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Case Report

A large congenital pancreatic cyst mimicking a macrocytic lymphatic malformation[☆]

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ABSTRACT

Congenital pancreatic cysts (CPCs) are rare developmental anomalies that arise in-utero from the pancreas. They are exceedingly rare in the literature, and most are discovered post-natally. Prenatal diagnosis is uncommon with only 21 published reports of prenatally diagnosed CPCs in the literature. CPCs may form unilocular or multilocular macrocysts which can distort normal anatomy. There is considerable overlap of imaging features with other macrocystic lesions of the neonatal abdomen. Ultrasound-guided biopsy and analysis of cyst aspirate for pancreatic enzymes may assist with obtaining an accurate preoperative diagnosis. We report a case of a 37-week gestational age female infant born with a known prenatal 9.5 cm macrocystic intrabdominal mass. An intrabdominal lymphatic malformation was initially diagnosed based on clinical and imaging features. Since conservative therapy with with cyst drainage and serial sclerotherapy was not effective, an ultrasound-guided biopsy was performed to rule out malignancy. Pancreatic tissue was identified on pathology. An exploratory laparotomy and total cystectomy was performed which confirmed the diagnosis of congenital pancreatic cyst originating from the pancreatic tail. This case highlights the diagnostic challenge of congenital pancreatic cysts and the importance of a multimodal and multidisciplinary diagnostic approach.

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Case report

At 19 weeks gestational age (GA), a female fetus was incidentally found to have an abdominal mass. Specifically, an ultrasound (US) demonstrated a septated cystic abdominal mass without internal color flow measuring 2.8 × 2.2 × 2.0 cm in

the left upper abdomen (Fig. 1A). The cystic lesion (arrow) appears superior to the left renal hilum (Fig. 1B) which raises the possibility of adrenal origin, including an evolving adrenal hemorrhage or cystic neuroblastoma. Alternatively, a duplex left kidney with an obstructed upper pole collecting system, or congenital cystic dysplasia of the upper pole could also have this appearance.

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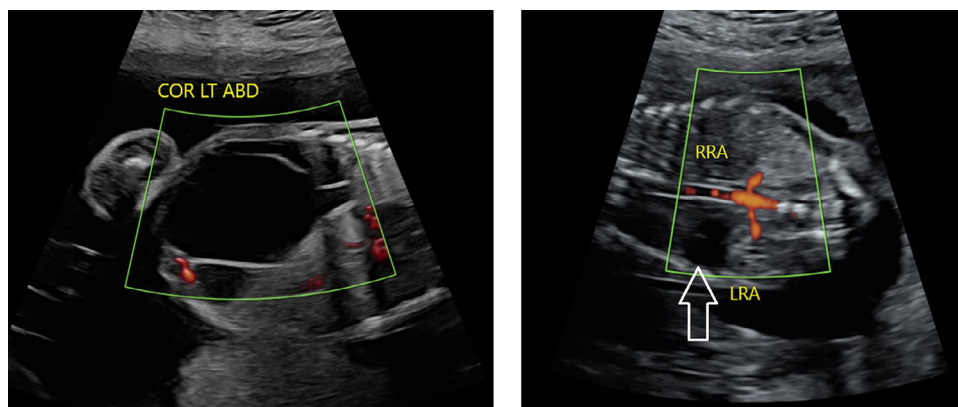


Fig. 1 – (A) Prenatal US at 19 weeks, 2 days gestational age showed a cystic lesion with internal septations and no internal color flow in the left upper abdomen. **(B)** A coronal image of the fetal retroperitoneum shows the cystic lesion (arrow) right (RRA) and left (LRA) renal arteries branching from the aorta, extending to the renal hila.

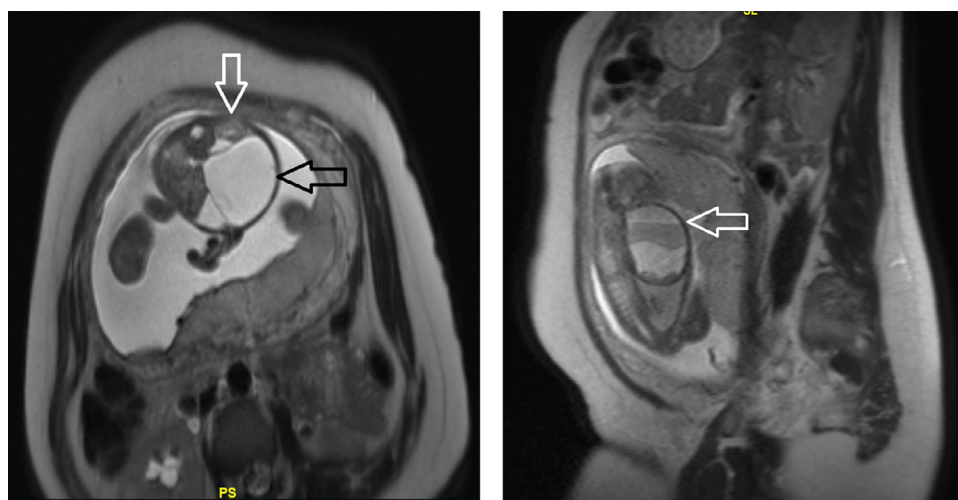


Fig. 2 – (A) Fetal MR performed at 26 weeks, 2 days shows interval growth of the left upper abdominal lesion. The cystic mass (black arrow) is separate from and compresses the left kidney (white arrow) posteriorly. **(B)** Fetal MR shows a hypointense-hyperintense fluid-fluid level within the multicystic lesion, suggesting interval hemorrhage on coronal T2-weighted sequences. **(C)** There is no finding to definitively attribute the lesion to its pancreatic tail origin, even in retrospect. The pancreatic tissue (white arrow) appears separate from the lesion and there is no visible continuity between the lesion and the pancreatic duct. The gallbladder (black arrow) is identified as an anatomic landmark as well.

Fetal magnetic resonance imaging (MRI) performed at 26 weeks, 2 days GA showed interval growth of the cystic mass to $5.4 \times 4.5 \times 6.1$ cm (Fig. 2A). The rapid growth, large size, multiple loculations, and relative anterior position to the left kidney expanded the differential to include a cystic ovarian lesion with possible torsion, or a mesenteric lymphatic malformation (LM). Additionally, a new fluid-fluid level was apparent, suggesting internal hemorrhage and/or proteinaceous debris (Fig. 2B) which are features often seen in both LM and ovarian lesions (particularly with torsion). The pancreas is identified just medial to the cystic lesion (Fig. 2C); however, its origin cannot be determined even in retrospect as there is no pancreatic tissue surrounding the lesion or clear connection between the lesion and the pancreatic duct.

The mother and fetus were monitored with serial US, and labor was induced at 37 weeks GA due to maternal hypertension. The infant was born via spontaneous vaginal delivery without fetal distress. She was asymptomatic, and on exam, her abdomen was soft with mild distention and no palpable mass. A postnatal US was obtained which showed continued growth of the mass to $3.8 \times 9.5 \times 9.1$ cm in the left upper abdomen near the spleen with extension toward the midline (Fig. 3). The mass was macrocystic with some echogenic debris and no internal color Doppler flow. The ovaries were not visualized.

An MRI was performed to further characterize the origin. The signal characteristics of the lesion were that of septated simple fluid, with pockets of T2 hyperintensity separated by

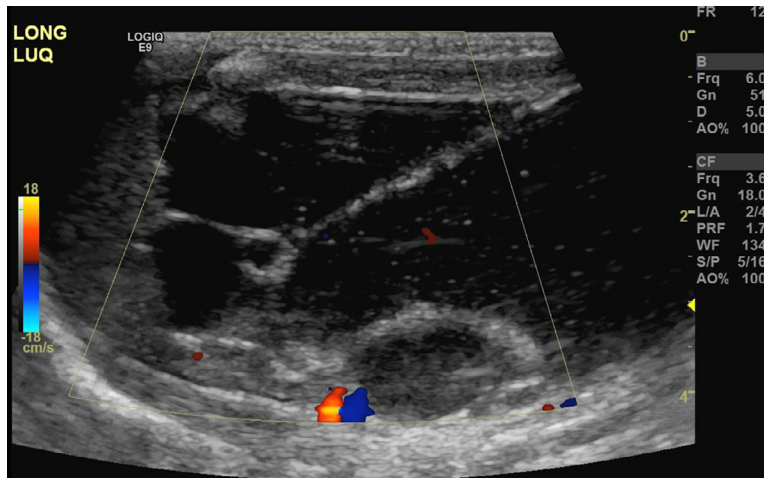


Fig. 3 – Postnatal US obtained immediately after birth demonstrates further interval growth of the multiloculated cystic mass to 3.8 × 9.5 × 9.1 cm. There are thin septations and echogenic debris, with extension to the midline. The ovaries were not seen.

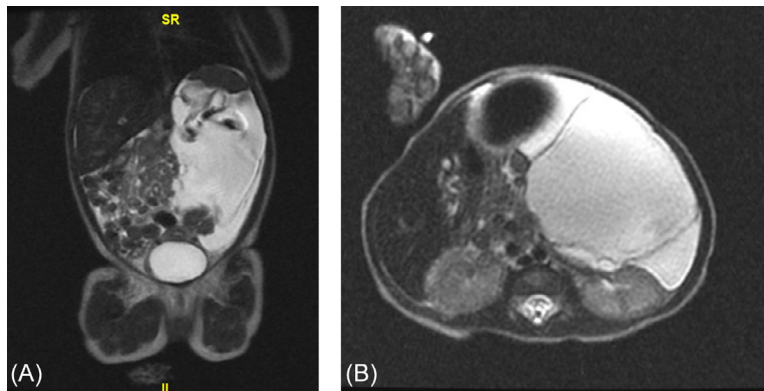


Fig. 4 – Postnatal MR was performed without contrast on the 2nd day of life demonstrating coronal (A) and axial (B) views of the known cystic multiloculated mass measuring 5 × 9.3 × 9.4 cm. The large size and thin septations have the appearance of a lymphatic malformation, with T2 hyperintense fluid signal. The normal ovaries were identified separately from the lesion.

T1 and T2 hypointense septae (Fig. 4). The ovaries appeared normal and separate from the cystic mass. Also, a normal left adrenal gland was seen. The mass appeared to be centered lateral to the descending colon, in the left paracolic gutter. Given these findings, an abdominal LM was presumed over other cystic masses. On the 3rd hospital day, she remained asymptomatic including tolerating feeds and having bowel function. The patient was discharged with close outpatient follow up for management of presumed LM.

At 2 weeks of age (39 weeks GA), she developed abdominal distension and some feeding intolerance which prompted evaluation and repeat US. This showed interval growth of the mass to 7.3 × 11.1 × 13.3 cm. She was readmitted to the hospital for further workup and management. To rule out a need for emergent surgical intervention (ie, bowel obstruction), upon admission a CT scan was obtained. This showed further interval growth (13.9 × 7.9 × 11.3 cm), again centered in the left paracolic gutter, with rightward/medial displacement of the

intestines (Fig. 5). There was no evidence of bowel obstruction or hypoperfusion. Given the enlargement of the presumed LM, she underwent cyst drainage followed by sclerotherapy with doxycycline by Interventional Radiology (Fig. 6). The cyst aspirate initially grew gram positive cocci resembling streptococci and gram-negative rods. Infectious disease was consulted and recommended initiation of empiric broad spectrum antimicrobial therapy. The working diagnosis was an infected LM due to contamination by surrounding gut microflora.

She responded well to drainage and sclerotherapy initially, with decreased abdominal distension. Her drain was removed when the drainage was <30 mL/day on the 2nd day after placement. Improvement was further corroborated by an interval MRI which showed mass reduction to 5.2 × 6.2 × 8.2 cm (Fig. 7). Interestingly, there were multiple newly visualized smaller cysts at the periphery of the central sclerosed macrocyst. There was no mass-like enhancement to suggest a solid component. It was unclear if these were de novo cysts

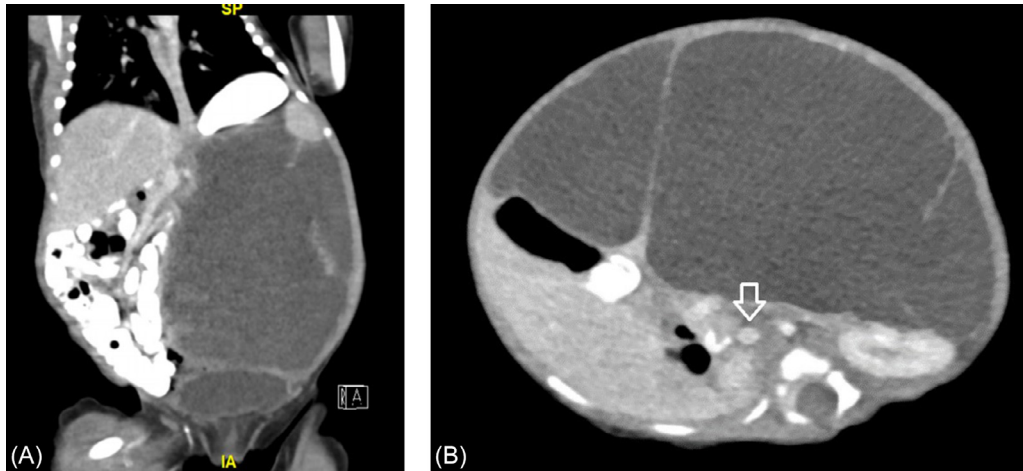


Fig. 5 – CT abdomen with enteral and IV contrast coronal (A) and axial (B) views demonstrating further interval growth of the cystic mass crossing midline. There are no solid or fatty components or calcifications. There was no decreased attenuation of the solid organs or hyperenhancement of adrenal lesions. The inferior vena cava (arrow) also maintains its rounded contour and is not flattened by increased intra-abdominal pressure. Such findings would suggest intra-abdominal compression syndrome. There are no dilated bowel loops to suggest bowel obstruction.

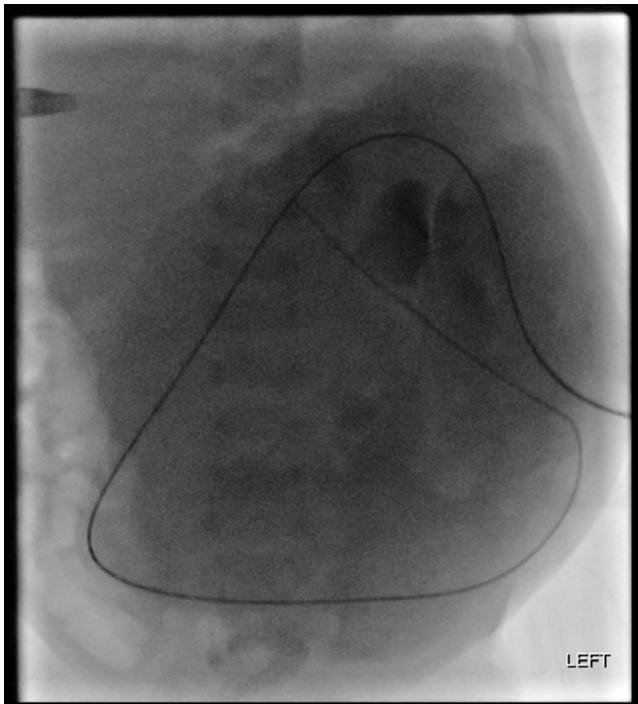


Fig. 6 – A percutaneous drain was placed to decompress the lesion due to the declining clinical status. The wire is shown within the contrast-filled cavity, which now takes up most of the intra-abdominal space. The lesion was sclerosed with doxycycline, as it was thought to be a LM.

or cysts that had become visible due to the decompression of the macrocyst from the aspiration and sclerotherapy.

An US-guided core needle biopsy was obtained of the cystic lesion. Although this biopsy showed no malignant cells, the

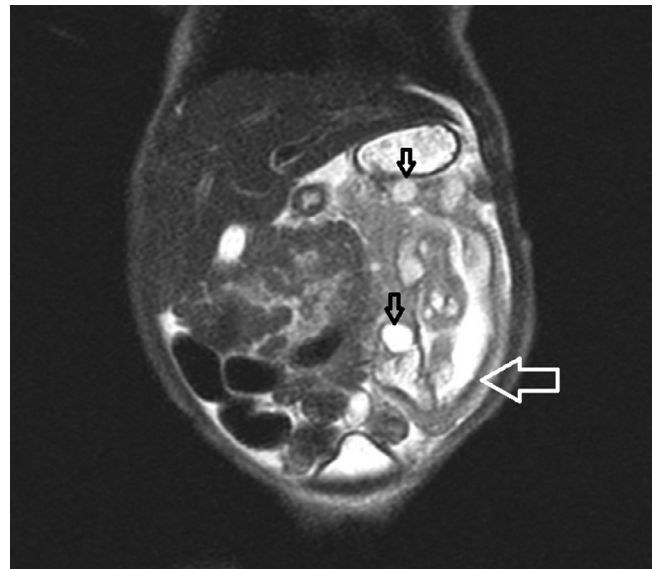


Fig. 7 – A post-treatment MR showed interval reduced size of the multilocular cystic mass, with expected changes of sclerotherapy including a contracting, thickened wall of the dominant cystic space (white arrow); however, numerous new, smaller cystic structures had also appeared at the periphery (black arrows).

presence of pancreatic acinar and ductal cells among fibro-inflammatory cells was surprising. The images of the US-guided biopsy were further reviewed to confirm inadvertent pancreatic tissue biopsies, and no US image contained pancreatic tissue (Fig. 8). Biochemical assays were pursued of both the previously aspirated cyst fluid and serum. These returned normal (serum amylase <5 U/L, serum lipase 55 U/L, and cyst amylase 21 U/L). Thus, the diagnosis remained ambiguous.

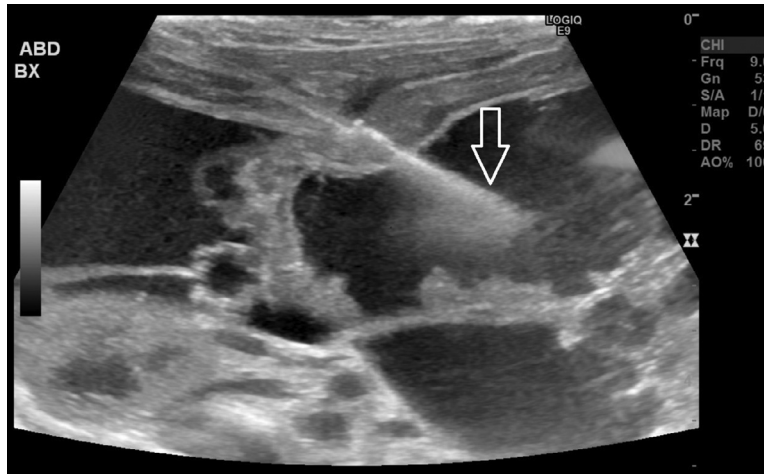


Fig. 8 – US-guided core needle biopsy of the lesion. The needle (arrow) is shown entering from the left lateral abdominal wall, with its tip in a cystic space. The fluid was aspirated and core samples were obtained of the lesion septations.

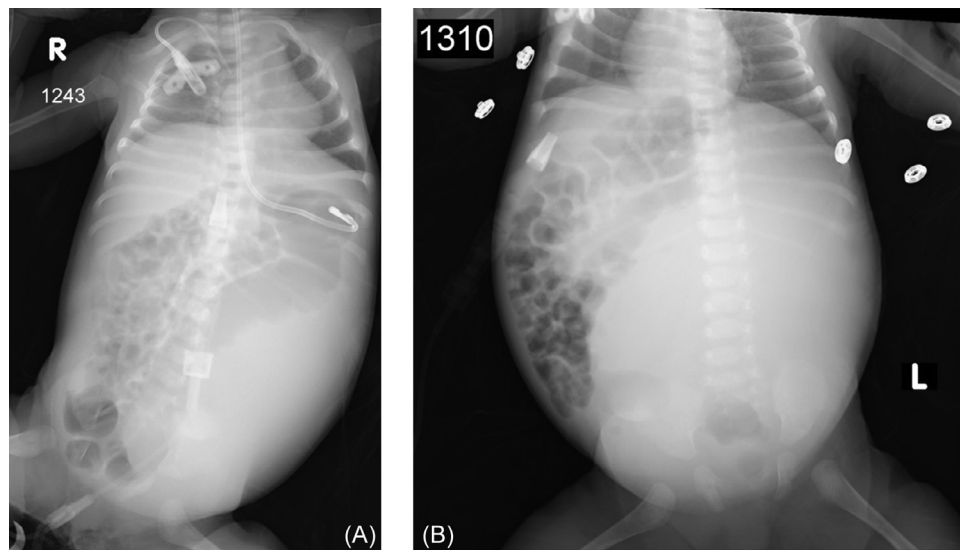


Fig. 9 – Plain abdominal x-rays. Interval placement of nasogastric tube for decompression (A), followed by rapid cyst recurrence (within 24 hours) with associated mass effect on the bowel (B).

The infant simultaneously developed recurrence of abdominal distention and feeding intolerance. Abdominal x-ray was concerning for growth of the mass again with displacement of the intestines to the right hemiabdomen (Fig. 9). Due to recurrent symptoms and concern for an underlying congenital pancreatic cyst (CPC) in context of the biopsy results, operative exploration and mass resection was pursued. A large loculated cystic mass was identified (Fig. 10A). It appeared to originate from the pancreatic tail (Fig. 10B). The mass was circumferentially isolated from all adherent structures and a total cystectomy with ligation of the pancreatic tail was performed. A cyst was also found on the colonic mesentery, and this too was excised. Final pathology of the mass showed squamous epithelium with associated pancre-

atic tissue and thus confirming diagnosis of CPC. The presumed mesenteric cyst returned as a benign lymph node.

The patient had an uncomplicated postoperative course and was discharged 6 days after surgical resection of CPC on oral feeds and cessation of antibiotics. She continues to be asymptomatic with appropriate growth parameters. On formal genetic testing, no mutations or syndromic associations with CPC were identified.

Discussion

Abdominal masses can be incidentally encountered on routine fetal ultrasound. Reportedly, 0.01 % of births are asso-

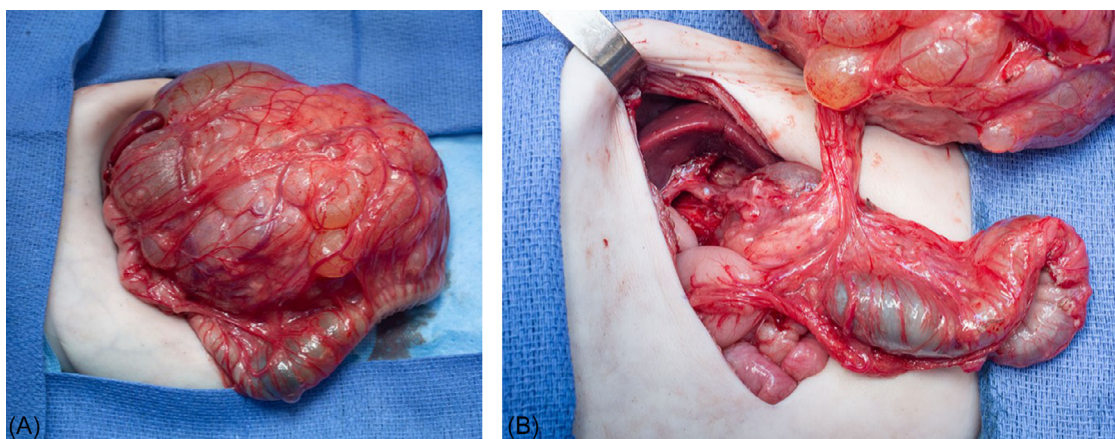


Fig. 10 – Intraoperative image of the congenital pancreatic cyst immediately upon abdominal access (A). Note origin of the cystic mass from the pancreatic tail (B).

ciated with an intra-abdominal mass [1]. The most common fluid-filled structure mimicking a cystic mass in the fetal and neonatal abdomen is hydronephrosis; [2] however, an ovarian cyst is the most common etiology for a true prenatal abdominal cyst in a female fetus [3]. Other lesions include enteric duplication cysts, mesenteric cysts, and choledochal cysts [4]. CPCs are true cysts that arise from the pancreas but are comparatively rare [5].

The first case of a prenatal CPC was reported in 1979 [6]. Hopper et al [6] described a bilobed-CPC causing duodenal compression resulting in polyhydramnios. The neonate expired shortly after delivery due to respiratory failure from pulmonary hypoplasia. Twenty-one reports of prenatally diagnosed lesions have since been published [6–24] and ours provides the 22nd. These cases primarily describe CPCs as simple-appearing, unilocular cysts of various sizes in the upper abdomen. The unique aspects of this case include the large size and multilocular nature of this CPC which more closely mimics a LM than other alternative diagnoses. The interval growth of the lesion, with the appearance of multiple additional smaller peripheral cysts after sclerotherapy prompted US-guided biopsy, with a result that led to surgery. This report highlights the importance of a multidisciplinary and multimodal diagnostic work-up to arrive to the diagnosis of a CPC.

Less than 5% of LM occur in the abdomen [25]. LM in this region may originate from the mesentery (most common), and infrequently from retroperitoneum, gastrointestinal tract, or solid organs, although these lesions are known to cross tissue planes to involve multiple anatomic spaces and organ systems [25]. Abdominal LM and CPC have overlapping imaging features. In the above report, the location of the presumed LM was both mesenteric and retroperitoneal (as specified on the fetal MR), given the extension into the left pericolic gutter on postnatal MR imaging. Lymphatic malformations are a type of vascular malformation and can occur as part of venolymphatic malformations which contain both cystic spaces and slow-flow venous channels [26]. On abdominal US, LM are cystic, often contain septations, and may demonstrate layers of echogenic debris within the cystic components attributable

to hemorrhage into the potential spaces [25,26]. There are no solid components.

US is often the initial diagnostic test pursued as it is widely available, rapidly obtained, and relatively cost effective. Cross-sectional imaging such as MRI or CT may be required for more detailed analysis such as anatomic origin and distribution [27]. On CT scan, abdominal LMs appear as fluid-density lesions with peripheral wall enhancement, with thin, enhancing internal septations and layering debris [25]. Like our report, others have described similar findings [12]. MRI of abdominal LMs likewise demonstrate fluid-intensity T1 hypointense, T2 hyperintense lesions with an enhancing wall and internal septations. Gradient sequences may highlight characteristics of blood products within layering internal debris, with variable T1 and T2 signal with patterns dependent on the chronicity of the hemorrhage [25]. However, despite such advanced imaging, even CT or MRI can be nonspecific to distinguish a LM from other cystic lesions. Given the rapid growth of our patient's lesion, it is important to highlight the possibility of an intra-abdominal cystic tumor in this age group, a known pitfall of attributing cystic abdominal lesions in the newborn to a benign entity.

The diagnostic work-up of CPC often includes biochemical analysis, namely cyst aspiration with amylase and lipase levels. CPC is often associated with elevated cyst amylase values [17,28]. Castellani et al [17] found cyst amylase levels as high as 2771 U/L and lipase >6000 U/l. Although cyst lipase was not assessed in our case, we found normal serum lipase/amylase and normal cyst amylase. Theoretical explanations include peripheral origin of the cyst and lack of communication with pancreatic ducts, lack of underlying pancreatitis in our patient, and disruption of the epithelial lining of the cyst by infection [17,29,30]. This case confirms that biochemical analysis is not sufficient alone for the diagnosis of CPC, and that pathologic tissue analysis should be pursued when there is a high index of suspicion.

Percutaneous core needle biopsy remains a safe and effective alternative to surgical biopsy and is a preemptive step to surgical resection for large cystic abdominal masses in

neonates. Biopsy should be considered particularly if there are concerns for an underlying malignancy or alterations in surgical management. Nonetheless, surgical resection was curative in this infant, a finding that is consistent with prior reports of CPC having higher recurrence rates with aspiration alone [17]. Recurrence after aspiration is likely aided by the inherent regenerative potential of the pancreatic epithelial cells that line the cyst [29]. This supports a low threshold for surgical exploration in neonates with symptomatic abdominal masses of questionable etiology or suspected CPC.

In conclusion, both prenatal and postnatal diagnosis of CPC is rare. We report the 22nd prenatal case. This is the first case in the literature that has detailed initial treatment of a CPC with sclerotherapy for presumptive diagnosis of a LM. This approach prompted additional clinical work-up, and eventually surgical cystectomy. Both CPCs and LMs are relatively rare causes of cystic abdominal masses in neonates; however, they should both be considered in the differential for masses that do not fit the clinical or radiographic features of more common etiologies. Tissue diagnosis via core needle biopsy or surgical excision is gold standard and should be pursued when the radiographic diagnosis is not definitive or does not fit the clinical status of the patient.

Patient consent statement

Written consent was obtained from the patient's parents prior to the publication of this case report.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.04.024.

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