Clinical symptoms or signs are not enough for differential diagnosis. IPAS is mainly located at the pancreatic tail as in 98% of the case reports reviewed here. In contrast, PNETs appear to be located throughout the gland at a Head: Body: Tail ratio of 7:1:1.5. Large lesion size is generally not compatible with IPAS but may be seen in PNETs. In the present survey, the size of IPAS as determined by imaging (CT, MRI, or EUS), did not exceed 3 cm in the largest diameter. For this reason, the possibility of IPAS should be carefully considered for lesions <3 cm. Tc-99mc- HDRBC scintigraphy can be useful to identify a focal high concentration of red blood cells such as seen in splenic tissue [20].

Rufini et al. [21] evaluated the value of ^{68}Ga -DOTANOC PET/CT in the diagnostic workup of patients with suspected PNET on the

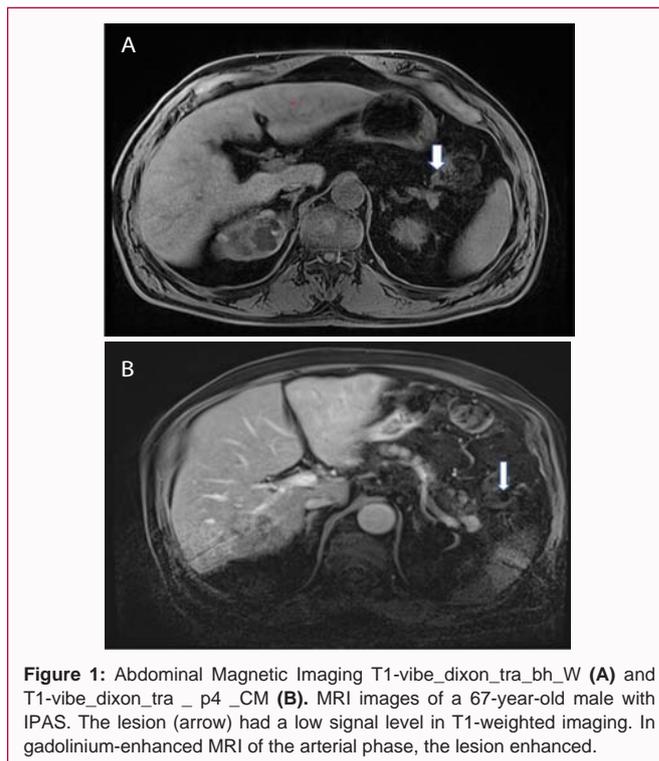


Figure 1: Abdominal Magnetic Imaging T1-vibe_dixon_tra_bh_W (A) and T1-vibe_dixon_tra_p4_CM (B). MRI images of a 67-year-old male with IPAS. The lesion (arrow) had a low signal level in T1-weighted imaging. In gadolinium-enhanced MRI of the arterial phase, the lesion enhanced.

basis of clinical and/or biochemical and/ or imaging data. Authors reported that ^{68}Ga -DOTANOC PET/CT was positive in 48 patients suspected PNET, negative in 7, and not evaluable in 3. Author's data indicate that up to 17% (8 patients) with positive ^{68}Ga -DOTANOC PET/CT in the pancreas and a suspicion of PNET may not have a PNET lesion. IPAS was confirmed in 6 patients (13%). Tc-99mc-HDRBC scintigraphy could have confirmed the diagnosis of IPAS and it's recommended. Bhutiani et al. [22] reported seven patients who underwent surgical resection of an intrapancreatic spleen. The most common pre-operative diagnosis was a Non-Functioning Pancreatic Neuroendocrine Tumor (NF-PNET). The most common operative approach was laparoscopic distal pancreatectomy and splenectomy.

Conclusion

An asymptomatic intra-pancreatic mass is detected, the possibility of IPAS should be considered. For the diagnosis of IPAS, the clinical picture, laboratory test and imaging test results must be analyzed together. Tc-99mc-HDRBC scintigraphy is recommended. IPAS must be considered as a differential diagnosis before surgery to avoid unnecessary pancreatic resections suggestive neuroendocrine neoplasia.

Authors Contribution

Conceptualization, G.Z., A.H; formal analysis, G.Z, A.H; data curation, G.Z, J.M.H; writing—original draft preparation, G.Z.; writing-review and editing, A.H. All authors have read and agreed to the published version of the manuscript.

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