Surgical management of bilateral thoracic outlet lymphaticovenous malformations causing recurrent cerebrovascular accidents

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ABSTRACT

Lymphaticovenous malformations (LVMs) are a rare subset of congenital vascular malformations that result from the defective development of the vascular and lymphatic systems during embryogenesis. LVMs can cause pathological mass effects or lead to thrombotic complications. We present a rare case of the surgical management of bilateral LVMs arising at the junction of the brachiocephalic and internal jugular veins in a patient with a patent foramen ovale, identifying the source of previously unexplained paradoxical cerebrovascular accidents. (J Vasc Surg Cases Innov Tech 2022;8:429-32.)

Keywords: Congenital vascular malformation; Lymphaticovenous malformation; Extratruncular lesion; Hamburg Classification

Congenital vascular malformations (CVMs) are a wide spectrum of rare structures created by interrupted embryogenesis. They may range from minor lesions to major disfiguring anomalies.[1] The Hamburg Classification System classifies CVMs based on their vascular component.[2,3] They are further subdivided as truncular or extra-truncular lesions based on embryologic developmental stage and by hemodynamic status.[2,4] These categories guide optimal surgical management.[1]

We present a rare case of bilateral lymphaticovenous malformations (LVMs) arising near the junction of the internal jugular veins and brachiocephalic vessels leading to thrombotic complications. The patient consented to publication of his case details and images.

CASE REPORT

A 33-year-old man with a history of HIV, patent foramen ovale (PFO), and multiple imaging-proven cerebrovascular accidents (CVAs) without residual deficits presented to an outside hospital with left-sided neck pain. Ultrasound examination demonstrated a 4.9 × 2.6-cm left-sided complex neck mass that was concerning for malignancy. The patient was transferred to our institution for further workup and management. On examination, he had a small, soft, palpable, and nonpulsatile mass just above the medial aspect of the left clavicle, along the lateral aspect of his neck. The patient had no history of trauma or vascular interventions. He had a history of tobacco use and intravenous drug use (IVDU), although he had been abstinent for 1 year. He reported compliance with antiretroviral therapy, but his absolute CD4 count was 168 cells/mL.

Duplex ultrasound examination and computed tomography angiography and venography showed bilateral masses at the confluence of the internal jugular and brachiocephalic veins (Fig 1), as well as a small saccular distal abdominal aortic aneurysm. Both masses seemed to be venous pseudoaneurysms without infiltration of surrounding tissue, but on ultrasound examination were noted to have additional connections. Given the unusual presentation, possible etiologies including connective tissue disorders, CVM, and infectious processes related to HIV or IVDU were discussed. Workup for other etiologies was negative and no other source of CVAs was found. This finding, along with evidence of thrombus in the malformations on ultrasound examination and his known PFO, made paradoxical emboli the favored etiology for his CVAs. He was subsequently started on therapeutic low-molecular weight heparin and discharged home with close outpatient follow-up.

Two weeks later, he returned to our vascular surgery clinic. Discussion of management options included observation with anticoagulation and endovascular intervention with the possible need for repeat procedures and the risk of thromboembolic events between interventions. The patient elected to undergo surgical resection, which he felt was the most definitive. One week later, he was admitted preoperatively and transitioned from low-molecular weight heparin to a heparin drip that was continued through the operation.

In the operating room, the patient underwent a median sternotomy and bilateral neck exploration to ensure adequate vascular control and exposure. The overlying muscles and thymic tissue were mobilized and divided. The anterior surface

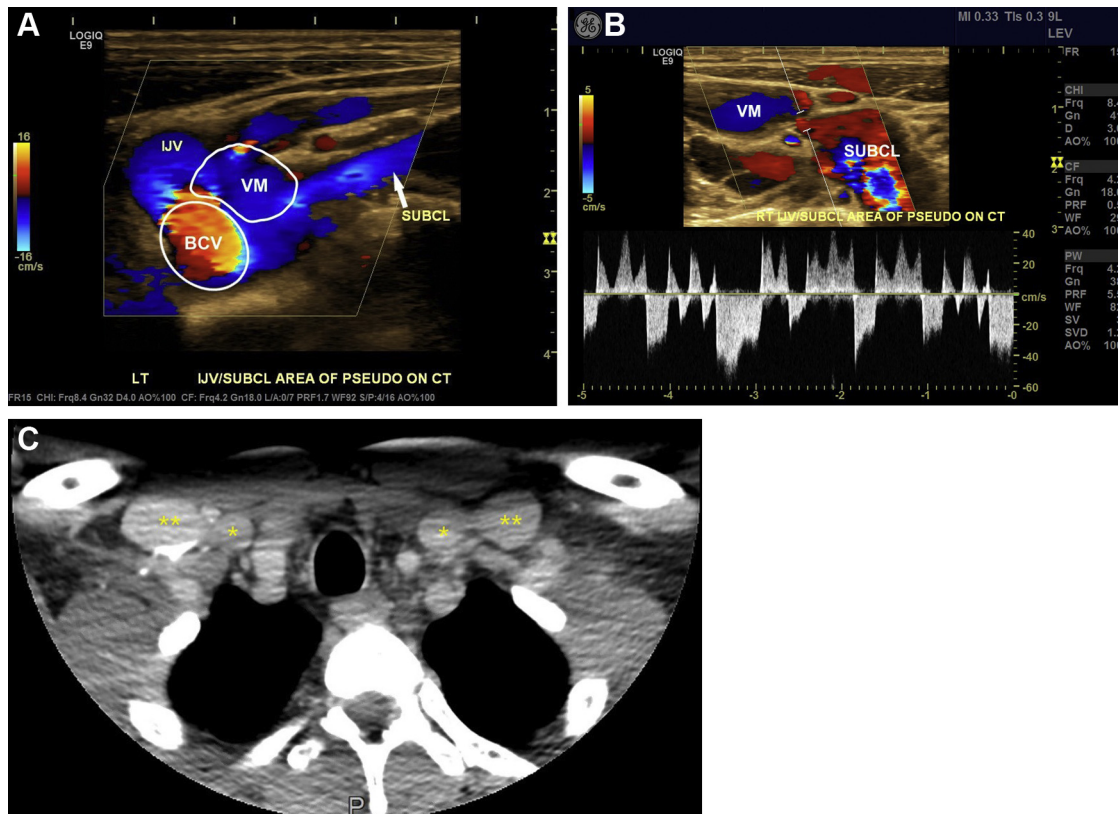https://doi.org/10.1016/j.jvscit.2022.06.010

Fig 1. (A) Duplex ultrasound image of the base of the left side of the neck. The suspected vascular malformation (*VM*) with a feeding vessel is seen at the confluence of the internal jugular vein (*IJV*) and subclavian vein (*SUBCL*, white arrow). Brachiocephalic vein (*BCV*). **(B)** Duplex ultrasound image of the base of the right side of the neck. Suspected VM. **(C)** Computed tomography (*CT*) venogram. The double asterisks mark the bilateral vascular malformations. The single asterisks mark the bilateral internal jugular veins.

of the left innominate vein was dissected. Near the junction of the left subclavian and internal jugular vein, the lesion was identified just lateral and posterior to the left internal jugular vein (Fig 2). A supraclavicular neck incision was made in continuity with the sternal incision for further dissection. The left-sided malformation was found to have multiple venous connections to the brachiocephalic and internal jugular veins and multiple lymphatic connections. The thoracic duct was not identified definitively or preserved. When the venous connections were ligated, the malformation turned from a normal venous-appearing color to white/yellow, which was consistent with presence of a significant lymphatic component. The remaining lymphatic connections were ligated, and the mass was removed. No significant surrounding fibrosis, adhesions, or inflammation were found, supporting the diagnosis of a congenital LVM. The right-sided malformation was dissected in a similar manner, but lacked a lymphatic component. Given the well-defined stalks connecting the masses to the major veins, venous reconstruction was not necessary. Each mass was approximately 6 cm in maximal diameter. Both were opened on the back table and found to contain copious fresh thrombus. Two flat Jackson-Pratt drains were placed, intravenous protamine was given, and the incisions were closed in multiple layers. The estimated blood loss was 250 mL.

The patient was monitored in the intensive care unit. Deep vein thrombosis prophylaxis began on postoperative day 1. Drains were removed by postoperative day 3. Full anticoagulation was restarted on postoperative day 4, and the patient was discharged home on postoperative day 6. He returned to vascular surgery clinic 3 weeks postoperatively. He was recovering well without complication, will continue therapeutic anticoagulation until planned PFO closure, and stop anticoagulation thereafter. The final pathology findings were consistent with LVM without evidence of an infectious or inflammatory etiology.

DISCUSSION

LVMs are a rare subset of CVMs composed of both lymphatic and venous components, commonly located in the neck and axilla.[5] The management of LVMs is highly dependent on location, associated symptoms, and classification.[6] Compared with truncular malformations, which directly affect the main vessels, extratruncular malformations are more variable in presentation and tissue infiltration. Because they originate from defective vascular development at the reticular stage of embryogenesis, extratruncular malformations possess mesenchymal cell characteristics. Under stimulatory
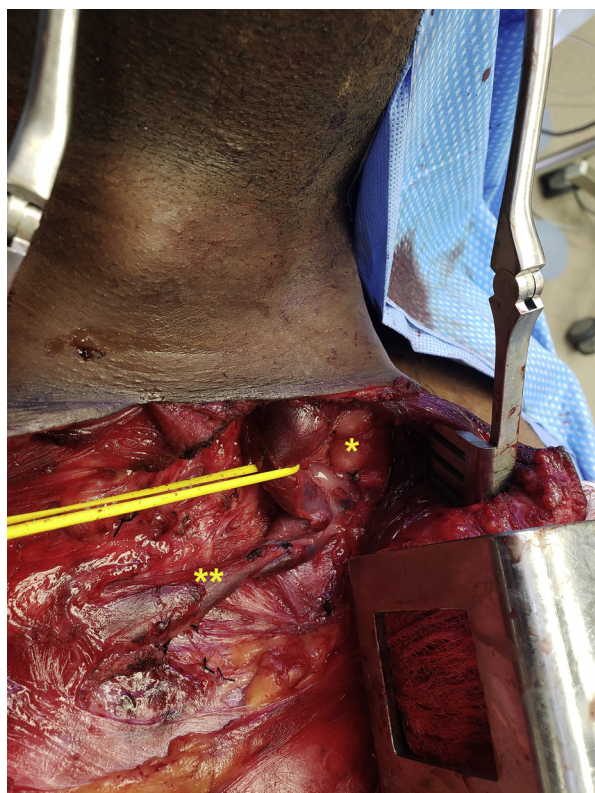
Fig 2. Intraoperative left side vascular malformation exposure. The yellow vessel loop marks the left internal jugular vein. The single yellow asterisk marks the vascular malformation that is white/yellow after ligation of venous connections. The double yellow asterisks mark the left brachiocephalic vein.

conditions, these malformations grow in parallel with the patient and may recur if incompletely resected.[1,7,8]

Depending on risk or symptom severity, CVMs may be treated with a step-up approach from symptom control, to anticoagulation, to procedural or surgical intervention.[9,10] Although endovascular intervention is possible, it is often not curative and is preferred for surgically inaccessible lesions or as an adjunct, particularly when malformations are infiltrating and involve nearby structures.[1,9,11,12] Densely associated tissue growth may require staged debulking with vascular reconstruction in such cases.[1,9,13] Thus, shared decision-making with the patient is paramount.

In our patient's case, we felt that endovascular embolization, sclerotherapy, or a stent may not adequately ablate or cover all connecting structures. And considering his PFO and prior CVAs, he preferred a full resection rather than risk interim thromboembolic events should an endovascular procedure later fail. Although the initial diagnoses were atypical-appearing jugular pseudoaneurysms, given our intraoperative findings of numerous bilateral venous and left-sided lymphatic connections to

a saccular structure, we concluded that our patient had bilateral limited extratruncular CVMs containing formed thrombus. Because his diagnosis was made intraoperatively, the risk of recurrence was discussed after resection.

We hypothesized that emboli were showering from his vascular lesions into his arterial circulation through the PFO, resulting in multiple ischemic CVAs. Preoperatively this factor was discussed with our cardiology colleagues, and we decided to first perform surgical resection followed by PFO closure later.

Other studies describing a similar pathology and presentation are few, although it is known that patients with PFOs can experience paradoxical emboli leading to CVAs.[11,14] We considered potential infectious etiologies given our patient's HIV status, low CD4 count, and prior IVDU, but the workup was unremarkable. The possibility of a connective tissue disorder was considered given the presence of a small, incidentally found aortic aneurysm, but ultimately considered unlikely.

As the literature suggests, anticoagulation was started at the time of presumed diagnosis and continued through the operation.[1,9] Although the postoperative risks for this patient included stroke, deep vein thrombosis/PE, and arm swelling, he experienced no complications and remains without apparent recurrence.

CONCLUSIONS

LVMs are rare vascular malformations that range in disease severity, size, symptoms, and location. Embryological characteristics are among the many factors that must be considered during interventional planning. The best surgical approach must be individualized and a multidisciplinary approach to management and treatment should be considered.

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