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CASE REPORTS

Congenital Splenic Cyst; Video Case Presentation and Literature Review

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Abstract Congenital splenic cysts have been reported to usually regress spontaneously within a few months postnatally. A splenic cyst was detected in the ultrasonography of a 27-year-old pregnant woman at her 34th week of gestation. The size of the splenic cyst remained constant in the follow-up of the patient. In the ultrasonography performed in the postnatal 1st year, it was determined that the cyst persisted and remained the same size. There is no algorithm for the follow up of cysts in the literature, and information about congenital splenic cysts will be presented in this case report and review.

Keywords Fetal ultrasonography · Splenic cysts · Embryology · Spontaneous regression · Perinatology · Fetal splenic cyst

Introduction

Congenital splenic cysts are benign lesions that are usually detected in the third trimester. On ultrasound, these are sonographically recognized as an anechoic cystic lesion in the left upper quadrant, behind the stomach. These cysts have been reported to usually regress spontaneously within a few months postnatally. In this study, we report a case of a congenital splenic cyst, which was detected in the third trimester and still persists at a one-year postnatal follow-up.

Presentation of Case

A 27-year-old woman (gravida 3 parity 2) no history of comorbidity, alcohol or smoking presented at our clinic. Her previous ultrasonography (USG) performed in the first trimester had normal findings and the triple test was negative.

In the ultrasound evaluation performed at the 34th week of gestation, a 17 × 16 mm cystic mass was detected in the left upper quadrant of the fetus's abdomen (Fig. 1). The stomach and left kidney were normal. The cyst was in the spleen. (Video 1). Color Doppler evaluation showed no abnormal blood flow around the cyst and the rest of the splenic tissue was normal. No abnormality was observed in other organ systems in sonographic follow ups and the size and shape of the cystic lesion was unchanged. For this reason, it was decided to follow up the cyst and not to make any intervention. The baby (4225 g) was delivered in the 39th week of pregnancy vaginally, with a normal 9 (1st minute) and 10 (5th minute) APGAR. No hypoglycemia or hyperbilirubinemia was observed in the neonatal period and blood values were within the normal range. All findings were reconfirmed postnatally. In our one-year sonographic follow-up performed at 3-month intervals, the cyst persists unchanged. The baby is now 1-year-old, does not have any disease and is healthy.

Discussion

In this case report, a case with a congenital splenic cyst at the 34th gestational week is presented. The majority of the cases reported in the literature regressed in the a few months

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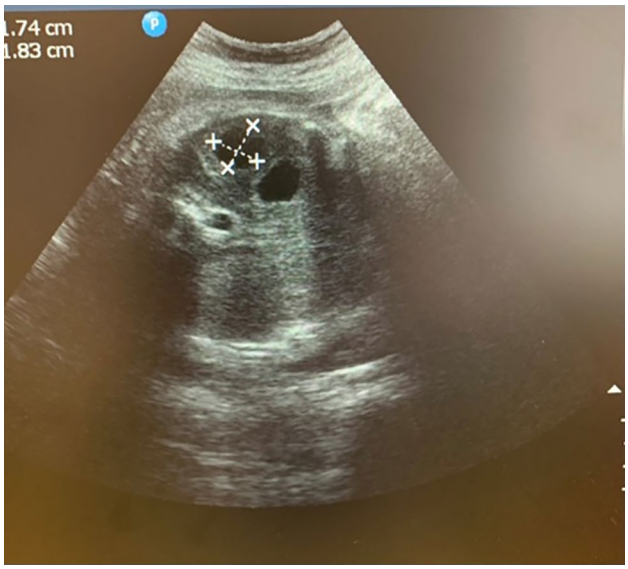

Fig. 1 Ultrasound of the fetal left upper abdomen at 34 weeks showing a single cystic mass within the spleen

postnatally and a persistent cysts after 1 year is rare. There is no specific follow-up algorithm in the postnatal follow-up of these cysts, and our case will contribute to the scientific literature. The review of the literature was performed through a computerized search of the PubMed/MEDLINE database combining the terms ''splenic cyst'' and with ''fetal'', ''prenatal diagnosis'', and ''antenatal diagnosis'' without limits of time and written in English. As a result of the scan, it was determined that there were 13 publications (43 cases in total) describing a congenital splenic cyst. Sonographic and clinical features of the cases are summarized in Table 1 [1–13].

The fetal spleen arises as an aggregation of reticular mesenchymal cells in the dorsal mesentery of the stomach during the 6-7th gestational weeks. A splenic cyst can be typically diagnosed as an anechoic cyst behind the stomach. The fetal spleen can be visualized at transverse sonographic view of the fetal left upper abdomen, behind the stomach at 20 weeks of gestation [2]. With the ongoing improvement of ultrasound technology, it is currently possible to detect splenic anomalies even earlier (1 case was described at 17 weeks) and also of smaller sizes [2].

Non-parasitic splenic cysts are divided into two types as true (primary) cysts with a cellular layer and pseudo (secondary) cysts with a fibrous capsule. True cysts include congenital, vascular, serous, infectious, and neoplastic (epidermoid, dermoid, mesothelial, lymphangioma) cysts. Pseudocysts may develop secondary to trauma, hemorrhage, infarction, inflammation, and are generally observed in the adult group. Congenital splenic cysts constitute 25% of true cysts [9, 14]. The pathogenesis of primary splenic cysts is not clear, but possible theories have been suggested: (1) Involution and metaplasia of pluripotent cells in the splenic parenchyma during fetal development (2) Inclusion of celomic mesothelium during organogenesis (3) Invagination of peritoneal endothelial cells (mesothelium) within the developing spleen (4) Dilatation of the normal lymphatic space [9].

Prenatal sonographic diagnosis was first reported by Lichman and Miller in 1988. In their report, a 13 mm sonolucent cyst in the left upper quadrant was detected at 33 weeks of gestation [4]. After the first report in 1988, a case with a 45 mm splenic cyst at 32 weeks of gestation was published [15]. Laparotomy was performed in this case because the cyst had grown up to 70 mm in the postnatal follow-up, histopathological examination was reported as an epidermoid splenic cyst. The literature was scanned and it is seen that congenital splenic cysts were generally asymptomatic and had a good prognosis. In 1995, Garel and Hassan reported spontaneous regression for the first time [1]. In 2017 a tertiary clinic reported 10-years' experience. According to this report, half of the cases regressed within the intrauterine period or postnatal six months [16]. The true incidence of congenital splenic cysts is unknown.

When a splenic cyst is detected, it is necessary to scan other organs (kidney, liver, pancreas, lung) because of the possibility of a polycystic pathology. Urinary system (renal cyst, cystic dysplasia, hydronephrosis, obstructed duplication), genital system in female fetus (ovarian cyst, giant hydrosalpenx) suprarenal, gastrointestinal system (pancreatic pseudocyst, mesenteric cyst, omental cyst, choledochal cyst, hepatic cyst in the stomach, duodenal atresia) must be reviewed [12]. Another reported rare association is abnormal pulmonary venous [13].

Congenital splenic cysts are rare pathologies. This is mainly a third-trimester finding, but with the advent of high-resolution ultrasound equipment and the increasingly frequent prenatal use of sonography, the detection week has shifted towards the second trimester. Splenic cysts are usually isolated. However, detailed sonography is required to exclude the differential diagnoses mentioned above. Although there are case reports confirming sonographic findings with magnetic resonance imaging, ultrasound provides sufficient information [17]. The longest reported time for cyst resolution is 2.5 years [18]. Therefore, when a splenic cyst is detected, follow-up in the postnatal period is recommended.

In conclusion, congenital splenic cysts should be kept in mind when a cystic structure is observed in the prenatal left upper quadrant. The size of these cysts should be followed. The family should be informed about the issue. These cysts are benign in nature, but; it should be explained that changes in the structure and size of the cyst must be followed with intermittent sonographic follow-up.

Table 1 Sonographic and clinical features of cases with congenital splenic cyst in the literature

	Maternal age	Parity	Gestational week	Cyst Size	Reported result
Lichman and Miller 1988 [1]	34	2	33	13	Unknown
Stiller et al. 1991 [2]	29	0	35	15	15 mm at postnatal 3rd month
Garel and Hassan 1995 [3]	Unknown	Unknown	32	10	Resolution at postnatal 2nd years
	Unknown	Unknown	28		16 mm at postnatal 3rd years
	Unknown	Unknown	33	16	17 mm at postnatal 6th month
Okada et al. 1995 [4]	32	1	17	13	Postnatal 8 mm
Yılmazer et al. 1998 [5]	26	0	31	12	Resolution at postnatal 7th month
Lopes et al. 2001 [6]	24	0	25	11	Resolution at postnatal 6th month
Kabra and Bowen 2001 [7]	30	0	20	6	4 mm at postnatal 6th month
Saada et al. 2006 [8]	26	0	23	10	Resolution at postnatal 2nd month
	33	0	25	10	Resolution at postnatal 3rd month
Chen et al. 2007 [9]	37	2	26	17	Resolution at postnatal 1st month
Dankovcik et al. 2009 [10]	30	3	27	29	Resolution at postnatal 6th month
	39	0	31	9	
	25	0	34	20	
Venkataraman et al. 2009 [11]	29	Unknown	23	12	Resolution at the postnatal 13th month
Sauvageot et al. 2018 (14 cases) [12]	Mean 30 SD (±3,8)	Unknown	Median 30 IQR (26–32)	Mean 9,9 SD (±3,5)	Half of the cases resolved in the postnatal 6 months
Sepulveda et al. 2018 (9 cases) [13]	Median 32 Range (22–39)	Unknown	Median 30 Range (22–37)	Mean 12 SD (±5)	In one case, the cyst increased from 8 to 11 mm. Other cases resolve in a maximum of 2.5 years

Implications for Clinical Practice

Congenital splenic cysts should be kept in mind when a cystic structure is seen in the left upper quadrant before birth. Malignant transformation of these cysts has not been reported. Sonographic follow-up of their size would be rational.

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Code Availability No code available.

Declarations

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval Informed consent of the patient was obtained prior to the writing of the manuscript.

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