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Successful Glue Embolization of a Retroperitoneal Lymphatic Malformation Complicated with Refractory Chylous Ascites: A Case Report

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Abstract

A patient diagnosed with inoperable retroperitoneal lymphatic malformation presented with progressive abdominal distention secondary to chylous ascites. This was believed to be secondary to lymphatic leakage from the lymphatic malformation into the peritoneal cavity. The chylous ascites was refractory to conservative management and drainage by a peritoneal catheter. Intra-nodal embolization with lipiodol reduced the amount of daily drainage from 4 to 2 L/day. Two weeks later, the most inferior parts of the lymphatic malformation were punctured, and a 1:6 mixture of *n*-butyl cyanoacrylate (glue) and lipiodol was directly injected into the lesion. Subsequently, the daily drainage decreased to less than 10 mL/day and the patient's symptoms resolved.

Keywords: Ablation, cyanoacrylate, cystic hygroma, lipiodol, lymphangiography, lymphangioma

Introduction

Lymphatic malformation commonly occurs in the head and neck region with 5% of the cases seen in the abdomen.[1] Lymphatic malformations are mostly small and localized, but rarely they can be diffuse and may encase several vital structures deeming curative surgery impossible. Thus, percutaneous sclerotherapy plays an important role in the management of lymphatic malformation.

A case of a retroperitoneal lymphatic malformation complicated with chylous ascites that was refractory to conservative therapy is reported, but it responded well to intra-nodal lymphangiography and embolization with a mixture of lipiodol and *n*-butyl cyanoacrylate (glue). Institutional review board approval was waived on this retrospective case report.

Case Presentation

A 17-year-old female with a history of inoperable retroperitoneal lymphatic malformation presented with abdominal distention and discomfort that slowly progressed over a 2-week period.

On ultrasound, a large retroperitoneal thin septated multicystic lymphatic malformation, extending from the celiac axis down to the pelvis, was seen. New large volume intraperitoneal anechoic free fluid was seen. Analysis of the intraperitoneal fluid showed triglyceride concentration of 2508 mg/dL with abundant lymphocytes in keeping with chylous fluid. Computerized tomography (CT) was obtained to reevaluate the patient for possible surgical resection (Figure 1).

An 8F pigtail catheter was inserted to drain the ascites. Despite conservative management by stopping oral intake with the initiation of low fat, total parenteral nutrition, 4 L of fluid were drained daily.

Due to the failure of conservative treatment, intranodal lymphangiography was performed. Under ultrasound guidance, the inguinal lymph nodes were bilaterally identified and punctured using a 22G needle. After verifying the position of the needle tip at the nodal hilum, lipiodol was injected using an infusion pump at a rate of 0.5 mL/min, up to a total of 10 mL. Fluoroscopic surveillance was performed for 60 min (Figure 2). The site of extravasation could not be demonstrated on lymphangiography. However, over the first 24 h, fluid drainage decreased from 4 to 2 L/day.

After 2 weeks, direct embolization of the lymphatic malformation with a mixture of *n*-butyl cyanoacrylate and lipiodol (diluted in the ratio of 1:6) was performed (Figure 3). Under moderate sedation, using a 22G Chiba needle and under ultrasound guidance, a total of 7 mL of this mixture was injected

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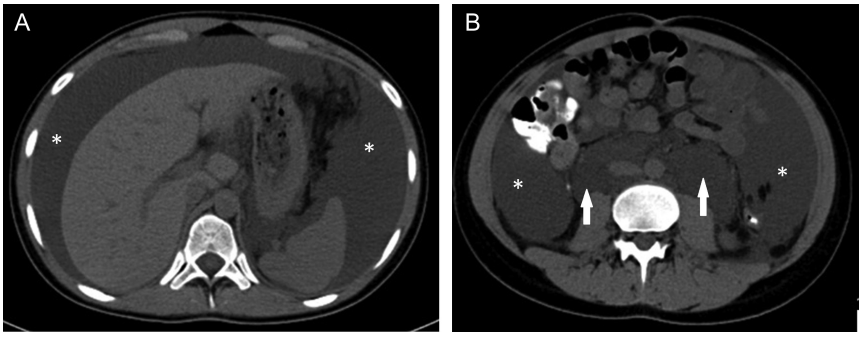

Figure 1. (A and B) Axial non-contrast CT images obtained at current presentation showing a large volume intraperitoneal fluid (asterisk) along with the retroperitoneal lymphatic malformation (arrow).

bilaterally in the most inferior components of the lymphatic malformation adjacent to the iliac arteries. The duration of the procedure was 10 min. Over the next 72 h, the peritoneal fluid drainage decreased from 2000 mL/day to less than 10 mL/day.

After the procedure, CT of the abdomen and pelvis showed scattered opaque droplets of lipiodol throughout the lymphatic malformation. The patient's abdominal distension resolved, and the pigtail was withdrawn 4 days after the second procedure. After 6 months of follow-up, the patient had no recurrence of abdominal distension and trace intraperitoneal fluid was seen on follow-up imaging (Figure 4).

Discussion

Most lymphatic malformations occur in the head and neck and less than 5% of them occur in the abdomen.[1] The older terms "lymphangioma" and "cystic hygroma" have been replaced by the terms "microcystic" and "macrocystic lymphatic malformation," respectively. In addition, a mixed form of lymphatic malformation also exists.[2] Intra-abdominal lymphatic malformations can present with pain, fever, abdominal distention from ascites, vomiting, and palpable mass.[3,4] Repeated paracentesis can provide symptomatic temporary relief, but reaccumulation of ascites usually occurs, and it is associated with electrolyte imbalance and hypoproteinemia. Death has also been described in this setting.[4]

Management of lymphatic malformation is challenging and conventionally it entails dietary modification for several weeks and surgery in refractory cases. Outcomes with traditional management are indefinite, and despite complete surgical resection recurrence occurs in about 12% of the cases. The recurrence rate could reach up to 53% when incomplete

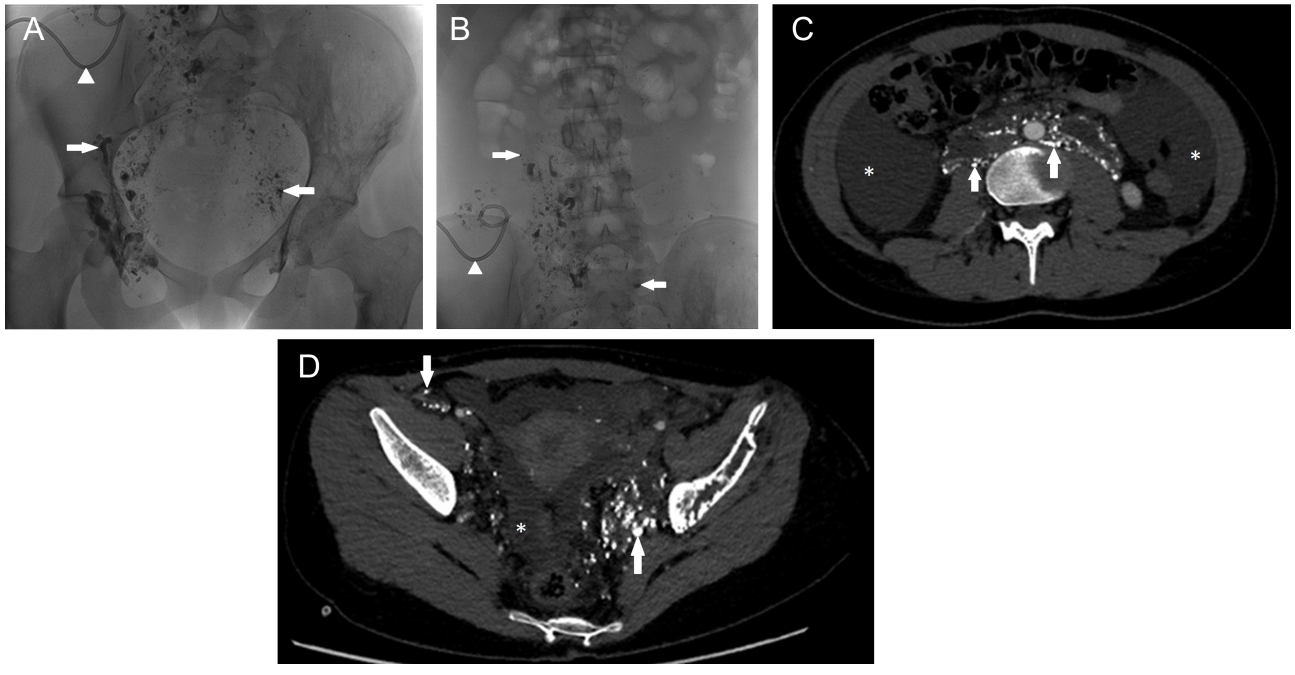

Figure 2. (A and B) Fluoroscopy images of the pelvis and abdomen obtained during intra-nodal lymphangiography. Opaque lipiodol (arrows) is seen scattered along the iliac and retroperitoneal lymphatic channels/lymphatic malformation. Lipiodol did not progress above the diaphragm. The site of lymphatic leakage could not be visualized. The pigtail catheter (arrowhead) can be seen in the right lower quadrant. (C and D) Non-contrast CT images obtained after lymphangiography showed lipiodol scattered throughout the retroperitoneal lymphatic malformation in the abdomen and pelvis. Intra-peritoneal fluid (asterisk) persisted after intra-nodal lymphangiography.

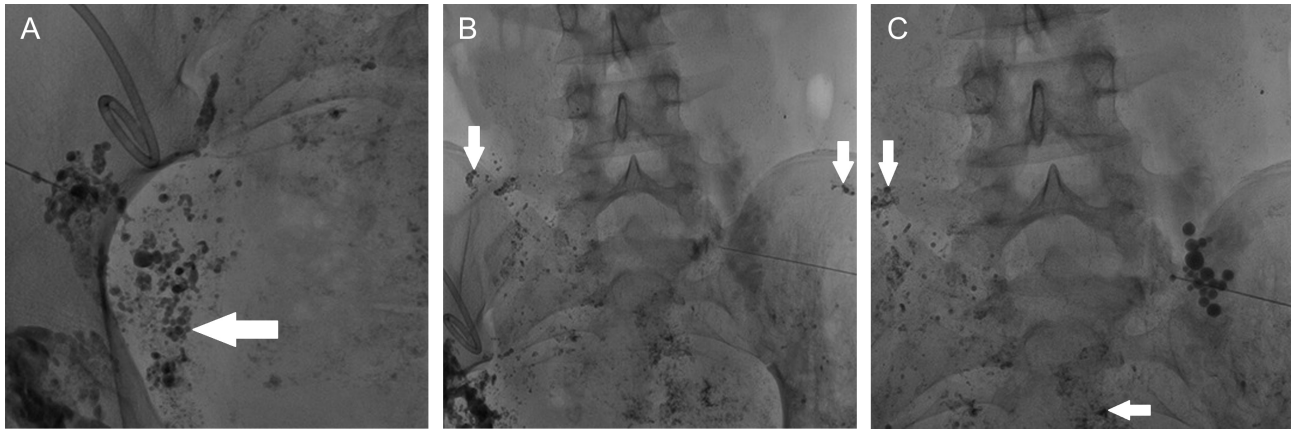

Figure 3. (A-C) Fluoroscopy images obtained during direct embolization of the lymphatic malformation with a mixture of lipiodol and glue (*n*-butyl cyanoacrylate). The most inferior components of the lymphatic malformation were identified with ultrasound, and they were punctured to inject the embolizing mixture. The opaque mixture (arrows) is seen scattered throughout the lymphatic malformation.

surgical resection is performed.[1] Over the last decades, several sclerotherapy modalities emerged as a feasible low-risk alternative which was shown to provide excellent/good clinical response in most cases of lymphatic malformation with a low incidence of complications. In addition, sclerotherapy was not found to complicate future surgery.[5]

Surgery is the most common cause of chylous ascites,[6] and the most common cause of atraumatic ascites is lymphatic anomalies (32%), which mainly include lymphangiectasia.[7] In one review, out of 190 cases of atraumatic chylous ascites only a single case was secondary to lymphatic malformation.[7]

In the current report, chylous ascites was believed to be secondary to leakage from the lymphatic malformation into the peritoneal cavity. Conservative management with dietary adjustment and percutaneous fluid drainage failed, which prompted the exploration of other treatment options.

Lymphangiography with lipiodol (ethiodized oil) is used as a diagnostic and therapeutic modality. It can show the site of lymphatic extravasation, and more importantly, it is an embolization procedure that can block the lymphatic structures causing the extravasation. It is thought that lipiodol causes a granulomatous and inflammatory reaction in the soft tissues at the site of extravasation, which can further help in closing the leak.[8] In a retrospective review of 31 patients, Nadolski et al. visualized the lymphatic leak site only in 17 (55%) of the cases on lymphangiography. In addition, spontaneous resolution of chylous ascites occurred in 7 out of 20 (35%) patients who underwent lymphangiography without any additional intervention.[9]

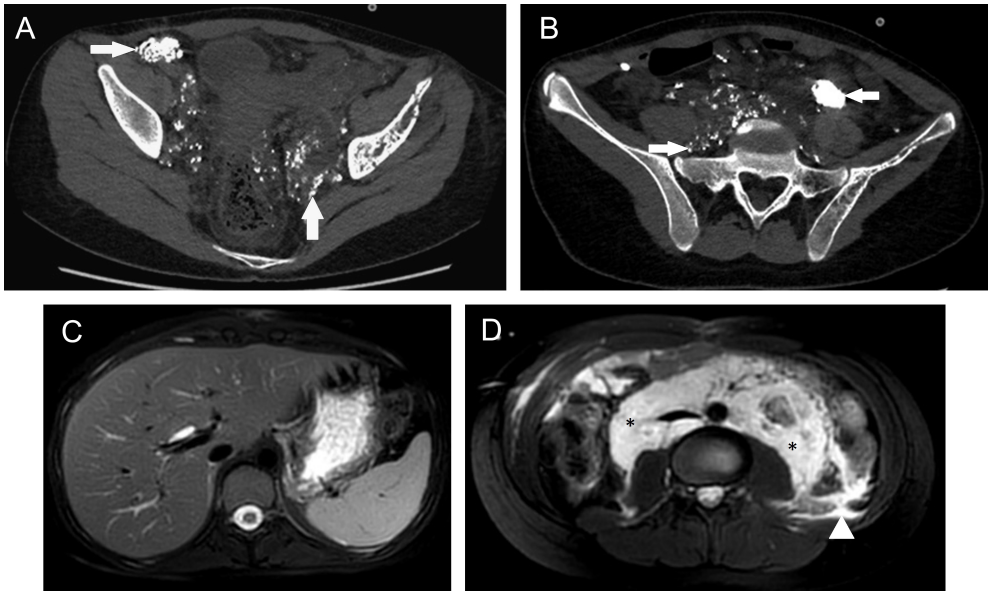

Figure 4. (A and B) CT images obtained after direct embolization of the lymphatic malformation with glue (n-butyl cyanoacrylate) and lipiodol mixture. The opaque mixture (arrows) is seen scattered throughout the retroperitoneal lymphatic malformation (asterisk). The intraperitoneal fluid has almost resolved completely. (C and D) Axial T2 weighted MRI images showing near total resolution of the intraperitoneal fluid (arrowhead).

In the present case, chylous drainage decreased from 4 to 2 L/day following intra-nodal lymphangiography with lipiodol. Since intra-nodal lymphangiography is time consuming and technically challenging, the authors chose to repeat the embolization by direct injection of the lymphatic malformation with a mixture of lipiodol and *n*-butyl cyanoacrylate. The amount of peritoneal fluid drained decreased dramatically within 24 h, and less than 10 mL of peritoneal fluid was drained over the next 24-72 h.

The interventional procedures described in this report were successful at stopping the lymphatic leak and were not intended to ablate the lymphatic malformation. The authors chose to puncture the most inferior part of the lymphatic malformation in the second procedure because they believed that the embolizing mixture would eventually travel superiorly throughout the lymphatic malformation along with the lymphatic flow, and eventually block the site of leakage.

No complication occurred secondary to this procedure. Resolution of the chylous ascites and the patient's symptoms were achieved despite the absence of a radiologically detectable leakage from the retroperitoneal lymphatic malformation.

Informed Consent: Informed consent is not necessary due to the retrospective nature of this study.

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