Conservative Management in Three Cases of Prenatally Recognized Splenic Cyst Using 2D, 3D, Multi-Slice and Doppler Ultrasonography

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Key Words

Prenatal diagnosis · Splenic cyst · Ultrasonography

Abstract

The aetiology, differential diagnosis and management strategies of the foetal spleen affected with a cystic lesion are discussed. In the current literature, there are very few reports that relate to antenatally diagnosed splenic cyst. Our study presents 3 case reports that were first suspected due to anisoechogenic structures detected during routine ultrasonographic examination at the 27th, 31st and 34th weeks of gestation. All 3 cases were further characterized by the lack of pathological power Doppler findings inside and around the lesions, and were morphologically refined by prenatal 3D ultrasound imaging. All findings were reconfirmed postnatally. No complications such as cyst expansion, subcapsular bleeding or acute abdomen have developed, and all 3 cystic lesions have regressed spontaneously after birth.

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Introduction

In human foetuses, the spleen starts to develop during the 6th to 7th week of pregnancy. It is formed through the aggregation of reticular mesenchymal cells in the

rear gastric mesenterium. It participates in production of megakaryocytes and in thrombopoiesis [1], but not in erythropoiesis [2]. The spleen can be identified by ultrasonography around the 20th week of pregnancy (W 20) [3]. Congenital splenic cyst is usually diagnosed in later childhood, when it may cause various difficulties (expansion, capsule bleeding or rupture, sudden abdominal incidents) [4]. Our study reports on 3 cases of congenital splenic cyst diagnosed retrospectively. After first diagnostic suspicion during routine ultrasonography (USG) in the 27th, 31st and 34th weeks of pregnancy, 3D MRI-like and Doppler ultrasound imaging were included in the framework of diagnostic algorithms in all cases. The cysts were confirmed postnatally in all 3 cases.

Methods

Between August 1999 and July 2007 (an 8-year period), 7,897 pregnancies were screened in our prenatal USG unit. There were no selection criteria applied, and all pregnant women referred to our unit were routinely screened for the presence of any foetal anomalies. USG examinations of the pregnancies described in case reports were performed on Accuvix QX supplied by Medison with the use of an abdominal 2D/3D/4D probe (including MRI-like 3D XI™ eXtended Imaging and Multi-Slice View) and/or on Voluson 730 Pro supplied by General Electric, using an abdominal 2D/3D/4D probe. Power Doppler ultrasound (PD-US) exami-

Fig. 1. Splenic cyst in the 28th week of pregnancy, diameter 25.5 mm.

Fig. 2. 3D MRI-like imaging of case 1. A series of 5-mm-thick sections was made from the 3D scan that confirmed the location of the cystic body in the spleen.

Fig. 3. Splenic cyst in week 30+5 of pregnancy located in the hilus. Diameter 9.1 mm. C = Cyst; S = spleen.

nation was performed to evaluate eventual flow parameters in the suspected lesions.

Case Report 1

The mother was 30 years old, terci para, and was referred for a superconiliary USG examination in week 27+4 of pregnancy due to a cystic body in the foetus located in the upper left quadrant of the peritoneal cavity. The previous USG examination performed in the first trimester was negative, as was the triple test. During our USG examination, a cystic body was found in the spleen, with a diameter of 25.5 mm (fig. 1). It was determined by MRI-like 3D XI and Multi-Slice View to be a splenic cyst (fig. 2). PD-US examination did not reveal any stream inside the cavity, and blood flow characteristics around the lesion did not differ from the rest of splenic tissue. All other sonographic findings were normal. During the following USG check-up in the 31st week of pregnancy, no change was observed in the size and nature of the cyst. The

baby was delivered in the 40th week of pregnancy (3,650 g/51 cm). The USG examination in the newborn confirmed the splenic cyst, which had a diameter of 26 mm. The follow-up ultrasonography 3 months later revealed a spontaneous regression in the cyst size by 10 mm, and 6 months after the delivery the cyst had regressed completely.

Case Report 2

The mother was 39 years old, primipara, and was admitted for a routine USG screening in the 31st week of pregnancy. A triple test in the 16th week of pregnancy was negative. Previous sonographic examinations showed no pathology. During our examination, a cystic body was found between the stomach and the spleen, with a diameter of 9.1 mm (fig. 3). 3D examination confirmed a splenic cyst. PD-US examination did not reveal any stream inside the cavity, and blood flow characteristics around the lesion did not differ from the rest of splenic tissue. During the

Fig. 4. Splenic cyst sized 2.02 × 0.91 cm found in week 34+0 of pregnancy.

USG follow-up in the 38th week of pregnancy, the cyst was of an identical size and flow characteristics. The baby (3,880 g/50 cm) was delivered in the 40th week of pregnancy by Caesarean section due to a non-progressing delivery. The USG examination of the newborn confirmed the splenic cyst. Normal structure of the spleen with no cystic alterations was found 6 months later during the sonographic check-up.

Case Report 3

The mother was 25 years old, primipara, and was sent for a superconsiliary USG examination due to a positive triple test. The Down's syndrome risk was 1 in 240. USG in the 19th week of pregnancy was without notable signs. During USG in the 34th week of pregnancy, a cystic structure was found in the abdomen located in the vicinity of the spleen, sized 2.02 × 0.91 cm (fig. 4). Splenic cyst was confirmed by means of 3D MRI-like imaging and 3D surface mode. PD-US examination did not reveal any stream inside the cavity, and the flow characteristics around the lesion did not differ from the rest of splenic tissue. USG in the newborn confirmed a splenic cyst. Follow-up USG 6 months later showed a normal splenic structure with no cystic alterations.

Discussion

In our 3 cases of foetal splenic cysts, no trauma, injury or parasitic disease were found in anamnesis or by clinical examination. In all 3 cases, there was a smooth-walled, anizoechogenic cyst of unknown aetiopathogenesis.

Literature Records on Prenatal Splenic Cyst

In 1988, Lichman and Miller became the first authors to report on prenatal USG-diagnosed splenic cyst [5]. The

majority of splenic cysts described in the literature were diagnosed between the 25th and 35th weeks of pregnancy [6, 7]. The spleen can be detected in foetus only after the 20th week of pregnancy determined by the last menstruation [8]. USG (with high-frequency probes and high-resolution capacity) has significantly improved the possibilities of prenatal diagnostics. Today, it is possible to detect cysts of several millimetres in size in the foetus, which were impossible to identify in the past. Kabra and Bowen [9] report on the detection of a 6-mm congenital splenic cyst and Lopes et al. [10] have diagnosed a 10-mm cyst.

Aetiopathogenesis and Classification of Splenic Cysts

Splenic cysts may be divided into 2 types depending on their epithelium: primary cysts and secondary cysts (or pseudo-cysts). The primary cyst capsule is formed from a cellular layer, while secondary cysts have a fibrous capsule. Primary cysts include serous, vascular, mesothelial, infectious and neoplastic (epidermoidous, dermoidous, haemangiomas and lymphoangiomas) cysts. Secondary cysts may be of a post-traumatic, inflammatory or a degenerative origin. Congenital cysts represent 25% of all splenic cysts.

In the majority of cases, the aetiology of splenic cysts is unknown. From the developmental aspect, splenic cyst formation may occur via the following mechanisms:

- (1) Invasion and metaplasia of pluripotent cells of various origins into the splenic parenchyma during foetal development.
- (2) Inclusion of a cellulose mesothelium into the spleen during organogenesis.
- (3) Invagination of peritoneal endothelial cells into the developing spleen.
- (4) Dilatation of lymphatic areas.

A typical USG image of a congenital splenic cyst includes an anizoechogenous formation with a smooth wall, located in the upper left quadrant of the abdominal cavity. Epidermoidal cysts are characterised by an irregular structure and thickened rear wall. Secondary cysts may also be of an anizoechogenous nature, but more often they display calcifications. Bleeding or haematomas are characterised by a hypoechogenous structure.

Differential Diagnosis

Differential diagnostics is necessary for any cyst located in the upper left lateral quadrant of the abdominal cavity. It may include a congenital cyst in the kidney, hydronephrosis, cystic dysplasia of kidneys, obstruction or duplication of the urinary tract, epinephral cysts, hepatic cysts, pancreatic cysts and pseudo-cysts, annular pancreas, mesenterial cysts, omental cysts, congenital stomach

duplication, duodenal atresia, ovarian cysts in females, and very rarely, hydrosalpinx. Cysts in kidneys can be excluded after detection and visualisation of the kidneys. Adrenal tumours are usually heterogeneous. Cysts in the gastrointestinal tract are more often located in the front right side of the abdominal cavity. It is currently unknown whether specific flow indices similar to Doppler examination (e.g. in adult ovarian tumours and some prenatal anomalies) may be reliably and routinely applied to prenatal cases of cystic lesions, but it is always advisable to screen for the signs of malignant nature (signs of angiogenesis, irregular blood flow, deviated shape of vessels, anarchic vascular architecture, irregular branching, vessel caliber and/or resistance changes, microaneurysms, vascular lakes, arteriovenous shunting, etc.), and further research in this area is necessary [11–13].

Clinical Management

Upon finding splenic cysts, it is necessary to examine other body organs (liver, kidneys, lungs and pancreas) for the presence of cysts so that congenital polycystic disease can be excluded. Subsequently, it is necessary to monitor the patient prenatally, and to monitor the newborn postnatally. It is important to observe any alterations of the cyst, its progression or regression up to its extinction. The majority of congenital splenic cysts are asymptomatic with a good prognosis. Usually, the cysts regress. In 1995, Garel and Hassan [14] became the first to describe spontaneous regression of congenital splenic cysts. Only a few case studies report the growth of a cyst and its complications. A growing cyst causes compression of other ab-

dominal cavity organs, it can burst spontaneously, demonstrate bleeding and cause sudden abdominal pain. This is the reason why a prenatally identified splenic cyst must be monitored up to its complete extinction. The management strategy recommends a conservative approach, and an invasive solution is only recommended in cases where complications arise. Depending on the severity of the situation, USG-controlled thin-needle aspiration of cysts, laparoscopic puncture and cystectomy can be performed. In emergency cases, treatment is partial or total splenectomy [9]. Any invasive methods should be applied only in cases when the cyst becomes symptomatic.

Conclusions

Spontaneous and asymptomatic regression of non-malignant splenic cysts is the usual fate of this developmental malformation. However, careful monitoring must be applied in all cases due to rare cases of life-threatening complications, such as cyst rupture and bleeding.

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