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Intrapancreatic accessory spleen in child

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ABSTRACT

Background. Intrapancreatic accessory spleen is a congenital abnormality with duplicated splenic tissue located in ectopic sites.

Case. We report a case of 10-year-old male patient with an infrequent finding of intrapancreatic mass. The examination of complete blood count, biochemistry, tumor markers were within the normal reference ranges. Imaging series found an intrapancreatic mass without wash-out effect on contrast-enhanced ultrasound, MRI-intensity equal to splenic one and no increased glucose metabolism on PET/CT. Follow-up of the patient did not demonstrate progression of the size or change of ultrasound characteristics of the lesion.

Conclusions. Intrapancreatic accessory spleen is an asymptomatic lesion and without the need of surgical therapy. It is important to differentiate it from pancreatic malignant tumors.

Key words: pancreatic tumor, accessory spleen, contrast-enhanced ultrasound, CEUS, PET/CT.

Intrapancreatic accessory spleen (IPAS) is a congenital abnormality with duplicated splenic tissue located in ectopic sites. The most common locations for accessory spleens are the hilum of the spleen, followed by being adjacent to the tail or other parts of the pancreas. The patients usually present with no clinical symptoms.[1,2]

Case Report

A 10-year-old male patient was referred to the Department of Pediatrics with a complaint of constant eructation during the day. There were no complaints when the patient was asleep. He had undergone an appendectomy three weeks earlier. The onset of belching was several days after the operation. Thalassemia minor was established at 1 year of age. An off-patient consultation was done due to eructation and a tumor mass was found in the pancreas.

The physical examination and laboratory data showed no abnormalities. Pancreas enzymes were normal. Tumor markers, including carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA 19-9) were within reference ranges.

Abdominal ultrasound of the pancreas appeared to be normal in size, without any remarkable findings in the parenchyma and ducts. In between the head and body of the pancreas a hypoechoic mass was visualized with dimensions 13 x 8 mm. On Doppler examination no vascularization of the mass was found. (Fig. 1)

The other parenchymal organs remained normal in size and echogenicity. No pathological findings were registered on Doppler examination of the major abdominal vessels. No free fluid was found in the abdominal cavity.

On contrast enhanced ultrasound (CEUS) the lesion appeared to be hypoenchanced during the early dynamic phase (Fig. 2) and isoenchanced

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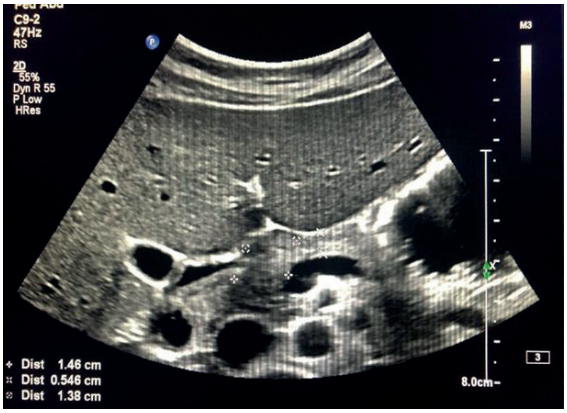

Fig. 1. B-mode ultrasonography examination presenting an intrapancreatic mass, located in the body of the pancreas.

during the late venous phase. No wash-out was observed (Fig. 3).

On gadopentetic-acid-enhanced MRI the lesion appeared round and localized on the ventral surface of the pancreas. The mass demonstrated relative hyperintensity in conventional T2-weighted images and isohypointensity in T1-weighted images. The lesion showed hyperintensity on diffusion-weighted imaging at b=0 and b=500 s/mm^2. After contrast application a formation increased in intensity characteristics equal to splenic parenchyma (Fig 4).

On fludeoxyglucose Positron Emission Tomography / Computed Tomography (18 F-FDG PET/CT) in the pancreas a mass was observed with unremarkable delineated borders and without increased glucose metabolism in the lesion. The standardized uptake value (SUVmax) was measured < 2.0. The conclusion was the absence of PET/CT evidence of malignant lesions. (Fig. 5)

The 6-month follow-up of the patient showed no increase of the lesion in size or change of ultrasound pattern. During work-up for intrapancreatic tumor, the eructation stopped.

Based on clinical and imaging features, our working diagnosis was accessory spleen in pancreas. No invasive treatment was planned. The initial complaint was considered as functional.

This case is an illustration that appropriate use of imaging series can avoid the need of invasive procedures and surgery in patients with intrapancreatic tumors.

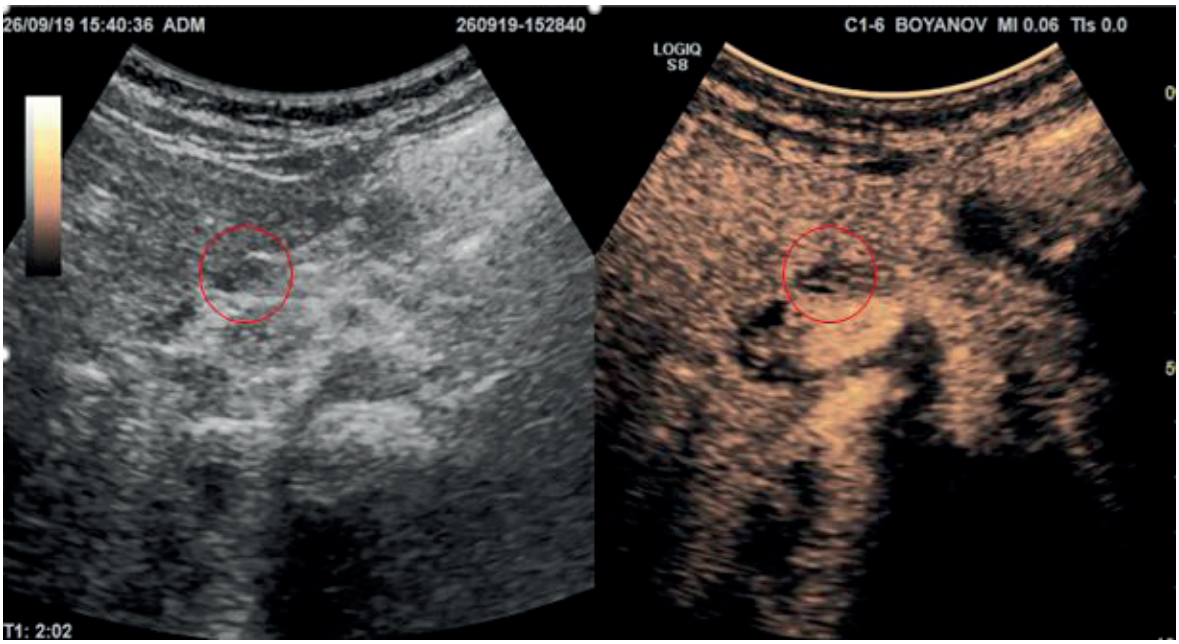

Fig. 2. Arterial phase of CEUS, presenting a hypoenhanced lesion at the site of the intrapancreatic mass.

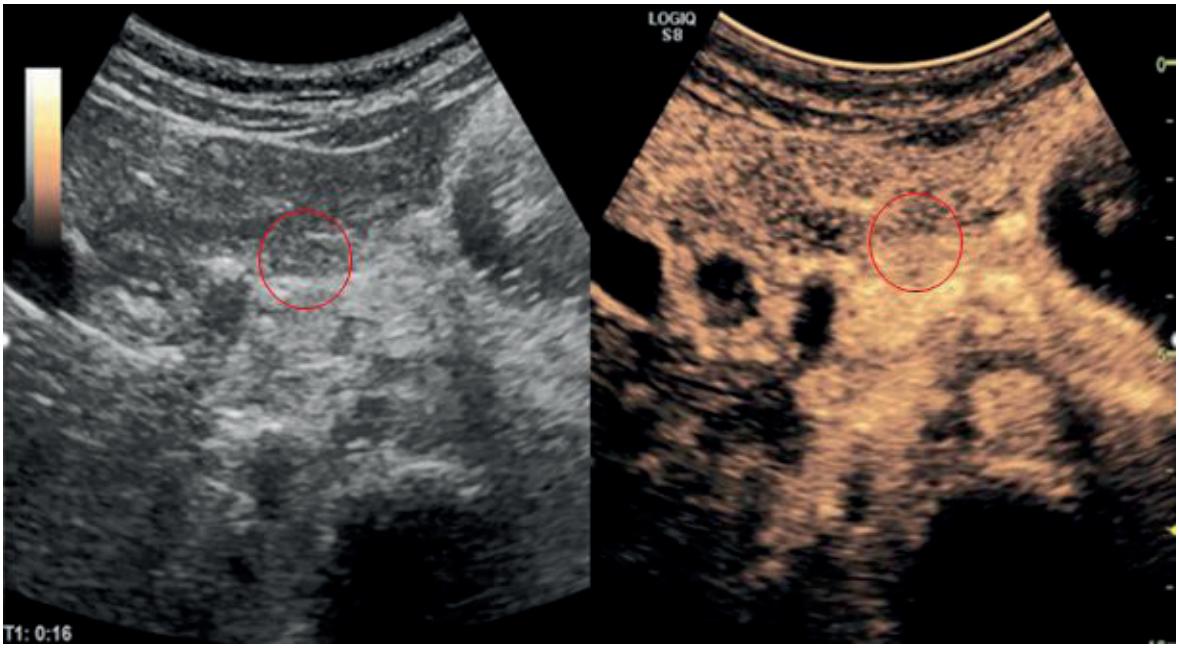

Fig. 3. Venous phase of CEUS presenting that lesion remain isoenhanced to the pancreatic parenchyma.

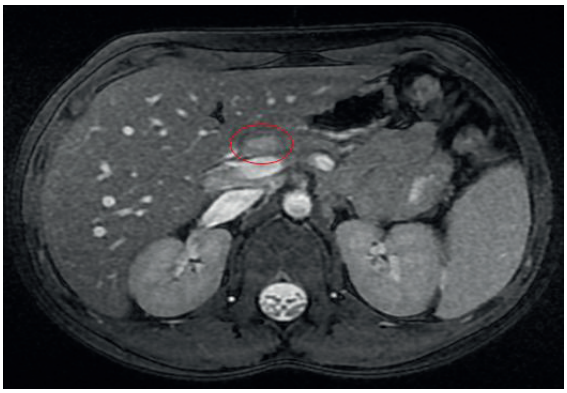

Fig. 4. Contrast-enhanced MRI hyperintense lesion at the site of intrapancreatic mass.

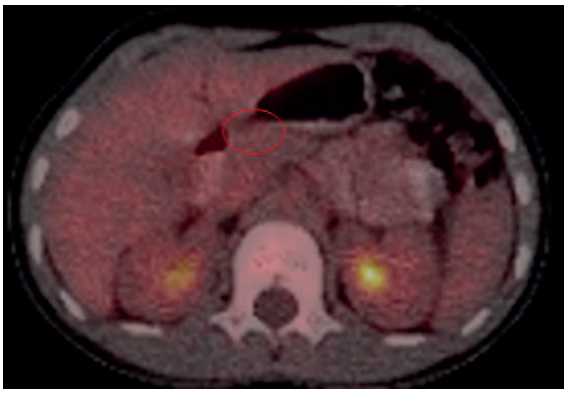

Fig. 5. PET/CT presenting absence of metabolic activity at the site of intrapancreatic mass.

Written informed consent was obtained from the patients'parents.

Discussion

When the solid mass was first discovered we primarily discussed the possibility of a neuroendocrine tumor (NET) with pancreatic localization. Johnson reported 60% inactive tumors in a studied cohort. They are usually large in size and often the metastases appear at diagnosis.[3] Stawarski et al. reported similar results.[4] Symptomatic tumors are most often insulinomas and present with hypoglycemia. Other possible manifestations are redness of the face and neck, tachycardia, dizziness, excessive sweating, diarrhea, bronchospasm, abdominal pain, weight loss, lack of appetite, and vomiting.[3,4] Due to the patient's lack of complaints and signs, a functionally inactive tumor was considered.

The second possibility was a benign mass. Usually, ectopic spleens (up to 80%) are located close to the splenic hilum, and 16% are in the tail of the pancreas. IPAS in other parts of the

pancreas is an occasional finding. In general, IPAS is an asymptomatic lesion and it does not need surgical therapy.[2]

Laboratory diagnosis of NET includes the examination of chromogranin A and chromogranin B.[3] The assay demonstrates high specificity, but low sensitivity and a high probability of interference. In the respective types of symptomatic NET, the level of the respective hormone can be examined, and various functional tests can be performed, as well.[3,4] After consultation with a pediatric endocrinologist, a possible inactive tumor was considered in our patient. During the follow-up, the child showed no complaints.

Sofuni et al. as well as the PAMUS study, concluded that ultrasound examination visualized well tumor formation and hypervascular enhancement pattern. CEUS demonstrated an excellent capability for the differentiation of the solid pancreatic masses.[5,6] D'Onofrio et al. reported that ultrasound techniques allow the differentiation of benign from malignant lesions in the pancreas, as well as solid from cystic lesions. CEUS pattern of NET is described as a rapid, intense enhancement of the lesion resulting in a hyperechoic appearance in the early contrast phases due to hypervascularity of the lesion. In the late phase NET appear hypoechoic due to the rapid wash-out effect. The minority of NET do demonstrate described characteristics.[7,8]

Rinzivillo et al.[9] reported that ^{18}F-FDG PET findings strongly correlate with disease behavior. In patients with unknown disease status, ^{18}F-FDG PET can provide relevant clinical information in patients with a positive examination. Calabrò et al.[10] concluded that ^{18}F-FDG PET is indicated in patients with high-grade NET or in those with the suspicion of rapid progression and for prognosis of the tumor stratification.

During the CEUS examination a hypoenchanced lesion in the early dynamic phase and an isoenchanced in the late phase is a typical US pattern. The IPAS is normally seen in magnetic resonance of hyperintense diffusion weighted in T2 and hypointense weighted in T1 compared to normal pancreatic tissue. The ^{18}F-FDG PET/CT or ^{68}Ga-DOTA-TOC PET/CT has a high specificity to differentiate IPAS from pancreatic adenocarcinoma or neuroendocrine tumors.[11,12]

A small proportion of cases of IPAS may occur secondary to other diseases, including epidermoid cyst and inflammatory pseudotumor.[13-15] In adults, IPAS needs histopathological conformation. In children, tissue material is usually obtained during surgical operations. Our patient demonstrated clinical and imaging presentation of a benign solid intrapancreatic lesion. That's why we accepted a "wait and watch" approach, but fine needle biopsy or laparoscopic examination remain options, in case of growth of the tumor or appearance of hypervascular pattern.

The diagnosis of IPAS may be difficult secondary lesions are present. Imaging studies certainly have an important role in IPAS diagnosis. Ultrasound examination, combined with CEUS, MRI and PET/CT revealed to be highly accurate methods in the diagnosis and differentiation of IPAS and small solid pancreatic tumors.[16-20]

IPAS is an asymptomatic lesion and surgical treatment and histopathological confirmation may not be needed if diagnostic imaging highly suggests IPAS in children. There is significant importance in differentiating IPAS from pancreatic malignant tumors. This diagnosis must be considered before surgery of solid pancreatic masses to avoid unnecessary pancreatic resections.

Author contribution

The authors confirm contribution to the paper as follows: study concept and design: IY; data collection: NB; analysis and interpretation of the data: IY, NB; draft manuscript preparation: IY. All authors reviewed the results and approved the final version of the manuscript.

Conflict of interest

The authors declare that there is no conflict of interest.

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