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Intrapancreatic accessory spleen in newborn with multiple congenital malformations

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ABSTRACT

A new case of intrapancreatic accessory spleen located in the tail of the pancreas in a premature male newborn is reported. The anomaly was unexpectedly found at the autopsy together with many other malformations including heart defects, polycystic kidneys, oxycephaly, facial dysmorphy, low set ears, webbing of neck and accessory fingers of right hand and foot. Microscopic examination of the pancreas revealed accessory spleen composed of red and white pulp. The spleen was partly well-demarcated but partly intermingled with pancreatic lobules. Many goblet cell-containing intralobular pancreatic ducts, some of them entrapped within spleen were noted. Splenopancreatic field abnormalities are known to be highly specific for trisomy 13 syndrome. Although karyotyping was not performed, on the basis of reveled malformations the syndrome can be suspected in the presented case.

Keywords: accessory spleen, anatomical variation, congenital malformation, trisomy 13 syndrome

INTRODUCTION

Accessory spleens arise form the failure of fusion of the splenic anlage located in the dorsal mesogastrium during the 5th week of gestation [1, 7]. In autopsy studies pancreas is relatively common location of accessory spleens, but in the computed tomography (CT) examination, it is affected only in 1.3% of patients with accessory organs [8]. Majority adults and some children cases of intrapancreatic spleens are sporadic findings [11]. However, many reports proved connection of that anomaly together with fusion of pancreatic tails with splenic hilum and pancreatic dysplasia with trisomy 13 syndrome as well as other congenital abnormalities, e.g., dysmorphy, osteocraniostenosis, prune belly syndrome or heart defects [3, 5, 9].

In the current paper a new case of intrapancreatic accessory spleen is presented. This is a second case of that malformation found at the Department of Clinical Pathomorphology, Medical University of Lublin during short period of time but with different clinical and morphological setting [11].

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MATERIALS AND METHODS

Of 84 autopsies performed in the period January-November 2012 in the Department of Clinical Pathomorphology, Medical University of Lublin, one new case of intrapancreatic accessory spleen was revealed. The clinical data on the patient was obtained from the medical history but postmortem findings from autopsy protocol. Microscopic examination of all samples taken during autopsy was routinely performed on the basis of hematoxylin and eosin staining using Olympus BX45 (Japan). For the pancreas alcian blue+PAS (pH 2,5) stain was additionally done. The photographs were taken by DP12 camera. The PubMed MEDLINE database was used for searching reports on intrapancreatic accessory spleen.

RESULTS

A male premature newborn was delivered at the 35-week of gestation by a 31-year-old woman (G2P2A0). Intrauterine growth retardation (IUGR) was suspected at pregnancy. Due to absence of respiratory effort after delivery and relatively low 1-minute Apgar score – 4, he was admitted to the Department of Anesthesiology and Intensive Care, Children's Clinical Hospital, Lublin. The child

died a few hours later. Congenital pneumonia, sepsis and bilateral pneumothorax were diagnosed. Furthermore, multiple congenital malformations including heart defects (common arterial trunk, atrial and ventricular septal defects), oxycephaly, facial dysmorphy, low set ears, webbing of neck and accessory fingers of right hand and foot were also revealed.

An autopsy of the newborn (weight 2362 g; length 46 cm) was performed. The direct cause of death was respiratory and circulatory failure due to the congenital interstitial pneumonia with hyaline membranes formations and focal intraalveolar hemorrhages in child with severe complex heart defect. Clinically found congenital malformations were confirmed. Polycystic kidney disease was also noted. Furthermore, microscopic examination unexpectedly revealed accessory spleen (dimension 6x3 mm) located in the tail of the pancreas (Fig. 1). It was partly well-demarcated but partly intermingled with pancreatic lobules. The spleen was typically composed of red and white pulps (Fig. 2). Many intralobular pancreatic ducts lined by one layer of the cuboidal epithelium and goblet cells, some of them entrapped in the spleen were also observed (Fig. 3). The lumen of ducts and goblet cells were filled with alcian blue-positive mucus.

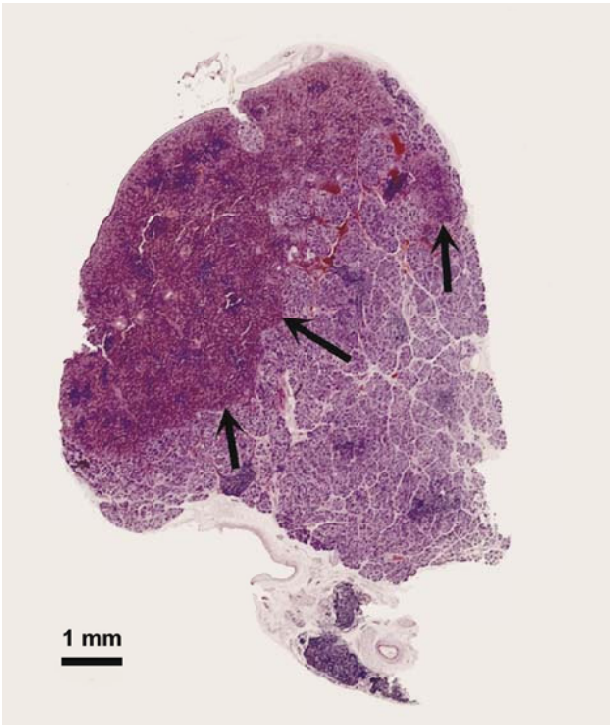

Fig. 1. Intrapancreatic accessory spleen (arrows) (hematoxylin &eosin)

Complex splenopancreatic field abnormalities are known to be highly specific for trisomy 13 syndrome. Although karyotyping was not performed, on the basis of reveled malformations the syndrome can be suspected in the presented case.

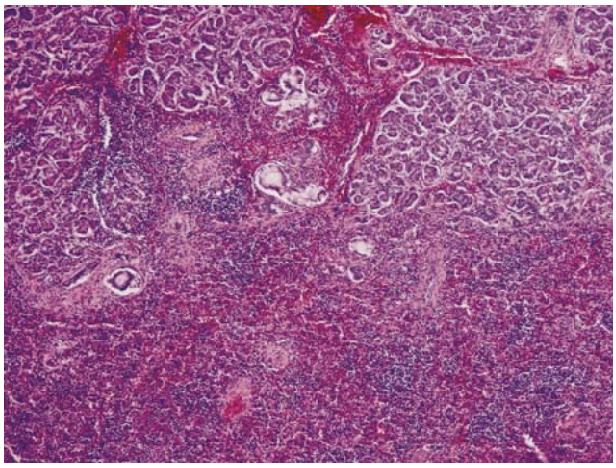

Fig. 2. Poorly-demarcated intrapancreatic accessory spleen composed of red and white pulp (hematoxylin&eosin; objective magn. 5x)

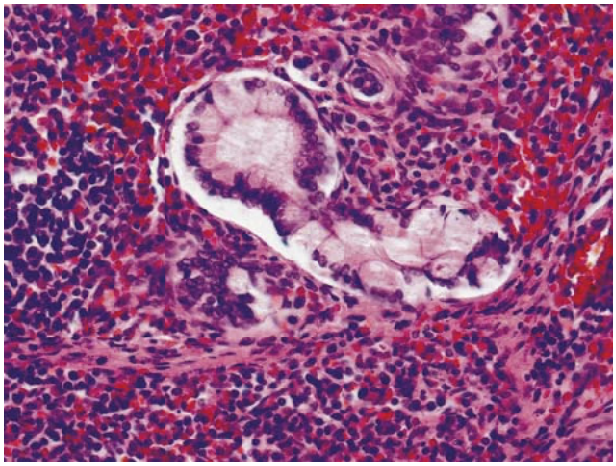

Fig. 3. Intralobular pancreatic ducts lined by columnar and goblet cells entrapped within accessory spleen (hematoxylin &eosin; objective magn. 20x)

DISCUSSION

The majority of intrapancreatic accessory spleens is asymptomatic and poses no clinical problem [7]. In newborns (including stillbirths) and infants with other severe congenital malformations they are just additional finding which does not influence general conditions but can be regarded as potential indicator of trisomy 13 syndrome, especially when accompanied by some microscopic findings like intralobular ducts with goblet cells, microcysts and ductulo-insular complexes, which are almost unique for the syndrome [3].

In most adult patients the lesions are incidental findings detected during otherwise unrelated diagnostic evaluation. However, in some cases it may mimic hypervascular pancreatic mass like an islet cell tumor, a pseudopapillary neoplasm or metastases [5]. It seems that due more frequent usage of modern imaging techniques it will be increasing problem in the near future. To avoid unnecessary surgery accurate preoperative diagnosis is important.

The intrapancreatic accessory spleen can be diagnosed by non-invasive imaging modalities like ultrasonography (US), contrast-enhanced US, computed tomography (CT) or magnetic resonance (MR). On gray-scale baseline US, the lesion is well-defined, round, ovoid or lobulated. The echogenicity is low compared with pancreatic parenchyma and identical to that of the main spleen. In contrast enhanced US using Levovist®, the intrapancreatic spleen appeared to be hyperechoic during all dynamic sonography phases. The echo enhancement of all intrapancreatic lesions, however, is identical to that of the spleen in all phases [7, 10]. The CT usually shows a spherical mass in or adjacent to the tail of the pancreas that is isodense to the spleen and brighter than the pancreas on arterial, pancreas and portal venous phases [4, 7]. The MR, fat-suppresed T1- and T2-weighted images usually shows round lesion with signal identical to spleen, and comparing to signal intensity of the surrounding pancreatic parenchyma, it is darker on the T1- weighted images and brighter than that of the pancreas on the T2-weighted images [7]. But some of the pancreatic tumors share signal intensity pattern with that of the spleen. In these cases dynamic gadolinium (Gd)-enhanced MR may be helpful as it shows characteristic heterogenous enhancement of intrapancreatic spleen similar to that of the adjacent normal spleen.

In most cases the CT and MR findings reveal similarity of intrapancreatic spleen to adjacent spleen in terms of density, attenuation values and signal characteristic. It is so because in majority of cases morphology of accessory and regular organs is identical [11, 12]. However, occasionally accessory spleen can have different white-to-red pulp ratio that may give higher signal intensity on T2-weighted imaging. In these cases, to exclude hypervascular pancreatic neoplasm, additional cytological examination, like endoscopic ultrasound-guided fine needle aspiration (EUS-FNA), should be performed. It revealed small lymphocytes with a subset of histiocytes, eosinophils, and plasma cells. An additional immunostaining for CD8 can confirm presence of sinus endothelial cells [10]. Non-invasive radiographic imaging accompanied by cytological examination allows to established definitive diagnosis and obviate unnecessary surgery.

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