Kopystecka Agnieszka, Lipinska Justyna, Kopec Izabela, Lipinski Lukasz, Wozniak Magdalena Maria, Jedrzejewski Grzegorz. Polysplenia and other anatomical variants of the spleen. Journal of Education, Health and Sport. 2022;12(3):128-138. eISSN 2391-8306. DOI http://dx.doi.org/10.12775/JEHS.2022.12.03.011
https://apcz.umk.pl/JEHS/article/view/JEHS.2022.12.03.011
https://zenodo.org/record/6366304

The journal has had 40 points in Ministry of Education and Science of Poland parametric evaluation. Annex to the announcement of the Minister of Education and Science of December 21, 2021. No. 32343.
Has a Journal's Unique Identifier: 201159. Scientific disciplines assigned: Physical Culture Sciences (Field of Medical sciences and health sciences); Health Sciences (Field of Medical Sciences and Health Sciences).

Punkty Ministerialne z 2019 - aktualny rok 40 punktów. Załącznik do komunikatu Ministra Edukacji i Nauki z dnia 21 grudnia 2021 r. Lp. 32343. Posiada Unikatowy Identyfikator Czasopisma: 201159.
Przypisane dyscypliny naukowe:Nauki o kulturze fizycznej (Dziedzina nauk medycznych i nauk o zdrowiu); Nauki o zdrowiu (Dziedzina nauk medycznych i nauk o zdrowiu).

Received: 01.03.2022. Revised: 16.03.2022. Accepted: 17.03.2022.

Polysplenia and other anatomical variants of the spleen

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Abstract

The anatomy, physiology and embryology of the spleen are essential fields of study for the determination of congenital varieties as well as the pathological processes occurring in this organ.The aim of this study is to summarize the current knowledge on the proper development of the spleen and to present structural variants that may be of clinical significance or be important from the radiologist's point of view.Structural changes of the spleen may result in non-specific clinical symptoms, which may confuse an inexperienced physician. Inaccurate diagnosis and late diagnosis have various negative, often even fatal, consequences. In order to make an appropriate diagnosis, and thus help the patient in a timely manner, it is necessary to have extensive knowledge about the variants of the structure of the spleen. Knowledge of anatomical variations is essential for the development of a differential diagnosis, which enables the correct diagnosis to be made.

Key words: asplenia; polysplenia; accessory spleen; ectopic spleen

Introduction

The anatomy, physiology and embryology of the spleen are essential fields of study for the determination of congenital varieties as well as the pathological processes occurring in this organ. Although many congenital lesions in the spleen are clinically significant, most of them are asymptomatic and are diagnosed accidentally after trauma or during non-invasive medical procedures. Significant modalities used to diagnose these anomalies are ultrasonography (US) and computed tomography (CT) (1). Congenital splenic variations include: asplenia, polysplenism, lobulation of spleen, accessory spleen, ectopic spleen, accessory spleen nodules, and organ heterotaxy (2). The knowledge of the characteristic features of anomalies in the structure of the spleen allows to distinguish them from abdominal pathological lesions (3).

Aim

The aim of this study is to summarize the current knowledge on the proper development of the spleen and to present structural variants that may be of clinical significance or be important from the radiologist's point of view. The gathered knowledge will facilitate the correct interpretation of imaging modalities results and will help to avoid misdiagnosis with clinically significant abnormalities, such as abdominal neoplastic diseases.

Method

A review of the scientific literature and publications of the PubMed database for the years: 2010-2020. Using medical records, we describe the essence of selected structural varieties of spleen and possible complications resulting from these anomalies.

Results

1. Anatomy of spleen

Spleen is the largest lymphatic organ in human body. This organ is the site for the removal of defective or "old" erythrocytes, lymphocytes and thrombocytes, and it is a reservoir of blood. Normally spleen is oval in shape and is located in the abdominal cavity, intraperitoneally under the left dome of the diaphragm, touching the ninth, tenth and eleventh ribs. In the supine position, its long axis corresponds to that of the tenth rib to the left (from behind and top (posterior pole) to the front and the bottom (anterior pole).). In healthy adults, the longitudinal dimension of the spleen ranges from 7 to 14 centimeters, and its circumference is on average 11 centimeters. Depending on the contents of the blood, the spleen weighs from about 150 to 200 grams (2). The size and weight of the spleen may vary by gender and race. Males, on average, have a larger spleen than females, and white people tend to have a larger spleen compared to African Americans. Usually, the spleen is not palpable on physical exam (4).

2. Embryogenesis of spleen

Spleen bud appears around the fifth week as a thickening of the mesenchymal cells inside the dorsal mesogastrium. The proliferating cells form the fibrous capsule, trabeculae and the reticular stroma of the spleen. (5). Initially, the bud, lying in the median plane, moves towards the left side of the abdominal cavity as the stomach rotates between the sixth and seventh week. The spleen continues to grow and connects to the posterior abdominal wall through the splenorenal ligament that connects the spleen to the left kidney. Then the spleen is fixed in the right place by subsequent ligaments that connect: the cavity with the stomach (gastrosplenic ligament), the spleen with the diaphragm (phrenicosplenic ligament) and with the pancreas (splenopancreatic ligament) (2). In utero, the spleen has a lobular structure, which later disappears and the pulp is differentiated into white and red pulp. From the fourth month, hematopoiesis begins, which lasts until about the eighth month, and the T and B lymphocytes penetrate into the organ (5). Spleen enlargement in prenatal life may occur in the case of congenital infections through the placenta, hematological diseases, immune disorders and lipidosis, while hypoplasia is observed in the case of Di George's syndrome and sickle cell anemia (6) .

3. Asplenia and hyposplenia

Asplenia is the term for the absence of a spleen. This may refer to the anatomical deficiency of this organ or functional asplenia secondary to the disease process - hyposplenism (4). The causes of asplenia and hyposplenismmay be associated with various diseases of hematologic, oncologic, gastrointestinal, rheumatologic, immunologic and congenital origins (7).This anomaly may be associated with decreased immunity, and functional assessment of the spleen is rarely performed in immunodeficient patients (7). The risk of severe infection complicated by sepsis in anyone with asplenia is estimated to be around 5%, but may vary with age and is higher in children. Mortality, on the other hand, is estimated at 50%. Asplenic patients have a 200-fold higher risk of dying from sepsis

compared to those with a normally functioning spleen (4). Enveloped bacteria such as Streptococcus pneumoniae (pneumococci), Neisseria meningitidis (meningococci) and Haemophillusinfluenzae type b (Hib) pose the greatest risk (8). Methods of preventing infection in such patients include education, vaccination (especially against enveloped bacteria) and antibiotic prophylaxis (7).

4. Polysplenia

The polysplenia is a multi-system congenital defect characterized by multiple (2-16 or more) accessory spleens(Fig 1) (2). The spleens are usually of a similar size, 1-2 larger spleens and a few smaller ones are less common (1). The synonym for polysplenia is left-sided isomerism, this defect belongs to the heteroataxy syndrome (9). The syndrome is characterized by an incorrect position of organs in the thorax and the abdominal cavity and is the result of abnormalities in embryogenesis. No single anomaly is pathognomonic, however, the presence of multiple spleens, most commonly associated with the stomach, is quite characteristic of heteroataxia (10). Contrary to asplenia, polysplenia is predisposed to the female sex. (9). The exact etiology of this defect is unknown, however, recent studies has identified genes such as CFC1, SHROOM3 and others, which occurs in people with heteroataxia syndrome (1).

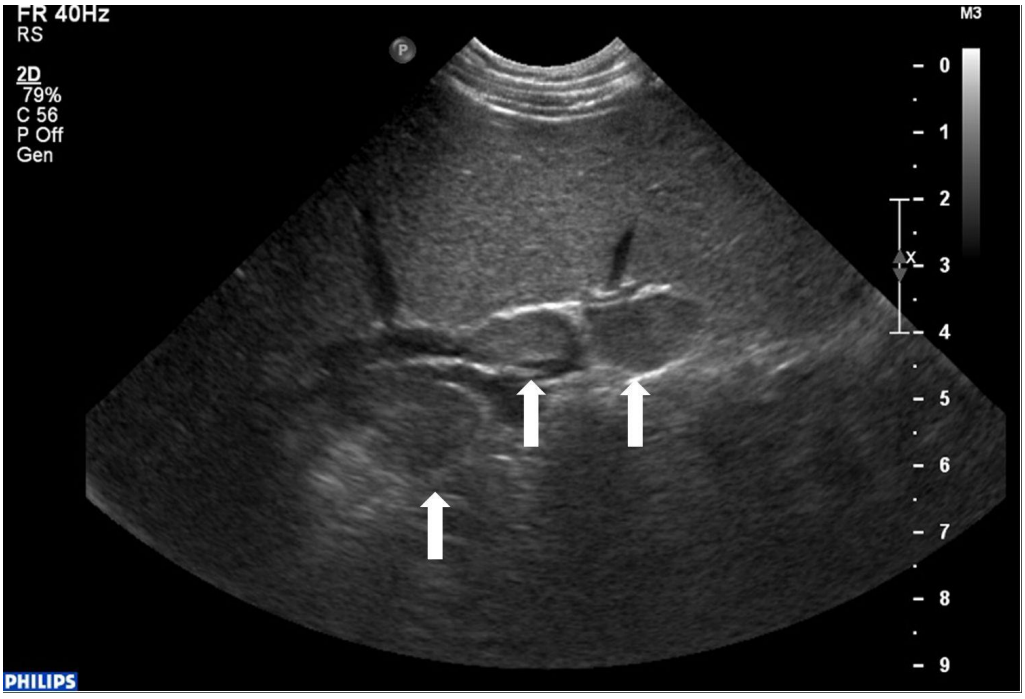

Fig. 1. Polysplenia. Ultrasound scan of the spleen, with three accessory spleens in the hilar area (arrows).

5. Lobulation of the spleen

Lobulation of the spleen is a normal variation of spleen shape, and it is not associated with any pathological conditions (2). The fetal spleen is lobulated, the lobules normally disappear before birth. Lobulation of the spleen may persist into adulthood and usually are seen along the medial part of the spleen (3). Deep clefts and notches, which are sometimes

called “crenaesplenis” may reach 2–3 cm in depth, and are located mostly on the diaphragmatic surface and, especially on the superior border (formerly known as the crenate margin) (Fig 2). Occasionally these lobulations may be misinterpreted as splenic lacerations in patients with abdominal trauma. (2). In some cases, the persistent lobule of splenic tissue extends medially anterior to the upper pole of the left kidney and, less often, posterior to the upper pole of the left kidney, displacing it anteriorly (3).

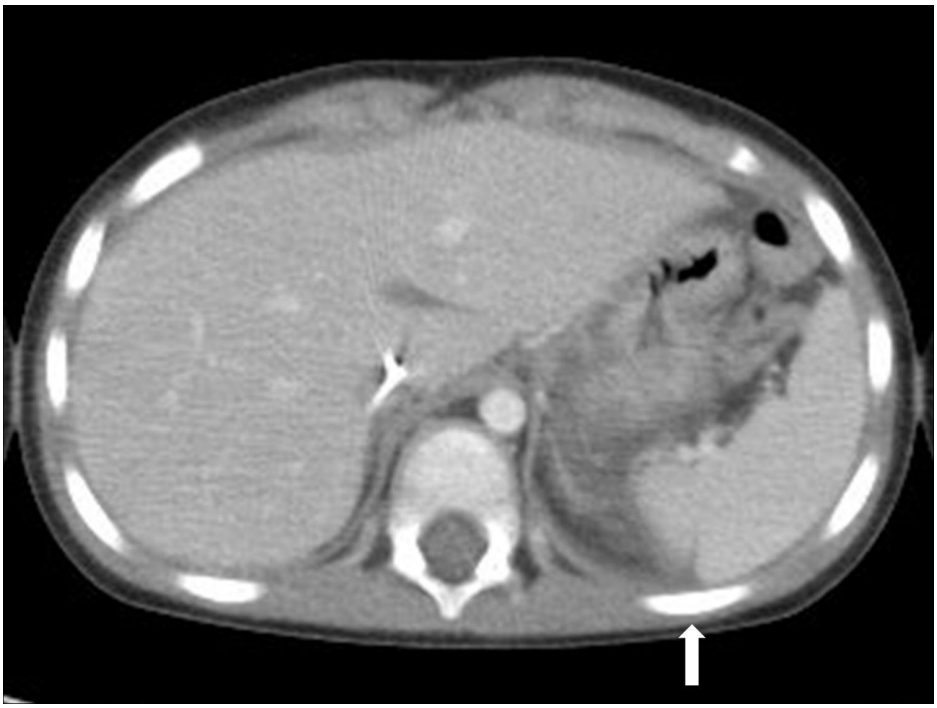

Fig. 2. Lobulation of the spleen. CT scan of the abdomen, with fissureon the diaphragmatic surface of the spleen,near the rear pole (arrow).

6. Accessory spleen

Accessory spleen, also called supernumerary spleen or splenules, is benign and asymptomatic condition where the spleen tissue is outside the normal spleen (Fig 3). Splenules are a relatively common phenomenon with an estimated 10% to 30% of the population having one (11). Accessory spleens are morphologically and functionally equivalent to normal spleens (2). The location is very diverse, but the most common locations are the hilum and vascular pedicle of the spleen, around the tail of the pancreas, greater omentum, along the greater curvature of the stomach, mesentery of the small and the large intestine, near the left ovary or left testis and in the pouch of Douglas (12). Typically, a person can have one to six accessory splenic buds. In addition, the size of the accessory spleen can also be different. (11). The accessory spleen is usually a round or oval structure between 1 cm and 3 cm in diameter, with most studies reporting a diameter of about 1.5 cm. Patients after trauma can present with a very large accessory spleen, reaching up to 10 cm. A smaller accessory spleen can be misdiagnosed as an enlarged lymph node or tumor (13). TerminologiaEmbryologica distinguishes accessory spleens based on their localization in organs, including the adrenal gland, pancreas, stomach, and intestine. In the current classification scheme, retroperitoneal and pelvic accessory spleens are missing (2). The

accessory spleen can simulate a tumor in the adrenal gland, pancreas, stomach, or intestine (3). Identification of the accessory spleen is especially important for patients with immune thrombocytopenic purpura (ITP), who require splenectomy, because an unrecognized accessory spleen can later develop symptoms that are resistant to treatment. The accessory spleen also can be the source of significant intraabdominal haemorrhage (13).

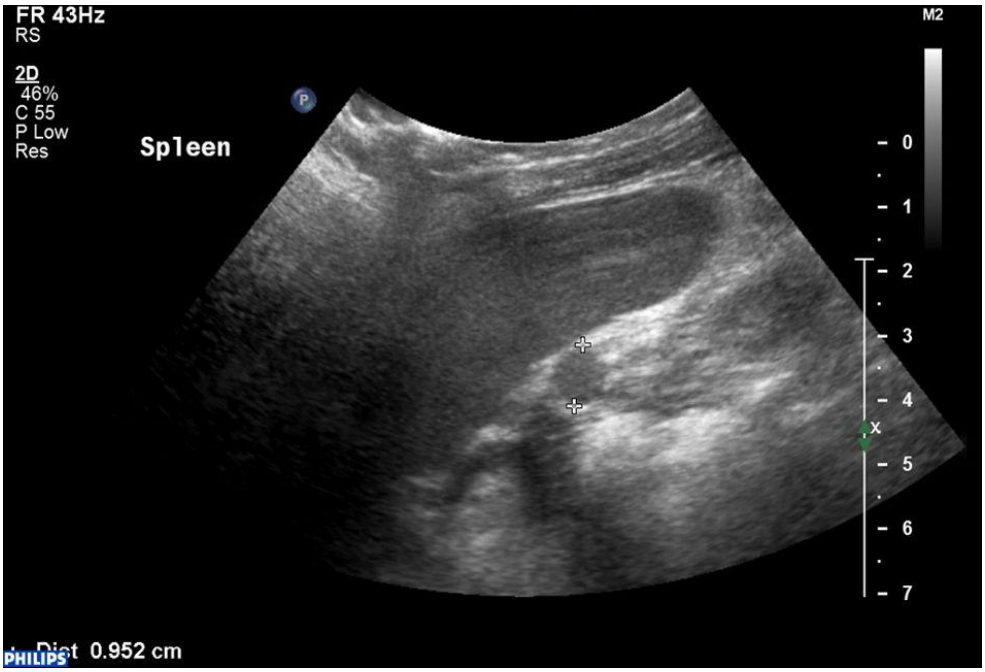

Fig. 3. Ultrasound scan of the accessory spleen (calipers).

7. Ectopic spleen

The migratory or ectopic spleen refers to the migration of the spleen from its normal location in the upper left quadrant to a more caudal location in the abdominal cavity due to the relaxation or abnormal development of the splenic support ligaments (gastro-splenic ligament and splenic ligament) (3). The ectopic spleen is often found in the pelvis (Fig 4), although it can sometimes remain in the upper left quadrant and simply have an abnormal rotation (14). It is a rare entity that is found by chance in less than 0.2% of splenectomy patients. It is usually detected between the ages of 20 and 40 and is more common in women (3). At reproductive age, the migratory spleen is diagnosed about 10 times more often in women, suggesting that the action of female sex hormones affects the flexibility of the ligaments associated with the spleen (2). Children account for a third of all cases, boys and girls under the age of 10 are equally affected (3). Ligament laxity can be acquired due to conditions such as spleen enlargement or pregnancy, but it is often congenital. Due to this laxity, there is an elongated vascular stalk that is prone to torsion and the resulting spleen infarction (14). Despite the fact that a migratory spleen carries an increased risk of vascular torsion (acute, intermittent or chronic torsion) or rupture following injury, it is assumed that up to 50% of all cases go undiagnosed (2).

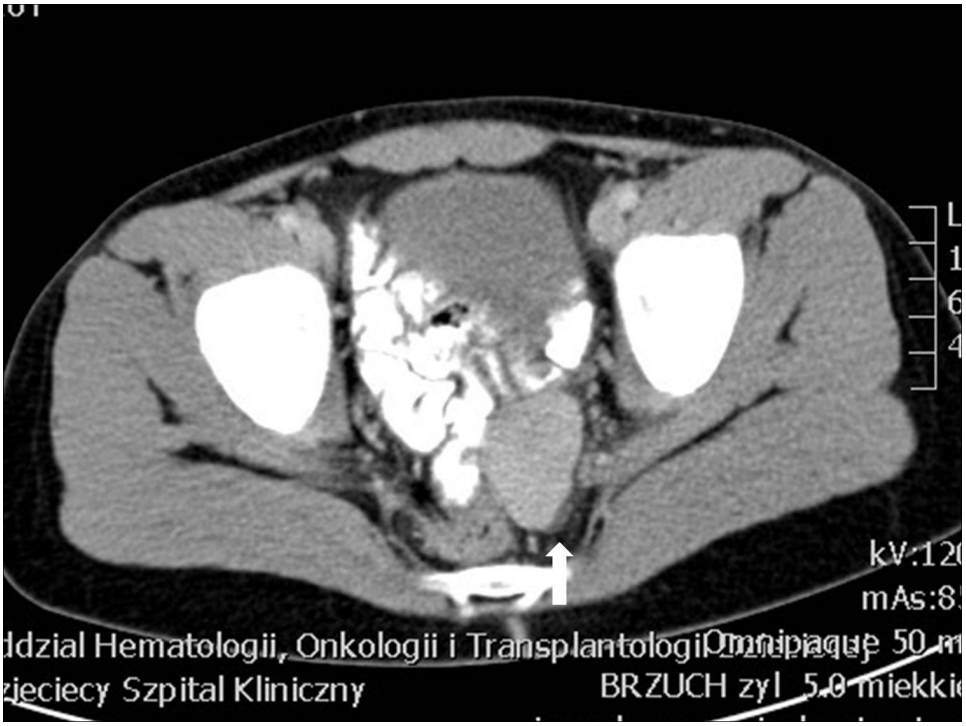

Fig. 4. The ectopic spleen. CT scan of the pelvis with the spleen tissue on the front of the sacral bone (arrow).

8. Splenogonadal fusion

Splenogonadal fusion (SGF) is a rare congenital anomaly defined as the presence of ectopic spleen tissue caused by the abnormal connection of the spleen to the gonad or mesonephric derivatives during the embryonic period. The first such case was described in 1883 by Farthouat et al. The age of the patients is less than 10 years in half of the described cases, and 82% of the cases concern young men under 30 years of age. SGF is a problem in the differential diagnosis of testicular tumors, which can lead to unnecessary orchiectomy. SGF usually affects the left gonad between 5 and 6 weeks of gestation (98% of cases) before the onset of gonadal descent (15). Fewer than 200 cases of SGF have been documented in the literature. This anomaly is often associated with other birth defects such as inguinal hernias and cryptorchidism, therefore clinicians should also be alert to any other associated birth defects in such patients. The fusion of the spleen and gonads is usually an asymptomatic mass of the testicles, but it can cause symptoms such as severe pain in the testicles, swelling due to trauma, and symptoms of ectopic infection of the spleen tissue.

A classification of SGF into continuous or discontinuous forms has been proposed. A continuous form is defined by an uninterrupted cord connecting the spleen and gonad. The discontinuous form is defined as the mesonephric splenogonadal structure that has lost connection with the primary spleen and is considered the accessory spleen variant, usually presented as a hard scrotal nodule imitating a testicular tumor, often asymptomatic (16). The continuous form accounts for approximately 55% of all reported cases and is associated with a five times the risk of congenital malformations compared to the discontinuous form (17).

The clinical picture of SGF is nonspecific and the diagnosis is often made by chance during surgery for inguinal hernia and / or cryptorchidism or by histological examination after orchiectomy for a tumor. Its association with germ cell tumors has been described in several cases, with particular emphasis on the fact that these patients had ipsilateral cryptorchidism.

9. Splenopancreatic fusion

Splenopancreatic fusion is a infrequently observed abnormality, it is in most cases associated with trisomy of chromosome 13, Schinzel-Giedion syndrome, trisomy 21 or other severe congenital abnormalities, usually it is the fusion of the spleen with the tail of the pancreas (2). Sometimes the accessory spleen may also be fused with the pancreas, or the spleen tissue may be found inside the pancreas (18).

10. Situs inversus and heterotaxy

Situs inversustotalis refers to an exact mirror-image of the usual organ arrangment, while heterotaxy is an abnormality in which the internal organs of the thoracic and abdominal cavity exhibit a misalignment across the left-right body axis. Heterotaxia is generally classified into two main syndromes, polysplenia and asplenia syndromes (1). Most patients with polisplenicheterotaxy patients have multiple spleens of varying size and number, which may be found on the left or right side of the abdominal cavity (19) (Fig 5).

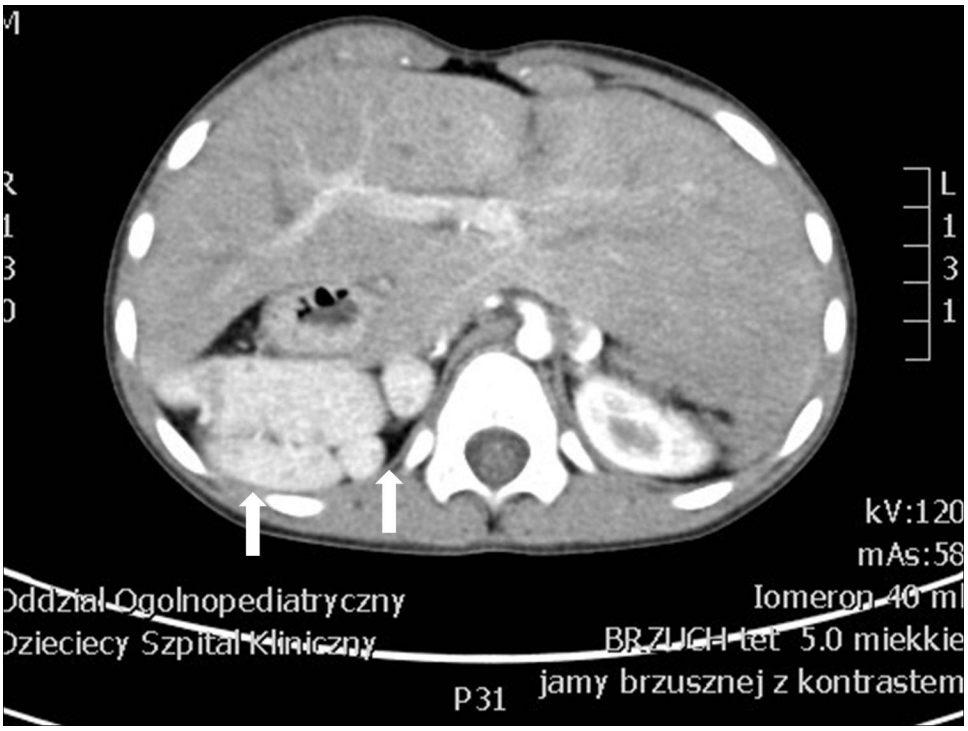

Fig. 5. Polisplenicheterotaxy. CT scan of the abdomen with the spleen and multiple accessory spleen on the right side of the abdomen (arrows).

11. The giant accessory spleen

The giant accessory spleen is a type of accessory spleen, which is characterized by a much larger size (Fig 6), and thus - it can give symptoms of pressure, gastrointestinal tract strain or

pain during exercise. The giant accessory spleen may be misdiagnosed as a retroperitoneal tumor. It appears as an enormous retroperitoneal mass (20). CT with contrast administration reveals a homogeneously strengthening mass. Clinical symptoms may also occur when complications such as torsion, necrosis, infarction or traumatic bleeding occur in the accessory spleen. In imaging studies, the arterial phase of contrast enhancement is significantly homogeneous, which is different from the enhancement pattern of the main spleen. This type of accessory spleen should be appropriately differentiated from the ecotopicphaeochromocytoma, which has different symptoms, and lymphoma that can occur in any part of the body, including lymph nodes, tonsils, spleen and bone marrow, with painless enlargement of the lymph nodes, local changes and systemic symptoms such as fever, weight loss or night sweats, which will not be observed in the case of the giant accessory spleen. The final diagnosis is made on the basis of the histopathological examination of the lesion. Laparoscopic or retroperitoneal laparoscopic resection of tumors is less invasive and helps to confirm the diagnosis, but it is more risky in the case of the giant spleen (20). During splenectomy, the giant spleen is at risk of rupture and hemorrhage. The giant accessory spleen is a rare cause of recurrent abdominal pain in children, and knowledge of the typical results of imaging modalities is essential for a precise preoperative diagnosis (21).

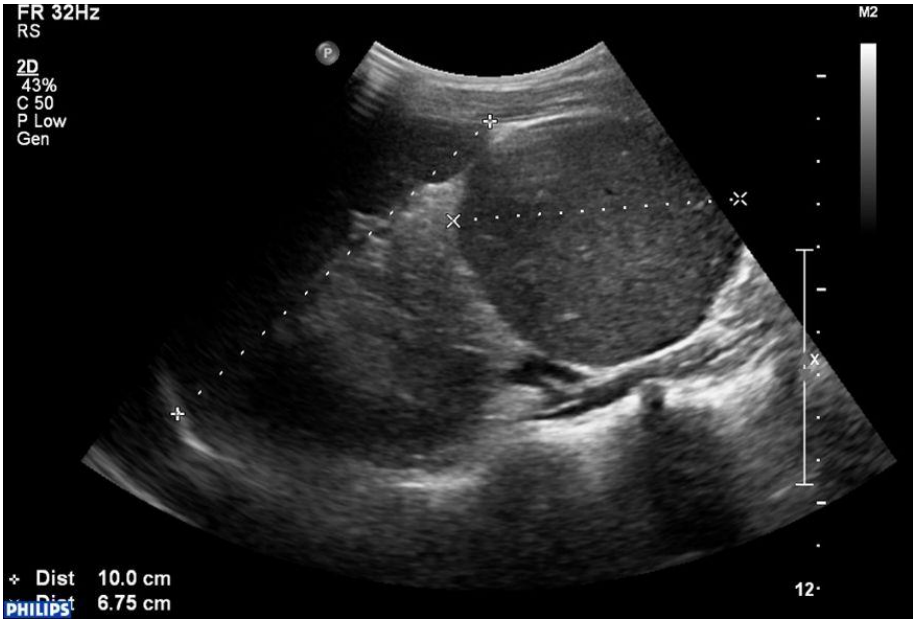

Fig. 6. The giant accessory spleen. Ultrasound scan of the splenic area with the giant, round accessory spleen, almost 7 cm in diameter (calipers).

Discussion

The anatomical varieties of the spleen have different clinical effects. The radiologist's knowledge of the various variants and abnormalities of the spleen is necessary for the development of a differential diagnosis and the correct diagnosis. In our work, we presented the aspects of the diagnosis of these lesions, important from the clinician's point of view, which directly determine further treatment. It is necessary to be aware of these rare changes as they may be life-threatening (in case of asplenia) due to risk of serious infection or sudden abdominal pain (in case of wandering spleen) or cause of misdiagnosis (differentiation of

giant spleen from ectopic phaeochromocytoma and lymphoma) or may be the cause of too relentless treatment (in the case of fusion of the spleen and gonads). The spleen is not considered as a vital organ and its congenital abnormalities are often overlooked.

Conclusion

Structural changes of the spleen may result in non-specific clinical symptoms, which may confuse an inexperienced physician. Inaccurate diagnosis and late diagnosis have various negative, often even fatal, consequences. In order to make an appropriate diagnosis, and thus help the patient in a timely manner, it is necessary to have extensive knowledge about the variants of the structure of the spleen. Knowledge of anatomical variations is essential for the development of a differential diagnosis, which enables the correct diagnosis to be made.

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