

case report

Superior vena cava stenting in IgG4-associated mediastinal fibrosis

Mohammed Khalid,^a Ihab Weheba,^{a,b} Abeer Abdelsayed,^{a,c} Leena Mohammad Zeitouni,^d Sarfraz Saleemi,^a Eid Al Mutaury,^d Syed Hassan^e

From the ^aDepartment of Medicine, King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia; ^bDepartment of Pulmonary Medicine, National Research Centre, Cairo, Egypt, ^cPulmonary Department, Faculty of Medicine, Ain Shams University, Cairo, Egypt, ^dCollege of Medicine, King Saud University, Riyadh, Saudi Arabia, ^eDepartment of Radiology, Johns Hopkins, Aramco Medical City, Saudi Arabia

Correspondence: Dr. Ihab Weheba · Pulmonary Medicine, National Research Centre, Cairo 12622, Egypt · ehabmokhtar@hotmail.com · ORCID: <https://orcid.org/0000-0003-4078-4869>

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We report a rare case of IgG4-associated mediastinal fibrosis with complete superior vena cava (SVC) obstruction successfully managed by thrombolysis and stenting in a 33-year-old male. The patient presented with a mediastinal mass lesion with clinical findings of SVC obstruction. Surgical biopsy of the mediastinal mass lesion with histology and immunohistochemistry staining established the diagnosis of IgG4 associated mediastinal fibrosis. The patient was treated with a systemic steroid and rituximab, but despite treatment, SVC obstruction and thromboses persisted, surgical intervention was declined by the thoracic surgeon due to extensive mediastinal fibrosis and an expected poor outcome. Percutaneous SVC angioplasty, intravascular thrombolysis with tissue plasminogen activator and afterward stent placement was done by the interventional radiology service. This intervention is rare and possibly was lifesaving as it restored complete patency of the SVC. Our case is probably the first with IgG4 mediastinitis and SVC complete obstruction relieved by intravascular thrombolysis and SVC stent placement. It demonstrates that SVC stenting can relieve SVC obstruction in patients with a high risk of surgery either due to medical comorbidities or an expected high surgical risk like bleeding in the mediastinal fibrosis, which in our case of SVC obstruction was due to a nonoperable mediastinal tumor.

SIMILAR CASES PUBLISHED: None to our knowledge.

Immunoglobulin G4-related disease (IgG4-RD) is an increasingly identified immune-mediated condition involved in a collection of multiorgan disorders that share certain pathologic, serologic, and clinical features. The hallmarks IgG4-RD are dense lymphoplasmacytic infiltrations with a predominance of IgG4-positive plasma cells in the affected tissue, associated with a degree of fibrosis that has a distinguished “storiform” pattern and is often characterized by obliterative phlebitis and an increased number of eosinophils with elevated serum concentrations of IgG4. Glucocorticoids are the main line of treatment with various courses of duration and relapse. Reported lung manifestations include solid nodules, bronchovascular thickening, alveolar interstitial with honeycombing, diffuse ground-glass opacities and pleural thickening. Mediastinal fibrosis is a rare presentation of IgG4 disease.¹⁻³

Superior vena cava thrombosis (SVC) was first described in 1912.⁴ Mediastinal compression syndrome due to SVC thrombosis is rare and

potentially fatal.⁵ It is usually associated with trauma to the SVC vein, coagulopathies, advanced malignancies as part of a paraneoplastic syndrome and direct compression of SVC by mediastinal tumors.^{6,7} The SVC may thrombose due to compression in diffuse mediastinal fibrosis as well, where maintaining SVC patency could be difficult.⁸ The aim of anticoagulation therapy for SVC thrombosis is to inhibit further thrombus formation and prevent embolization.⁹ Only a few case reports in the literature review the use of thrombolysis either systemic or intravascular to establish patency of SVC.¹⁰ We present a rare case of IgG4-related mediastinal fibrosis causing SVC thrombosis and stricture, conventional medical therapy with steroids, rituximab and anticoagulation therapy to relieve the mechanical obstruction and associated thrombosis. Due to extensive mediastinal fibrosis, surgical intervention was considered technically difficult to restore SVC patency. Intravascular catheter thrombolysis and placement of SVC stent resulted in complete patency of SVC.

CASE

We report a rare case of a 33-year old man who presented with mediastinal compression syndrome. His condition started in 2009 when he presented with a 3-month history of progressive exertional dyspnea. He was found to have constrictive pericarditis and underwent total pericardiectomy.

Pericardial histology showed evidence of granuloma but AFB stain, polymerase chain reaction and tissue cul-

ture for mycobacterial tuberculosis were negative. He was given empirical anti-TB treatment for nine months. Four years later, he presented with progressive shortness of breath, an engorged neck vein with neck and facial swelling. A chest X-ray showed speculated right hilar mass and right upper lobe infiltrates.

A contrast CT upper body and head showed extensive thrombosis of internal jugular veins bilaterally which extended superiorly to the sigmoidal sinus and inferiorly to the SVC. In addition there was worsening of mediastinal fibrosis causing SVC stricture and obstruction. A follow-up surgical biopsy of the hilar mass and CT-guided core biopsy of the paravertebral mass showed inflammatory infiltrates with background fibrosis (**Figure 1**); immunohistochemistry staining indicated IgG4-related disease as the cause of fibrosing mediastinitis (**Figure 1**). The patient was started on an IV heparin infusion. In view of worsening mediastinitis, he was given steroids and initiated on rituximab, but the recommended therapy failed to relieve the SVC obstruction.¹¹ Surgical intervention to relieve SVC obstruction was declined due to a possible poor outcome in view of the extensive fibrosis. An interventional radiologist was consulted and we reached a consensus to intervene using recanalization.

Recanalization procedure

The right basilic vein was punctured and a venogram showed total occlusion of the SVC with significant collaterals. The left brachial vein was punctured and the veno-

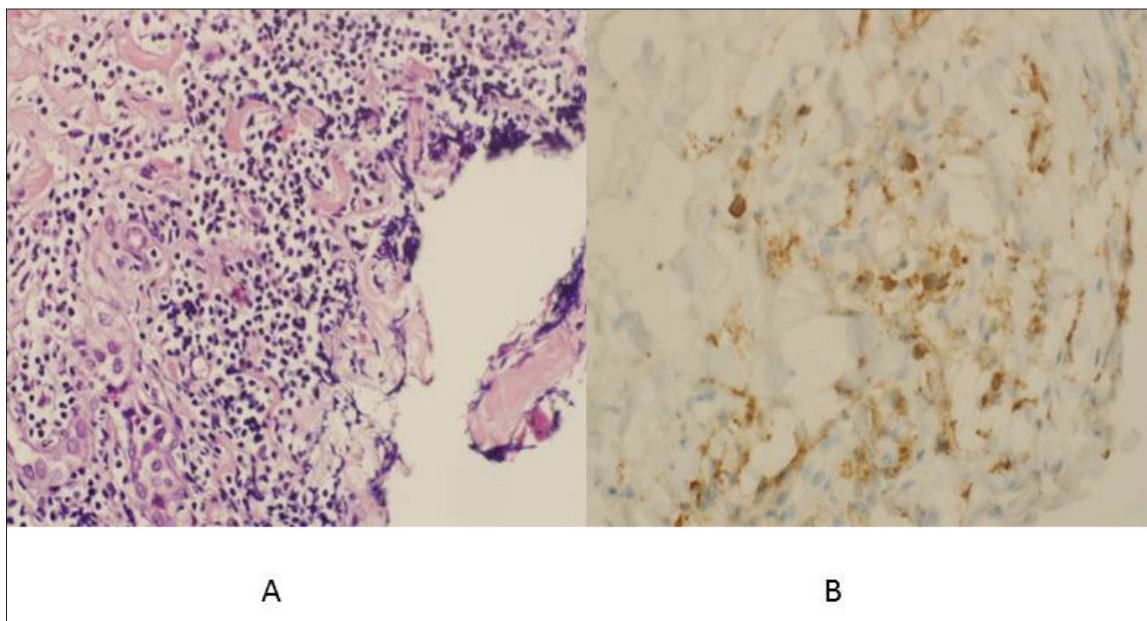


Figure 1. Lung biopsy showing IgG4-related mediastinal fibrosis (A) and immunohistochemistry (B).

gram showed an occluded left innominate vein with significant collaterals. Using a combined angled catheter and Terumo wire, both side occlusions were successfully crossed to the SVC and IVC, the Terumo wire was exchanged with the exchange Rosen. Serial balloon dilatation and post angioplasty venogram showed patent veins with multiple filling defects. After that the Asprix was used throughout the occluded portions. The postthrombectomy venogram showed persistent filling defects. Over a guidewire bilateral overlapping 10 cm effusion catheters were placed, and the patient underwent infusion thrombolysis over 24-hours (25 mg TPA in 500 mL of normal saline on each side) along with continuous heparin infusion. The next day, the contrast showed improvement of the thrombus with the remaining filling defects. An AngioJet thrombectomy device from both sides was used and postthrombectomy angiograms showed improvement of the thrombosis with residual filling defects. Bilateral kissing stents were placed and a poststenting venogram showed significant improvement of the flow with resolution of the collaterals (**Figures 2, 3**).

DISCUSSION

Multiple case reports exist in the literature of SVC thrombosis and obstruction. Intravascular catheter thrombolysis and, in some case reports, placement of an intravenous (IV) stent could resolve patient symptoms. Our case is probably the first case report of IgG4-related mediastinal fibrosis causing SVC thrombosis and stricture relieved by intravascular thrombolysis and SVC stent placement

In our case, the patient had extensive mediastinal fibrosis secondary to mediastinal inflammation with almost complete obstruction and thrombosis of the SVC vein. Medical therapy with steroids and rituximab failed to establish the patency, and the thoracic surgery team declined to operate on the patient fearing a poor outcome. The interventional radiology service was able to successfully do percutaneous angioplasty, thrombolysis and stent placement with complete reversal of vascular obstruction.

In conclusion, IgG4-related mediastinal fibrosis is rare, surgical correction for SVC obstruction where thrombosis could be difficult, A simple IV thrombolysis and placement of SVC stent resolved our patient symptoms. In cases with mediastinal involvement of non-operable tumors and mediastinal fibrosis in high-risk

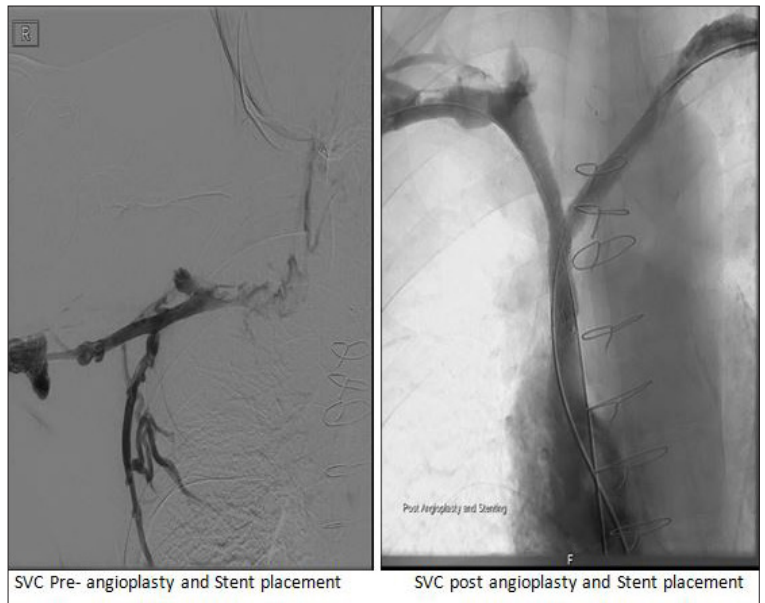


Figure 2. SVC angioplasty and stenting.

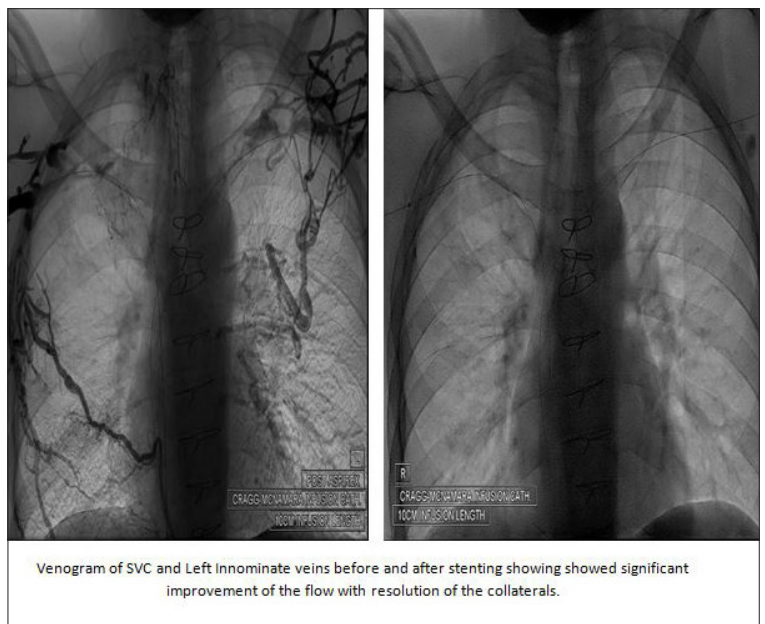


Figure 3. Venogram of SVC and left innominate veins before and after stenting showed significant improvement of the flow with resolution of the collaterals.

patients with comorbidities causing SVC obstruction, a minimal procedure associated morbidity of IV thrombolysis and placement of SVC stent could provide an excellent palliative care without major surgical intervention.

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