VIA MEDICA

CASE REPORT

Folia Morphol.
Vol. 71, No. 1, pp. 45–47

ISSN 0015–5659
www.fm.viamedica.pl

Intrapancreatic accessory spleen

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[Received 23 September 2011; Accepted 1 December 2011]

A case of accessory spleen located in the tail of the pancreas in a stillbirth male foetus is reported. The congenital anomaly was revealed at autopsy. The intrapancreatic spleen was well demarcated and was composed of red and white pulp; however, same pancreatic ducts were intermingled with the splenic parenchyma. As well as the intrapancreatic lesion another minute accessory spleen was also found at the hilum of the proper organ. Since a lack of morphological features of trisomy 13 syndrome were found in the foetus, the ectopic spleens were regarded as incidental findings. (Folia Morphol 2012; 71, 1: 45–47)

Key words: accessory spleen, anatomical variation, congenital malformation

INTRODUCTION

Accessory spleens are a relatively common congenital abnormalities, and they are found in 10–30% of non-selected autopsies [3]. Moreover, the contrast-enhanced abdominal computed tomography (CT) examination revealed that the anomaly is visible in about 16% of consecutive patients [12]. Its prevalence is much higher in children who have undergone splenectomy — 45–65% [1]. At autopsy most accessory organs (80%) are located at or close to the hilum of the normal spleen [3, 12]. The second most common site is the tail of the pancreas [3]. However, in the CT examination intrapancreatic location was rare — seen in about 1.3% of patients with accessory organs [12]. In the current paper a new case of that anomaly is presented.

CASE REPORT

A male stillborn was delivered at the 39th week of gestation by a 34-year-old woman (G4P4A0) at the Gynaecological and Obstetric Ward of Lublin' District General Hospital in February 2011. An autopsy of the foetus (weight 3030 g; length 510 mm) was performed at the Department of Clinical Pathomorphology of the Medical University of Lublin. The features of intrauterine asphyxia with minute ecchymoses of the pericardial sac, focal pulmonary intraalveolar haemorrhages, and amniotic fluid aspiration were revealed. Major congenital malformations were not found. The intrauterine death was most likely associated with the pathology of the placenta and the umbilical cord; however, it was not received for the gross and histological examination.

Upon dissection of the abdominal cavity, a small, 6 mm accessory spleen located at the hilum of the main proper organ was seen. Furthermore, at the longitudinal section of the pancreas a minute dark-red area in the tail was unexpectedly noted arousing suspicion of local haemorrhage. However, the microscopic examination showed another — an intrapancreatic accessory spleen (Fig. 1). The lesion was well circumscribed but unencapsulated. It was composed of the typical red and white pulp (Fig. 2). A few pancreatic ducts lined by one layer of the cuboidal epithelium and entrapped in the spleen were also observed.

To highlight some details of the morphology of the pancreas a set of immunohistochemical reactions were performed on formalin-fixed paraffin-embed-

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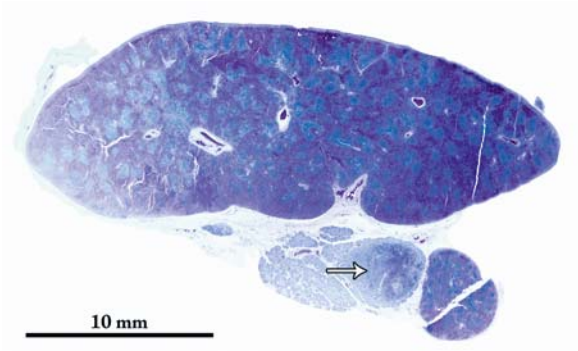

Figure 1. Normal spleen as well as hilar and intrapancreatic (arrow) accessory spleens (haematoxylin & eosin).

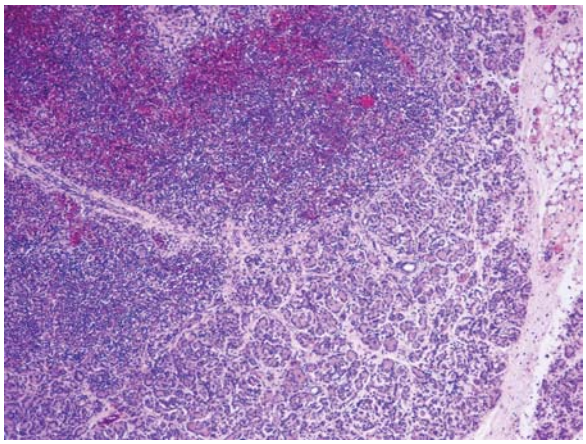

Figure 2. Well-demarcated intrapancreatic accessory spleen composed of red and white pulp surrounding a few pancreatic ducts (haematoxylin & eosin; objective magn. 5×).

ded sections. Monoclonal mouse anti-human antibodies against epithelial cells — pancytokeratin (clone MNF116; dilution 1:100) as well as against lymphocytes — leukocyte common antigen CD45 (clones 2B11+PD7/26; dilution 1:100), B cells — CD20 (clone L-26; 1:100), and T cells — CD3 (clone F7, 2.38; dilution 1:50) were applied. The incubation with antibodies was performed for 0.5 hours at normal temperature. According to the manufacturer's directions the main reactions were preceded by antigen retrieval procedures, i.e. for pancytokeratin — pretreatment with proteinase K (20 μL/mL) for 4 min, for CD45 incubation with 0.01 M citrate buffer (pH 6.0) in microwave oven (3 × 5 min), and for CD20 and CD3 incubation in water bath (temp 95°C) with target retrieval solution (pH 9.0) for 20 min. EnVision™+/HRP visualisation system was used for 0.5 hours at normal temperature and then 3,3'-diamonobenzidine (DAB) as a chromogen was applied. All reagents were from Dako, Denmark.

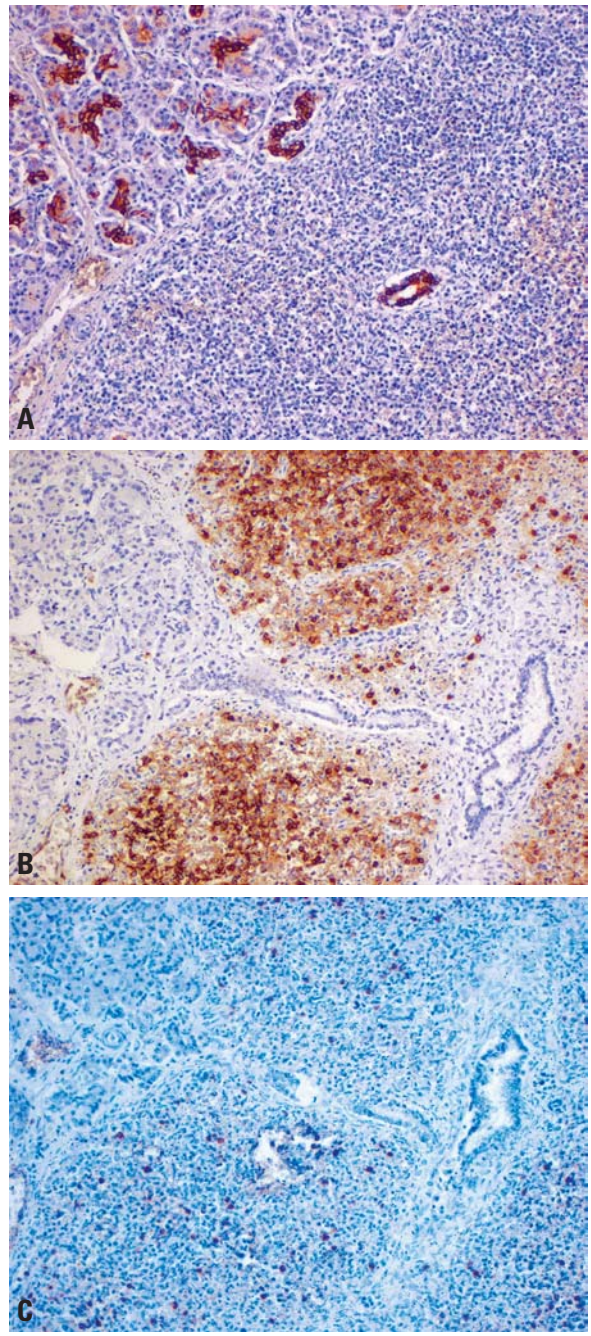

Figure 3. Positive immunostaining for (**A**) pancytokeratin in epithelial cells of pancreatic acini and ducts, as well as for (**B**) CD20 in B — and for (**C**) CD3 in T cells of white pulp of intrapancreatic spleen (DakoEnVision™+/HRP; objective magn. 10×).

Positive immunostaining for cytokeratin was seen in epithelial cells of pancreatic acini and ducts (Fig. 3A). Lymphocytes forming white pulp of the accessory intrapancreatic spleen exhibited a strong positive reaction for CD45. They were predominantly CD20-positive B cells (Fig. 3B), whereas CD3-positive T cells were rare (Fig. 3C).

DISCUSSION

The accessory spleens originated most likely from hillocks of mesenchymal cells in the dorsal mesogastrium, which migrated between leaves of the mesentery and did not fuse to create the main spleen at the 5th week of gestation. Nests of those cells may be entrapped by the pancreatic tail, which, at the same time, derived from the mass of cells situated caudally to the splenic hillocks [4].

Some childhood (including that presented here) and probably most adult cases of the intrapancreatic spleen seem to be sporadic lesions without genetic predisposition. However, splenopancreatic field abnormalities are typical for the trisomy 13 syndrome (Patau syndrome) [2, 5]. The incidence of accessory spleens among individuals with such chromosomal abnormality is much higher than in the normal population, reaching 30–60% [5]. In the large series by Gomi et al. [2] the classification of gross splenopancreatic abnormalities, intrapancreatic accessory spleens, and/or fusion between the pancreatic tail and the spleen/accessory spleen was found in all children with trisomy 13 (n = 21). Conversley, those anomalies were noted just in seven cases of non-trisomic controls (n = 1060). Furthermore, pancreatic dysplasia, including intralobular ducts with goblet cells, microcysts, and ductuloinsular complexes, are typical of the trisomy [5]. Splenopancreatic field abnormalities were also reported in new-borns with an abnormal karyotype (47, XX + 21) and dysmorphic features, osteocraniostenosis, and prune belly syndrome or with congenital heart malformations [13].

Since intrapancreatic spleens are usually asymptomatic, they are regarded as lesions without special clinical significance. Occasionally, they can be a source of secondary pathological lesions like the epidermoid cyst [7, 8], serous cystic neoplasm [6], or littoral cell angioma [14]. Nevertheless, the most important clinical problem is their incorrect diagnosis as a true pancreatic neoplasm, usually a non-secreting endocrine tumour [10, 11]. Appropriate diagnosis is a crucial for further treatment. The intrapancreatic spleen does not require any treatment, but for true tumours surgical resection is recommended. Radionuclide imaging with ^{99m}Tc-sulfur colloid or ^{99m}Tc-tagged heat-damage erythrocytes as well as superparamagnetic ion oxide (SPIO)-enhanced magnetic resonance imaging or intravenously enhanced ultrasonography are non-invasive and sensitive methods of visualisation of the splenic parenchyma [9, 10]. Endoscopic ultrasound-guided fine-needle aspiration biopsy can also be helpful in confirmation of the diagnosis [11]. Large aggregates of CD8-positive spindle cells found in smears seem to be typical of intrapancreatic accessory spleens complicated by epidermoid cyst [8].

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