Development of intractable ascites due to thoracic duct hypertension

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ABSTRACT

We describe a 69-year-old dialysis-dependent patient who developed intractable ascites after zone 2 aortic reconstruction for a type IA thoracic endovascular aneurysm repair endoleak. Investigation as to the cause of ascites revealed a unique set of clinical circumstances leading to intractable bloody ascites. Investigation included imaging and invasive testing to diagnose the culprit mechanism. Ultimately, interventional catherization of the left subclavian vein illustrated an abnormally elevated pressure in the left subclavian vein. It was thus determined that, owing to the combination of a left brachiocephalic (innominate) vein occlusion after surgical ligation and in situ left brachiobasilic arteriovenous dialysis graft, there was overcirculation through the thoracic duct. Retrograde flow through the pop-off thoracic duct led to hematogenous ascites. Ligation of the left brachiobasilic arteriovenous dialysis graft resulted in near instantaneous and complete resolution of the ascites. (J Vasc Surg Cases and Innovative Techniques 2021;7:189-92.)

Keywords: Computed tomography; Dissection; Hypertension

CASE REPORT

Patient consent was obtained for publication of this case report. The patient is a 69-year-old man who was being evaluated for surgery of a persistent type IA thoracic endovascular aneurysm repair (TEVAR) endoleak. The patient's pertinent past medical history includes hypertension, dialysis-dependent end-stage renal disease, and atrial fibrillation. Nine years prior, at the time of index presentation, the patient had a developed an uncomplicated type B aortic dissection. He was taken to the operating room for a TEVAR for uncomplicated type B aortic dissection aneurysmal degeneration, which was complicated by retrograde type A dissection. This retrograde type A dissection was surgically repaired with a valve-sparing root replacement and ascending hemiarch.

He was followed with routine surveillance for 9 years with yearly computed tomography scans. After this index operation, he developed end-stage renal disease and underwent hemodialysis through a left upper extremity brachiobasilic arteriovenous (AV) graft. This graft was functioning well through the surveillance period. Unfortunately, during the surveillance period he slowly developed a type IA endoleak with increasing distal aortic arch diameters measuring 6.1 cm × 5.1 cm. It was deemed medically necessary to address the type IA endoleak. He was taken to the operating room for a two-stage operation.

First, he underwent a successful left carotid subclavian artery bypass to provide adequate landing zone for the distal aspect of the future zone II arch graft. Second, he was taken to the operating room for a surgical repair of the proximal type IA endoleak. This was performed by initiating peripheral cardiopulmonary bypass through the right axillary artery and femoral vein. A median sternotomy was performed and after cooling to 22°, the aorta was cross-clamped, and the old Dacron graft was transected. A combination of retrograde cardioplegia and direct ostial cardioplegia was used to arrest the heart. Deep hypothermic circulatory arrest was commenced and anterograde cerebral perfusion at 7 to 10 mL/kg/min was performed. It was found that the edge of the TEVAR graft had eroded into the distal edge of the hemiarch graft and that this was the source of the type IA endoleak with flow in the residual false lumen. First the metal of the old TEVAR graft was resected. A zone 2 arch reconstruction with distal anastomosis to the resected clean edge of the prior TEVAR graft was performed using felt—clean resected TEVAR edge—felt to new zone II graft as a sandwich with running 3-0 Prolene. The distal aspect of the TEVAR was left intact. The left carotid was connected to the zone 2 graft using a hybrid 9-mm Gore graft and the innominate artery was directly anastomosed to the zone 2 graft. The proximal extent of the zone 2 aortic arch graft was then anastomosed to the old Dacron graft from the initial valve-sparing root replacement. Of note, during dissection and access through the redo sternotomy, the left innominate vein was ligated to expose the ascending aorta and arch vessels.

The postoperative course was complicated by prolonged requirement of inotropic and vasopressor support and

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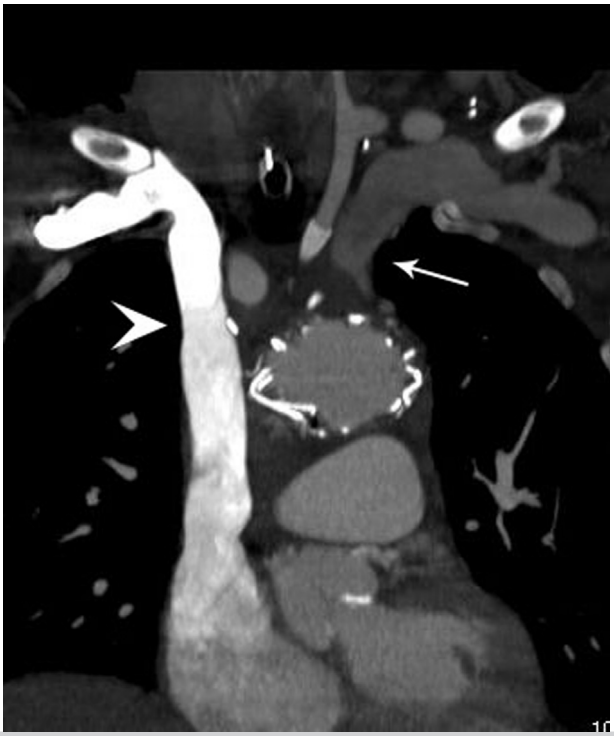

Fig 1. Computed tomography angiography reconstruction, demonstrating complete obstruction of the left brachiocephalic vein (*arrow*). Note patent right brachiocephalic vein and SVC (*arrowhead*).

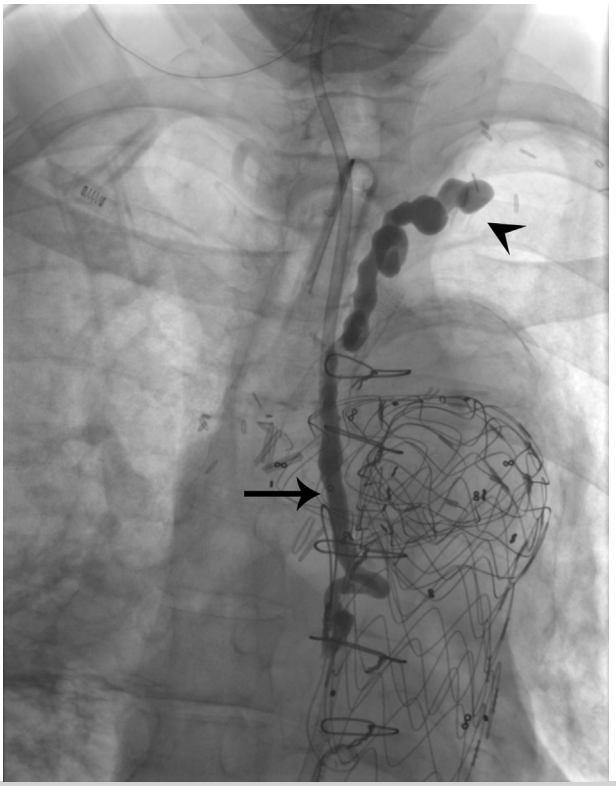

Fig 2. Fluoroscopy image of the injection of the contrast in the dilated and tortuous thoracic duct (TD; *arrow*). The distal part of the TD (*arrowhead*) is functionally obstructed by elevated central venous flow.

continuous renal replacement therapy. A tracheostomy was placed because of the continuous need for ventilator support. A protracted course in the intensive care unit revolved around a profound, prolonged refractory-to-treatment complex ascites that required paracentesis of 3 L several times a week.

Initially, the ascites was attributed to volume overload, malnutrition, and his critical illness. Physical examination revealed a patient who was cachectic with tense ascites that resolved with paracentesis and reaccumulated every 5 days. There was a well-healed sternal scar and a tracheostomy in place. A palpable thrill was appreciated in the AV graft and no major left upper extremity or facial edema was appreciated. Cytology from the paracentesis was negative for malignancy. Culture of the ascites was also persistently negative. Ascites chemistry revealed red blood cells consistently above 10,000/μL with several series well above 1,000,000 red blood cells/μL, albumin level between 1.4 and 1.7 g/dL, fluid triglycerides of 30 to 170 mg/dL, lactate dehydrogenase of approximately 70 μ/L, and glucose of 100 mg/dL.

Ultrasound examination demonstrated a nodular contour of the liver surface suspicious for liver cirrhosis; however, the transhepatic venous gradient was 1 to 2 mm Hg, which was not consistent with portal hypertension and a hepatic etiology of the ascites. Right heart catheterization and pulmonary wedge pressure revealed a right atrial pressure of 9 mm Hg with a ventricularized waveform consistent with tricuspid regurgitation and reduced RV compliance, mild pulmonary hypertension (pulmonary artery mean pressure of 29 mm Hg) with a high but normal pulmonary capillary wedge pressure (13 mm Hg) and normal pulmonary vascular resistance. Systemic vascular resistance was low (6 Wood units) and cardiac output was elevated (11.7 L/min; cardiac index, 6.75 L/min/m^2) consistent with a high output state. Transthoracic echocardiography demonstrated an ejection fraction estimated at 60% with mild tricuspid regurgitation. Computed tomography angiography of the chest revealed an occluded left innominate vein (Fig 1).

Owing to the slight elevation of the triglycerides in the ascitic fluid, chylous ascites was considered.

The patient underwent bilateral groin lymphangiography and thoracic duct (TD) catheterization that demonstrated dilation and tortuosity of the TD (Fig 2). The pressure in the TD was measured and found to be 21 mm Hg. The left subclavian vein was cannulated and the same mean pressure was measured in the left subclavian vein, where the TD drained. The pressure in the superior vena cava was measured and found to be 5 mm Hg. It was therefore suspected that the reason for the increased pressure in the left subclavian vein was due to surgical ligation of the left innominate vein, during entry and exposure of the type IA endoleak repair, and the overcirculation of blood secondary to the presence of the left upper arm brachiobasilic AV graft. It was decided that this increased pressure and

circulation in the TD, which was acting as a low pressure retrograde pop-off conduit, was the cause of the bloody ascites.

Recanalization of the left innominate vein was deemed unfeasible owing to terminal surgical ligation of the vein. Rerouting of the TD to the right-sided venous circulation was considered to be technically challenging owing to the prior left carotid subclavian bypass and in situ tracheostomy. It was decided to ligate the brachiobasilic AV graft to decrease the pressure in the left subclavian vein. This was done surgically with a small incision over the AV graft and suture ligation. Simultaneously, a temporary right internal jugular tunneled dialysis line was placed. The ascites resolved immediately after surgery. The sequelae of the increased abdominal pressure leading to ventilator dependency also resolved and the patient was rapidly weaned from the ventilator in 48 hours. The patient was then transferred to the floor and subsequently to a rehabilitation facility within the same week of ligation of the AV graft after 6 months in the ICU.

DISCUSSION

In this case report, we describe a unique situation where the combination of the occlusion of the left innominate vein and presence of a left arm brachiobasilic AV dialysis graft caused a significant increase of the pressure in the subclavian vein, thus impeding lymphatic flow from the TD, resulting in the development of intractable ascites. Ligation of the graft brought about resolution of the symptoms. The differential diagnosis of the development of intractable ascites includes liver cirrhosis, hepatic veno-occlusive disease, volume overload, heart failure-related ascites, chylous ascites from surgical disruption of the lymphatic channels, cancer-related ascites, infection-related ascites, ascites, nephrotic syndrome, pancreatitis, and anatomic causes.

The main function of the lymphatic system in the body is to remove fluid from the interstitium back into venous circulation. When the capacity of the lymphatic system reaches a threshold, the fluid starts to accumulate in the tissues and cavities.[1] One of the first theories of the development of ascites in patients with congestive heart failure and liver cirrhosis was a failure of the lymphatic system.[1] The relationship between increased pressure in the TD and symptoms in patients with heart failure was first suggested by Earnest Starling.[2] In his seminal work "The Arris and Bale lectures on some points in the pathology of heart disease" he stated: "It seems probable that the obstruction to the flow of lymph from the thoracic duct into the blood, as well as the distension of the duct from the largely increased lymph flow from the liver, may contribute to the production of oedema in the rest of the body."[3] The Starling hypothesis was further bolstered by experimental and clinical data over the next half a century. External, surgical decompression of the TD in patients with acute exacerbation of congestive heart failure and ascites in liver cirrhosis was performed, resulting in resolution of the symptoms in all patients within hours.[4]

Allan Dumont proposed that the flow capacity of the TD is limited by the lymphovenous connection in the distal part of the TD, which is a cause of ascites in patients with liver cirrhosis.[5] Several groups reported improvement of ascites in approximately 50% patients with liver cirrhosis after surgical TD-venous anastomosis.[6-8] To further test the concept of TD decompression, Cole et al[9] anastomosed the TD to the pulmonary vein in dogs with experimental heart failure, induced by a combination of surgically created pulmonary stenosis and tricuspid insufficiency, thus decompressing the TD into a lower pressure circulation. They demonstrated a significant decrease in ascites in the anastomosis group compared with controls. In an attempt to decompress the lymphatic system in single-ventricle patients with Fontan circulation complicated by pleural effusions and protein-losing enteropathy, which are known lymphatic complications, Victor Hraska, anastomosed TD to the right atrium, resulting in resolution of the symptoms in both patients.[10]

TD access is performed as previously described and ultrasound guidance is used to directly access bilateral inguinal lymph nodes using a 25G spinal needle, connected to the small connection tubing (BD Medical, Franklin Lakes, New Jersey, NJ).[11] After accessing the hilum of the lymph node, the position of the needle is confirmed by injecting an oil-based contrast agent (Lipiodol, Guerbert, Villepinte, France) using fluoroscopic guidance. When the cisterna chyli is identified, it is accessed transabdominally with a 22G, 15- to 20-cm Chiba needle (Cook Medical, Bloomington, Ind). As double wall penetration of the duct is achieved, a stiff 0.018-in. guidewire (V18 Control, Boston Scientific, Natick, Mass) is engaged to probe for the TD. Successful access of the TD is confirmed by easy advancement of the wire into the upper thorax. A 3F, 65-cm microcatheter (Renegade, Boston Scientific, Marlborough, Mass) is advanced over the wire into the TD. Using a 3-mL syringe, water-soluble iodinated contrast is injected into the TD to confirm the position.

This experimental and clinical evidence of the relationship between the pressure in the TD and development of ascites supported our decision to ligate the dialysis graft to decrease the pressure in the left subclavian vein and TD and bring about the resolution of the ascites. After a 6-month ICU stay, the patient was discharged to a rehabilitation facility. He was ventilator liberated, requiring 2 L of O_2, ambulating more than 100 m, and ascites free.

This unique clinical scenario, however, can have a broader significance and application for the treatment of ascites in patients with heart failure and liver cirrhosis. The lymphatic system is one of the main components of the fluid metabolism in the body and often overlooked in the evaluation of the patients with ascites and/or edema owing to a lack of readily available diagnostic techniques. New lymphatic imaging and interventional

techniques can provide a better understanding of the pathophysiologic mechanisms attributable to the lymphatic circulation, which will in turn assist in developing novel treatment options for these patients.

This case highlights the role of interventional diagnostic catheterization of the left subclavian vein and the TD to evaluate the pressure gradients in the TD. Early assessment of the TD pressures can be instrumental in the diagnosis of TD hypertension. Although an invasive procedure, this case highlights the advantages of TD pressure monitoring. Increased pressure in the TD can result in the development of intractable ascites. Novel lymphatic imaging and minimally invasive lymphatic interventional techniques can be used to evaluate and intervene on the lymphatic system in patients with cardiovascular diseases. Furthermore, interventional techniques of ligation and embolization can also be used for TD pathologies if necessary. Although this case highlights that left brachiobasilic AV graft ligation in the setting of left innominate stenosis or obstruction can relieve TD pressure, our patient was left with a temporary solution of a tunneled dialysis line. Right upper extremity and lower extremity AV access options were considered for long-term dialysis access. After deliberation, the patient was schedule for an outpatient right upper extremity HeRO graft owing to his prior central stenosis. Peritoneal dialysis was considered a suboptimal option owing to the prior recurrent ascites and failed peritoneovenous shunt. Although not an optimal solution, the resolution of refractory bloody ascites and weaning from ventilator dependency with hospital discharge resulted in an optimal outcome for the patient, who remains hospitalization free after discharge.

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