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Case Report

Combined endolymphatic and surgical treatment of a leaking central conducting lymphatic malformation in a neonate

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ABSTRACT

Lymphatic malformations are congenital alterations of normal embryonic lymphatic development. We present a case of a premature 7-week-old male with a large central conducting lymphatic malformation and significant abdominal chylorrhea. He was successfully treated with combined endolymphatic and surgical approaches. To the authors' knowledge, this is the first case to be described.

Introduction

Lymphatic malformations (LMs) result from the abnormal embryologic development of the lymphatic system during fetal growth. Cystic LMs are the most common congenital manifestation of this entity and are often found in the head and neck with only 5%-10% occurring in the abdomen. However, an anomaly of the central conducting lymphatic system may also occur and can lead to impaired systemic lymphatic flow [3-5]. Resulting chylous hypertension can cause channel engorgement, disruption, and subsequent leakage into surrounding cavities [1-5]. Presenting symptoms are largely dependent on lesion location and severity, but in childhood central conducting lymphatic malformations (CCLM) tend to present acutely [4,5]. In the abdomen, clinical findings include distension, nau-

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Fig. 1 – Coronal image from contrast-enhanced abdominal computed tomography scan shows large ascites and displacement of abdominal viscera from mass effect.

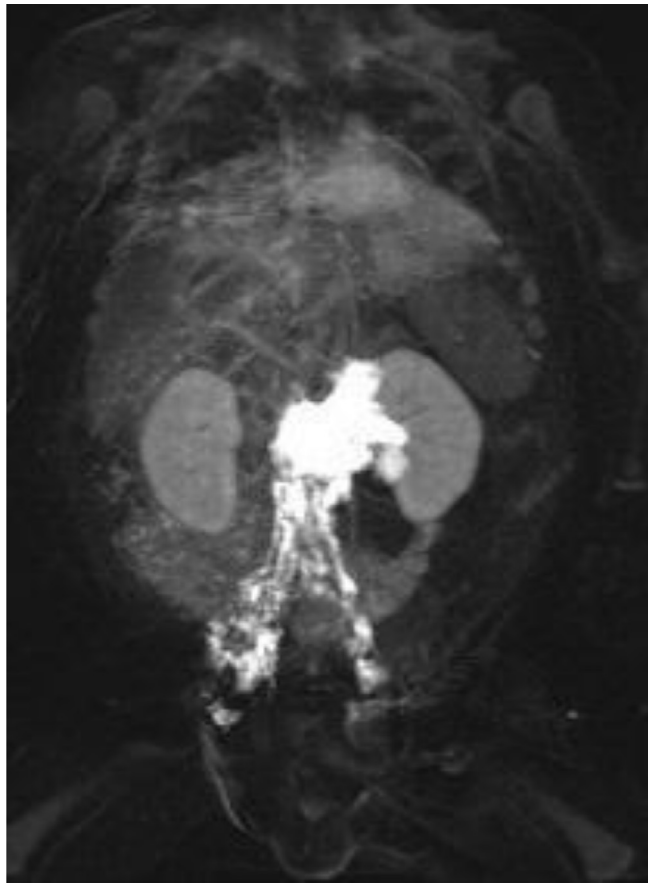

Fig. 2 – Coronal maximum intensity projections from dynamic contrast enhanced MR lymphangiogram shows large extravasation from disrupted central abdominal conducting lymphatic channels.

sea, vomiting, ascites, and respiratory compromise [1-5]. Diagnosis in the pediatric population may be made antenatally with ultrasonography [4]. Postnatal diagnostic confirmation may be challenging, but magnetic resonance lymphangiography can provide additional clarity [2,4]. Similar to LMs elsewhere in the body, CCLM may be managed conservatively, with embolization of lymphatic vessels or surgery reserved for medically refractory cases [1-4]. To our knowledge, this is the first reported case of a CCLM resulting in severe ascites and respiratory compromise in a neonate, managed with combined percutaneous n-BCA glue embolization and surgery.

Case report

We present a case of a male premature neonate born at 31 6/7 weeks, weighing 2.4 kg, with an antenatally diagnosed intra-abdominal cystic mass and hydrops. He was delivered via urgent C-section for fetal bradycardia and placental abruption. Ultrasound (US) at 1 week showed what was interpreted as an intact cystic LM. Parents noted worsening abdominal distention 3 weeks later. Subsequent US showed diffuse ascites. He was transferred to our institution at 7 weeks of age for abdominal distension, emesis, and decreased stool frequency. Repeat US showed septations and frond-like projections within the left upper quadrant ascitic fluid. Abdominal computed tomography showed large ascites (Fig. 1).

He was initially managed conservatively with parenteral nutrition, fluid resuscitation, octreotide and albumin infusions. He required serial large volume paracenteses for persistent distention (averaging 600-700 cc/procedure, max >1 L/day). At hospital day 28, approximately at 10 weeks of age, he became febrile with respiratory compromise requiring endotracheal intubation. Due to concern for abdominal compartment syndrome, a peritoneal drainage catheter was placed, and he was empirically started on intravenous antibiotics.

At 11 weeks old, dynamic magnetic resonance lymphangiography was performed via inguinal lymph node cannulation and injection of gadolinium, consisting primarily of T2-weighted and postcontrast dynamic T1-weighted imaging. This demonstrated a large dysplastic central abdominal lymphatic conducting channels with extensive intra- and retro-peritoneal extravasation (Fig. 2). Multiple lymphatic conducting abnormalities were also present, including: (1) large multiseptated LM in the upper peritoneal cavity with retroperitoneal and left renal hilar components connecting to the posterior mediastinum, infiltrating the pancreas and encasing major blood vessels; (2) dilated pelvic and retroperitoneal LM with contrast spillage at the lumbar region, possibly at the expected cisterna chyli, extending superiorly; and (3) early pelvic/lumbar venous plexus diffuse enhancement, suggesting direct lymphatic-venous connec-

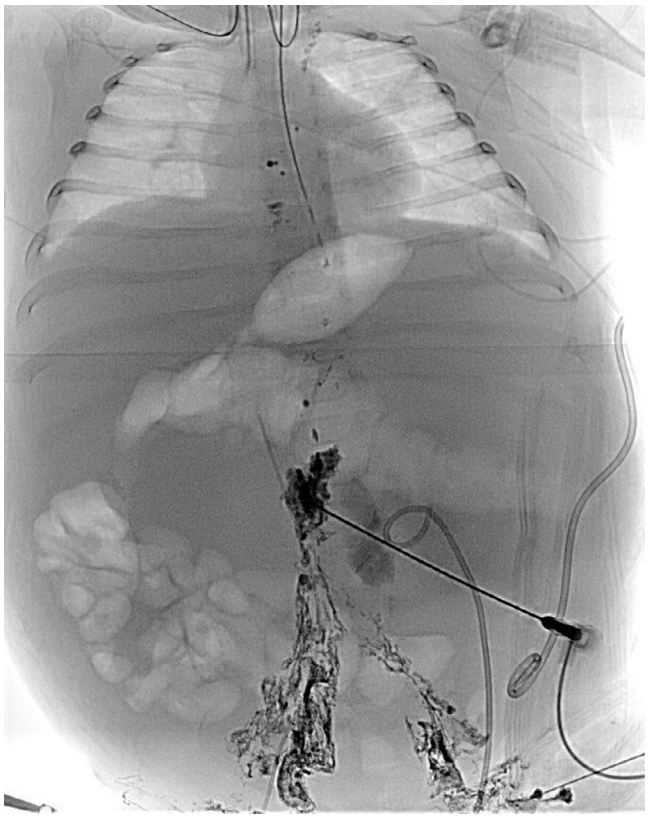

Fig. 3 – Conventional lymphangiogram showing the large central abdominal disruption and subsequent embolization with n-BCA glue using direct percutaneous access.

tion. Conventional lymphangiography with percutaneous n-BCA glue embolization (Histoacryl, B. Braun, Melsungen AG: Melsungen, Germany) was performed at age 13 weeks, targeting disrupted lymphatic channels (Fig. 3). By this point, the patient had a protracted course complicated by pleural effusions requiring chest tube placement, right adrenal hemorrhage, acute kidney injury, line-associated IVC thrombosis, and multimicrobial infections.

His peritoneal drain output dramatically improved to <300 cc/day until 15 weeks of age, when he again had significant output. A second lymphangiogram with percutaneous n-BCA glue embolization was attempted, but drain output (up to 2 L/day) was not reduced. With increasing large volume outputs and significant fluid weight gain, he ultimately underwent multiple surgeries starting at 16 weeks old with an exploratory laparotomy, lymphatic ligation, retroperitoneal dissection, and oversewing of multiple retroperitoneal lymphatic leaks. Extensively abnormal lymphatics of the descending and distal transverse colon were seen with associated cystic structures (Fig. 4). A partial colectomy was performed with temporary colostomy in order to control lymphatic leak from the colonic mesentery. Postoperatively, fluid losses progressively resolved and the patient was weaned from parenteral nutrition. After an uneventful but lengthy recovery, he was discharged at 6.5 months old.

Subsequent follow-ups showed appropriate developmental progress and weight gain. He has been feeding via a gastrojejunostomy tube without issue and gaining weight appropriately. His colostomy was reversed at 14 months. He developed a symptomatic anastomotic stricture soon after and underwent segmental resection and re-anastomosis. He subsequently recovered well and has not had any further complications. Follow-up at 1.5 years of age revealed he was without symptomatic recurrence. At 2 years old, his gastrojejunostomy tube was removed, and he was tolerating full oral nutrition.

Discussion

This report describes a case of a leaking abdominal CCLM in a neonatal boy treated with 2 percutaneous glue embolization procedures and multiple surgeries for control of severe lymphorrhea. The overall incidence of all LMs range considerably from 1 out of 2000 to 20,000 live births, which may be partially attributed to inconsistent definitions and terminology including lymphangiomas, cystic hygromas, etc [3,4]. Abdominal LMs account for roughly 5% of all LMs, most frequently found in the lymphatic-rich mesentery. The retroperitoneum, as in our patient, is the rarest accounting for 12%-14% of abdominal lymphatic malformations (ALMs) and less than 1% of all LMs. The second most common abdominal LM site is the omentum, with all three structures sharing a common embryologic origin [4].

LMs may be diagnosed sonographically in-utero as early as the second trimester. The appearance often is a multiloculated thin-walled cystic structure [3,4]. MR imaging may provide further information, characterized by hyperintense signal on fluid-weighted sequences, variable T1-weighted signal intensity based on serosanguinous or proteinaceous contents, and variable enhancing septations [2-4]. Central conducting lymphatic anomalies in the retroperitoneum, however, are poorly described in current literature [5]. Lymphangiectasia may be found, often with large, elongated lesions, that traverse adjacent anatomic regions [4]. If not captured through imaging, CCLMs may not be evident until significant acute symptoms occur, particularly from major vessel rupture/leakage or infection [3,4]. Severe ascites resulted in respiratory compromise for our patient, requiring serial paracenteses and intubation. Indolent growth manifests with nonspecific presentations such as abdominal pain, distension, nausea, vomiting, diarrhea, constipation, or possibly a palpable mass [1-5].

Differentiation of abdominal cystic LMs from other diagnoses is important. In pediatric patients, attempt at distinction from cystic neoplasms may depend on such factors as serologic tumor markers and suspicious imaging features like associated enhancing soft tissue nodularity. Cystic LMs can be distinguished from ascites by the presence of septa, compression on adjacent bowel loops, and absence of the position-dependent free fluid. Appearance and location may be undistinguishable from cysts of other organs, particularly enteric duplication and ovarian cysts [2,4].

The management of LMs begins with conservative measures, including dietary modifications/parenteral nutrition, somatostatin analog infusion, and drainage of fluid collec-

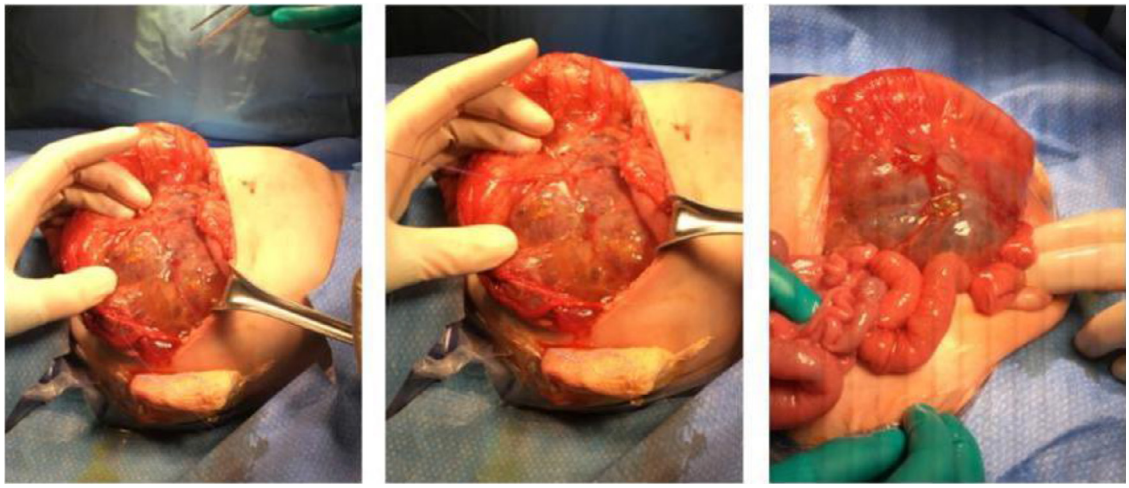

Fig. 4 – Intraoperative images taken during initial operative exploration. These photos demonstrate extensive mesenteric lymphatic abnormalities with cystic component in the descending colon mesentery.

tions [1,3]. Unfortunately, drainage is usually palliative and provides temporary relief. When these attempts fail, sclerotherapy is used for macrocystic lesions (using agents such as doxycycline, sodium tetradecylsulfate, bleomycin, OK-432, or sirolimus), but microcystic and CCLMs are frequently refractory to both medical and surgical treatments. Lymphangiography may identify a leak with subsequent embolization of faulty vessels or ducts, but leaks are often multifocal in nontraumatic cases [1-3]. Complications of lipiodol and glue embolization include pulmonary embolism of glue material, glue migration into systemic circulation via shunts, abdominal or lower extremity swelling, and chronic diarrhea [2,3]. Surgery, such as lymphovenous anastomosis or ligation, is reserved for advanced cases [1,3]. For all LMs combined, recurrence rate is 10%-40% after incomplete resection and 17% after macroscopic complete resection.

To the authors' knowledge, this is the first case of an abdominal CCLM resulting in severe ascites and respiratory compromise in a neonate. With only documented cases of cystic LM in the abdomen, our progressive approach for the management of central conducting anomalies is demonstrated with percutaneous n-BCA glue embolization and surgery. The case presented special challenges given the unique anatomy of a neonate and tools and strategies utilized in treating this rare pathophysiology.

Declaration of Competing Interest

None.

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