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Case Report

Invasive Abdominal Lymphatic Malformation: a Case Report with a Combination of Therapeutic Interventions

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Abstract

Abdominal lymphatic malformations account for less than 5% of all congenital lymphatic malformations. Although antenatal diagnosis is ascertained in some cases, postnatal imaging is essential to confirm the diagnosis, delineate the extent of the lesion, and provide an insight for possible therapeutic interventions. While surgical excision is considered the only curative measure, conservative management is recommended when the lesion is small and does not affect the anatomy or function of other intra-abdominal organs. Sclerotherapy is seldom used as the only therapy but is indicated when the lesion is large and invading nearby structures, although surgical excision might be warranted as an emergency intervention when the lesion is complicated by rapid enlargement, hemorrhage, infection, volvulus, or acute compartment syndrome.

We hereby present a case of abdominal lymphatic malformation diagnosed antenatally at 24-week gestation as suspected intestinal obstruction. Postnatal imaging confirmed a massive retroperitoneal macrocystic lymphatic malformation invading nearby structures. Conservative management was initially attempted, local drainage was achieved at the age of three weeks, sclerotherapy was done at two months, and laparotomy with excision performed at four months with an excellent overall outcome.

Keywords: *Abdominal Lymphatic Malformations; Neonate; Sclerotherapy; Antenatal*

Case Report

Clinical information

A male baby was delivered by cesarean section at 37+4 weeks of gestation to a 21-year-old mother. The mother had regular antenatal care. Ultrasound (US) study at 24-week gestation showed suspected intestinal obstruction. Subsequent US scans showed the same suspicion with no polyhydramnios and no evidence of hydrops fetalis. The baby was born with a birth weight of 4545 grams (99th centile), length of 52 cm (94th centile), and APGAR scores of 8 and 9 at 1 and 5 minutes respectively.

Physical examination at birth showed a male baby with no dysmorphic features, distended abdomen with no palpable masses, the anus was patent. No other abnormalities were detected. The baby was kept to nil per os, nasogastric tube was placed for decompression, and intravenous fluids started.

Imaging work up

X-ray abdomen was done after admission and showed dilated bowel loops mostly located in the upper abdomen nonspecific for a particular intestinal obstruction. On the second day of life, abdominal US was performed which reported the presence of fluid-filled bowel loops compressing and displacing the surrounding structures, a picture suspicious for intestinal atresia.

Gastrografin contrast enema study was then performed to rule out lower intestinal obstruction and was reported normal.

As a result, abdominal US was repeated and reported the presence of multilocular retroperitoneal anechoic cystic lesion with internal septations of variable thickness with echogenic soft tissue component; the lesion seen was occupying the abdominopelvic cavity, displacing and compressing the solid organs. The US findings were concerning for macrocystic abdominopelvic lymphatic malformation (Figure 1).

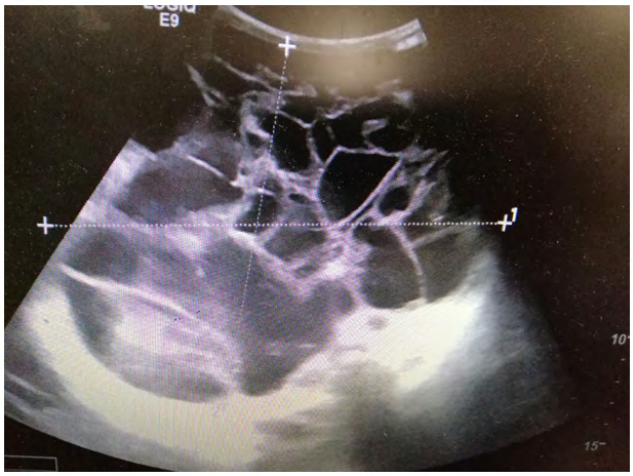

Figure 1: *Abdominal ultrasound scan showing cysts.*

Magnetic resonance imaging (MRI) of the abdomen was then performed to gather more accurate assessment of the mass origin and extent. It showed large multiloculated cystic lesion (highT2/highT2- SPIR signal intensity) with thin enhancing internal septations measuring about 13.6 x 11.1 x 7.5 cm occupying most of the abdominal cavity elevating the diaphragm, displacing and compressing the small bowel (which appear clumped anterio-superiorly) and the solid organs laterally, encasing the colon, and compressing the kidneys/IVC posteriorly. The lesion extends through the right inguinal canal into the right hemiscrotum. There was a concern for an enhancing component within the lesion just anterior to the right kidney. Overall MRI findings were concerning for large retroperitoneal lymphatic malformation with possible small venous malformation component.

Therapeutic approach

Since the mass was not well-demarcated to be eligible for complete resection and as the baby was clinically stable, enteral feeding was started attempting conservative management with watchful waiting. The baby tolerated enteral feeding very well, did not show any worsening abdominal distention or any new concerning physical signs and he was stooling and voiding regularly. The patient was kept in the hospital for 11 days then discharged home and parents were given a multi-disciplinary follow-up program.

Follow up and progress

On the 21st day of life, the patient presented for regular follow up at the pediatrics surgery clinic where he was noticed to have increased abdominal distension with no rigidity or tenderness. He was hemodynamically stable with no other clinical concerns. The baby was admitted to the hospital for work up and management. Abdominal US showed increased size of the cysts (16.1 x 13.1 cm). So, US-guided pigtail catheters were inserted into the lesion by interventional radiologist and a total of 300 ml of lymph was drained over six days. After stabilization, the baby was discharged home and scheduled for elective sclerotherapy.

At the age of 58 days, sclerotherapy was performed using a total of 1000 mg Doxycycline (10 capsules of 100 mg) mixed with 60 ml 0.9% NaCl together with 40 ml contrast media injected through a pigtail tube that confirmed good distribution of the sclerosing agent. Another similar injection was done on the next day. The baby tolerated the procedure very well and discharged home within few days.

At four months of age, the patient electively underwent laparotomy with excision of the mass from surrounding structures except part of the small bowel, with partial resection of the terminal ileum, cecum and appendix with ileo-colic anastomosis (Figure 2). Histopathologic report showed a completely excised cystic lymphangioma with foci of serosal foreign-body giant cell granulomas.

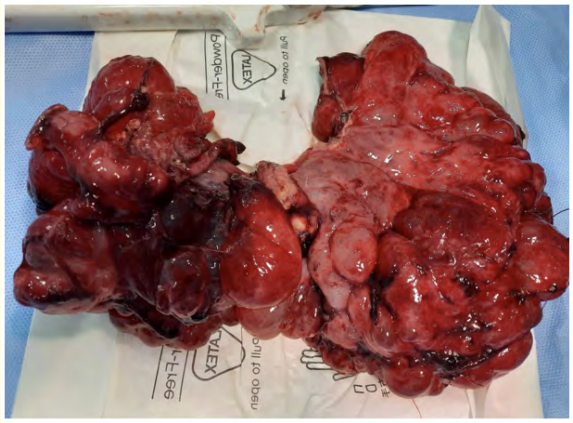

Figure 2: *Gross appearance of the resected mass.*

Currently, the patient is 19 months old, his weight is 12 kg. His growth and development are appropriate for his age. He is tolerating solid food intake with no concerns.

Discussion and Conclusion

Congenital Lymphatic malformations (CLM), known as lymphangiomas, are rare anomalies caused by a defect in the development of lymphatic structures and improper drainage into the venous system. They can affect any body organ and are mostly seen in the head and neck (> 75%) [1]. Abdominal lymphatic malformations (ALM) account for less than 5% of all CLM cases. ALMs are subcategorized into microcystic and macrocystic lesions [2,3].

The most common type of ALMs originates from the mesentery and the affected patients clinically present with intestinal obstruction, volvulus, acute cystic bleeding, or asymptomatic abdominal mass. Other types of ALMs arise from solid organs (liver, spleen, or pancreas) or from the gastrointestinal tract. Omental and retroperitoneal ALMs are the least common types that might not present until late childhood or even later [2,4].

The diagnosis and management of ALMs are challenging as they have a wide range of clinical presentation at different age groups, and complete surgical excision is not possible often times when the lesion is not well-demarcated which makes complete cure not easy to achieve [5].

ALMs can be diagnosed antenatally or postnatally. Antenatal detection rate is relatively higher in non-abdominal CLMs. Antenatal US sometimes cannot exclude intestinal obstruction or other abdominal cystic lesions [6]. Fetal US assesses the mass morphology and vascularity. It also helps screening for possible associated syndromes. At the same time, it is used to identify any possible complications such as intestinal obstruction, hydrops fetalis, and polyhydramnios [3].

In the majority of cases, the diagnosis is achieved postnatally as an incidental finding or when the affected patients develop intra-abdominal complications such as volvulus, cystic rupture, hemorrhage, or infection and as a result presented with abdominal mass, abdominal distention, abdominal pain, or feeding intolerance [7].

The final diagnosis of ALMs is confirmed by histopathology after surgical excision [8]. However, imaging studies are helpful in establishing the diagnosis and delineating the extent of the lesions before medical or surgical intervention. US is considered the best initial imaging study; it shows anechoic cystic spaces with or without echogenic debris. CT scan with contrast is the gold standard modality for evaluation of ALMs. CT images show the presence of multilocular septated masses with(out) hemorrhage and/or calcification. MRI does not provide more information than US or CT scan but is more preferred in neonates and children to avoid the radiation exposure encountered during CT scan [6].

Our case was antenatally suspicious for intestinal obstruction, but ALM diagnosis was established with postnatal US and MRI. Paolo Gasparella., *et al*. reported a case of giant ALMs in a 33-week preterm neonate that was diagnosed by fetal US at 18-week gestation and confirmed by postnatal US and MRI [9]. Sangho Lee., *et al*. reported 12 cases of ALMs diagnosed among children between 3 months and 17 years of age in the period 1999 to 2017. The primary presentation was abdominal pain, abdominal mass, and distention. All cases in their report were diagnosed postnatally by US or CT scan [4].

Currently, there is no standard approach regarding the optimal treatment modality. Conservative management is recommended for small or regressing and uncomplicated cases. Complete surgical excision is the best curative choice in well demarcated lesions. The proper timing of surgery is determined by the size of the mass, occurrence of complications, and the presenting symptoms among affected children. However, this may necessitate intestinal resection if the lesion is invading the bowel [1].

On the other hand, sclerotherapy is the best therapeutic modality for unresectable lesions that are not located within deep organs [3]. Sclerotherapy, which is reported to be safe and effective, is achieved by injecting sclerosing agents like OK 432, Doxycycline, or Bleomycin directly into the lesion. The best success rate is achieved in microcystic lesions, reported complications include pain at the injection site and local skin ulcerations [10,11]. Emerging systemic therapies have been used with good outcome especially when there is an element of vascular malformation, those medications include Propranolol, Sildenafil, and Sirolimus [12].

Olivera., *et al*. reported 5 prenatally diagnosed ALM cases from Canada in the period 2006 to 2008. One case spontaneously involuted, one treated with sclerotherapy only, one with sclerotherapy and surgical excision, and two cases by surgical excision only [13].

In our case, all therapeutic options were applied at different stages. Initially, conservative management with watchful waiting was attempted as the patient was clinically stable and tolerating feeds. Subsequently, the patient presented with worsening abdominal distention and had two pigtail tubes placed, the cyst was adequately drained with excellent recovery. After that, at the age of two months,

sclerotherapy was achieved using Doxycycline in an attempt to shrink the size of lesion and to make surgical excision possible. Finally, surgical excision was successfully achieved at the age of four months, partial ileal resection with end-to-end anastomosis was performed during the same procedure.

Similar approach was followed in the case reported by Gasparella., *et al*. where conservative management was initially attempted. Subsequently, the baby underwent emergent laparotomy and excision of the cyst with ileocecal resection due to intracystic hemorrhage that predisposed acute abdominal compartment syndrome at the age of two weeks [9].

Among the 12 cases reported by S. Lee., *et al*. seven cases underwent complete surgical excision of the mass with intestinal resection performed in 3 cases. Two patients had incomplete excision as the mass was extensive and invaded superior mesenteric vessels [4].

Incomplete resection is associated with a risk of recurrence in nearly 10% of cases [14]. S. Lee., *et al*. reported a risk of recurrence among one out the 12 included patients [4]. Our the patient is currently 19 months old with appropriate growth and development and no clinical concerns for recurrence, but he is scheduled to have a follow up MRI in the next few months.

In conclusion, ALMs are rare congenital anomalies that carry a significant risk of complications [9]. Diagnosis and management remain challenging. Conservative management with watchful waiting is recommended with close monitoring if the patient is stable. Sclerotherapy might be indicated as an exclusive therapy or as a bridge to surgical excision depending on the site and extent of the lesion. Surgical intervention with complete excision, if feasible, is the only curative intervention to relieve patients' symptoms and avoid or minimize the risk of recurrence.

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