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MINI-FOCUS ISSUE ON CARDIAC MASSES

CLINICAL CASE

Surgical Management of Fibrosing Mediastinitis Presenting as Superior Vena Cava Syndrome

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ABSTRACT

Fibrosing mediastinitis is a rare chronic inflammatory condition characterized by excessive fibrotic process in the mediastinum. Although various infectious processes are more common causes, it can also be related to several malignancies. We report a case of a 28-year-old woman with fibrosing mediastinitis related to an aggressive primary gray-zone lymphoma causing complete occlusion of the superior vena cava (SVC) and the innominate veins. She underwent several unsuccessful attempts at stenting and angioplasty. Due to worsening symptoms of SVC syndrome, she was ultimately treated with surgical reconstruction of the SVC and innominate veins using the Y-synthetic graft technique. A high degree of clinical suspicion should be maintained to diagnose fibrosing mediastinitis in onco-cardiovascular patients. With careful patient selection, review of techniques and imaging, surgical reconstruction may be a viable strategy for patients with SVC syndrome secondary to malignancy who have exhausted endovascular options. (JACC Case Rep. 2025;30:102854)

Fibrosing mediastinitis (FM) is a rare, chronic inflammatory complication of various infectious processes including histoplasmosis.[1] The excessive proliferation and infiltration of acellular collagen and fibrotic lesions can cause obstruction of the SVC, with the syndrome estimated to occur in 19% of FM cases.[2] Although FM has been shown to mimic a malignancy,[3] we describe a novel case of SVC syndrome caused by FM secondary to mediastinal gray-zone lymphoma (MGZL), an aggressive subtype of lymphoma with features of both Hodgkin's and primary B-cell lymphomas.[4]

HISTORY OF PRESENTATION

A 28-year-old woman presented to our institution with worsening facial swelling, shortness of breath,

TAKE-HOME MESSAGES

- This case illustrates the importance of maintaining a high clinical suspicion for FM secondary to a primary malignancy leading to SVC syndrome.
- Surgical reconstruction of the SVC and innominate veins can be achieved successfully in carefully selected patients with extensive tumor-vessel involvement who had unsuccessfully undergone other systemic and endovascular approaches.
- In high-risk onco-cardiovascular patients, a careful, multidisciplinary approach should be used to optimize the patient's quality of life and minimize postoperative complications.

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ABBREVIATIONS AND ACRONYMS

FM = fibrosing mediastinitis

GVHD = graft-versus-host disease

PBSCT = peripheral blood stem cell transplant

MGZL = mediastinal gray-zone lymphoma

SVC = superior vena cava

ocular angioedema, blurry vision, and unstable gait concerning for exacerbated compression of SVC. On admission, her heart rate was 105 beats/min, respiratory rate of 20 breaths/min, and blood pressure was 143/90 mm Hg. Her pulse oxygen saturation was 95% on room air but she reported feeling uncomfortable with neck swelling when lying flat, which raised concerns about impending airway compromise. She reported feeling an "impending sense of doom" and intermittent headaches.

PAST MEDICAL HISTORY

Two years before presentation, she had a diffuse hilar mass, which seemed to have originated from mediastinum. This was later diagnosed as diffuse mediastinal B-cell lymphoma. She was promptly treated with 6 cycles of rituximab, etoposide phosphate, prednisone, vincristine sulfate, cyclophosphamide, and doxorubicin hydrochloride (R-EPOCH); however, within 1 year of completing chemotherapy, she developed relapsed primary MGZL. Four months before presentation, she received a peripheral blood stem cell transplant (PBSCT), which was complicated by graft-versus-host disease (GVHD) in the guts. She underwent several unsuccessful attempts at endovascular recanalization and stenting due to significant occlusions of the SVC and innominate veins.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis included MGZL, previous catheter-associated thrombosis, and mediastinitis. While her SVC syndrome may be secondary to chemotherapy port-related strictures, preoperative imaging suggests that the diffuse fibrosis may have had a direct compressive effect on the SVC.

INVESTIGATIONS

Diagnostic venogram demonstrated no discrete communication between bilateral innominate veins and the SVC connecting to the right atrium (**Figures 1A to 1C**). Using radiofrequency wires, multiple attempts, both superiorly and inferiorly, were made to recanalize the innominate veins and the SVC without success. Her contrast-enhanced computed tomography angiography confirmed total occlusion of the proximal SVC and a prominent 3.2 × 2.2-cm mass on the anterior aspect of the proximal SVC (**Figures 2A and 2B**). The patient underwent positron emission tomography-computed tomography, which revealed low anterior mediastinal mass tracer activity, suggesting the mass was likely not an active tumor. Preoperative and intraoperative transesophageal echocardiograms revealed normal heart function with no intracardiac shunts.

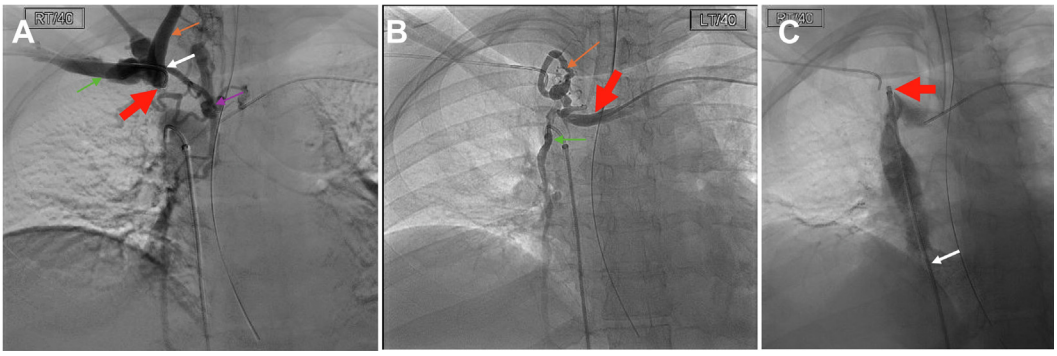
MANAGEMENT

Given these impressive symptoms, she underwent reconstruction of the SVC. The chest was accessed via median sternotomy. With notable tumor necrosis, the anterior mediastinal mass had completely occluded flow in the innominate vein closest to the SVC junction (**Figure 3A**). However, the lumen could be visualized at the level of the confluence of the right internal jugular and right subclavian vein (**Figure 3B**). She was systematically heparinized before applying a partially occluding clamp on the SVC. We reconstructed the left innominate vein via end-to-end anastomosis in an open fashion with a 14-mm ringed fluorinated ethylene propylene Gore-Tex graft (Gore Medical). Heavy pressurized bleeding could be observed when the occluding lesions were cleared from the SVC (Video 1). We then performed a graftotomy to anastomose an 8-mm ringed polytetrafluorethylene Gore-Tex graft (Gore Medical) to the SVC, establishing a Y-shaped venous drainage into the right atrium (**Figures 1C and 2C**). Adequate flow through the prosthetic grafts were confirmed by Dopplers intraoperatively. Pathological examination of the excised mediastinal mass revealed dense fibrosis.

DISCUSSION

The goal of managing malignant SVC syndrome remains twofold: alleviate obstructive symptoms and treat the underlying malignancy. In the absence of randomized controlled trials, there are currently no evidence-based guidelines regarding the management of complex, malignancy-related SVC syndrome. In general, our multidisciplinary strategy begins with considering chemotherapy and endovascular approaches as first-line treatments in severely symptomatic patients with chronic malignant SVC syndrome.[1,5] This is especially true should there be any threat to possible airway compromise. However, our patient had previously undergone unsuccessful rounds of systemic chemotherapy and attempts at endovenous recanalization with stenting to no avail. Notably, radiation therapy is no longer considered a first-line treatment option in potentially life-threatening malignant SVC syndrome, particularly as this can hinder histological analysis of the primary tumor if performed before biopsy.[5] Indeed, open-lumen surgical approaches do not represent the gold

FIGURE 1 **Diagnostic Contrast-Enhanced (Omnipaque 350) Venography Still Images of the Patient at Admission**

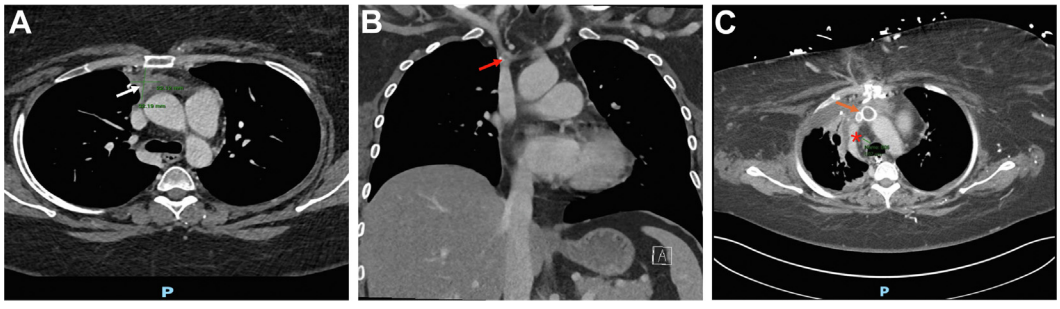


(A) Preoperative image demonstrates abrupt cutoff of right innominate vein (red arrow) without discrete communication with the superior vena cava or left brachiocephalic vein. Green arrow shows contrast-enhanced right subclavian vein. Orange arrow shows contrast-enhanced right internal jugular vein. Purple arrow shows collateral veins. White arrow shows guidewire that was introduced superiorly from the right brachial vein. (B) Preoperative image demonstrates the origin of multiple collateral veins (orange arrow) arising from the central left innominate vein (red arrow), which communicate with the azygous vein (green arrow). (C) Preoperative image demonstrates abrupt cutoff of the lower superior vena cava (red arrow) without discrete communication with bilateral brachiocephalic veins (white arrows indicate guidewire).

standard in these cases, even among hemodynamically stable patients. However, it may be reserved for cases of extensive occlusion or high risk of venous thrombosis. In particular, when the occlusion involves the confluence of the innominate veins, an open-lumen reconstruction of the SVC bifurcation may be warranted.[5,6] Furthermore, in patients who may require local resection of the lesions, surgical bypass may be necessary.

There have been series of cases describing a range of potentially suitable techniques, albeit not exclusively for malignant causes of SVC syndrome. In cases with >50% vessel wall involvement without evidence of occlusion, the SVC may be repaired using pericardial or bovine patches without the need for cross-clamping to a cardiopulmonary bypass machine.[6,7] In more complex cases with tumor invasion greater than one-half of the total SVC length, total graft reconstruction should be considered.[8] Synthetic grafts are attractive choices for most because of their variety of sizes, flexibility, and ease of modifications. Polytetrafluorethylene grafts may be preferred with

FIGURE 2 **Contrast-Enhanced Computed Tomography Images of the Patient With Fibrosing Mediastinal Mass Related to Primary Mediastinal Gray-Zone Lymphoma Presenting as Superior Vena Cava Syndrome**



(A) Preoperative axial section demonstrates approximately 3.2 × 2.2-cm anterior mediastinal mass (white arrow). (B) Preoperative coronal section demonstrates prominent stricture at the confluence of right and left innominate veins (red arrow). (C) Two-month postoperative axial section demonstrates Y-shaped Gore-Tex vascular grafts (orange arrow) and enlarged 1.1-cm high paratracheal lymph node (asterisk).

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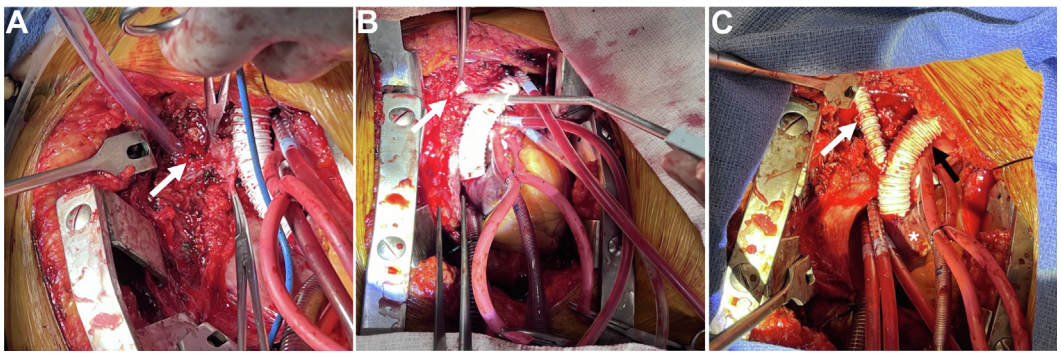
FIGURE 3 Intraoperative Findings



(A) Image demonstrates diffuse fibrosis causing occlusion of venous flow from the superior vena cava (white arrow) to the left innominate vein. (B) The lumen of the superior vena cava (white arrow) is visualized and checked thoroughly. (C) Image shows Y-shaped reconstructed left innominate vein (fluorinated ethylene propylene graft; black arrow) and superior vena cava (polytetrafluorethylene graft; white arrow) establishing drainage into the right atrium (white asterisk).

up to 95% rate of 1-year patency in patients with malignant causes of SVC syndrome.[9] The decision making regarding the size of the graft merits further discussion. The SVC normally experiences high flow at relatively low pressures compared with its arterial counterparts. Therefore, although the reported average diameter is between 18 and 20 mm, the size of the graft should be carefully selected to match the patient's anatomical features to decrease the risk of flow restrictions and to prevent future thrombotic events.[8]

Akin to our case, when tumor invasion extends to both innominate veins and the SVC, the Y-graft technique may help to establish multiple, larger routes of venous drainage into the right atrium. More commonly, the left innominate vein is replaced before ligating and reconstructing the right innominate vein with the Y-graft to the SVC conduit.[6] We began by first reconstructing the left innominate vein leading down to the right atrial appendage. This allowed us to remove the obstruction from the SVC and examine the site of vein-graft anastomosis while maintaining right-sided drainage before performing a graft-to-graft anastomosis to the existing larger left innominate vein conduit. Surgical reconstruction has also been successfully performed after failed attempts at stenting or angioplasty in a patient with post-radiation fibrosis. Thus, given the inherent risks, surgery may be reserved for complicated or failed endovascular stenting.[10] Taken together, we highlight the importance of identifying the optimal patient population that may reap the most benefit from a surgical intervention. These patients should be symptomatic, yet hemodynamically stable and contraindicated for alternative endovascular or systemic approaches. Although several techniques have been described, the choice of surgical approach is largely dependent on the surgeon's expertise and preference. Surgical reconstruction of the SVC and innominate veins can be achieved using the Y-graft in patients with complete FM and multi-vein involvement.

FOLLOW-UP

Because of the high risk of thrombus, she required anticoagulation on Coumadin 5 mg with goal international normalized ratio between 2 and 3. Her international normalized ratio at discharge was 2.93. The postoperative course was notable for hypoxia inherent to the procedure for which she required high-flow O_2 and aggressive diuresis in the hospital. She was discharged on postoperative day 15 after being weaned from supplemental O_2 and transitioning to oral Lasix 40 mg twice daily. Her creatinine at discharge was 1.6 mg/dL. She noted significant improvements in facial and generalized edema. However, her subsequent recovery alternating between rehabilitation and acute care facilities was challenging. Although she regained most of her function, she continued to experience unexplained left arm pain. Three days after admission to the rehabilitation facility, she had worsening dyspnea that required a tracheostomy. She also developed kidney injuries that were medically managed. More seriously, she presented with hematochezia 3 months after her

surgery. Subsequent esophagogastroduodenoscopy and flexible sigmoidoscopy-guided biopsies revealed worsening GVHD from her prior PBSCT. Percutaneous endoscopic gastrostomy tube was placed to aid with her nutrition. However, she developed further gastric perforations requiring exploratory laparotomy and significant wound complications leading up to her death 5 months after presentation.

CONCLUSIONS

Given the progressive onset of symptoms, a high degree of clinical suspicion should be maintained to consider FM as the underlying etiology of SVC syndrome. Surgical reconstruction of the veins in the mediastinum, although challenging, may be a viable treatment for select patients with extensive compression and infiltration of the SVC due to an aggressive malignancy. It is important to consider the impact of this procedure on the patient's quality of life. In our case, most of the major complications appeared to have been a sequela of her underlying malignancy and GVHD due to PBSCT. We highlight the need for careful, multidisciplinary approach in complex onco-cardiovascular patient populations. Thus, this case serves as a cautionary tale of the risks and benefits of SVC reconstruction in patients with mediastinal malignancies.

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APPENDIX For a supplemental video, please see the online version of this paper.