



Giant Ruptured Azygos Vein Aneurysm in a 33-Year-Old Woman

Murat Uzdenov¹  Johannes Kroll¹ Fatos Ballazhi¹ Martin Czerny¹

¹Department of Cardiovascular Surgery, University Hospital Freiburg, Freiburg, Germany

Address for correspondence DR. MED. Murat Uzdenov, Department of Cardiovascular Surgery, University Hospital of Freiburg, Hugstetter Str. 55, 79106 Freiburg, Germany
(e-mail: murat.uzdenov@uniklinik-freiburg.de).

Thorac Cardiovasc Surg Rep 2023;12:e36–e40.

Abstract

Keywords

- aneurysm
- angiogenesis
- imaging
- venous disease

The rupture of an azygos vein aneurysm is a very rare but catastrophic complication. Careful differential diagnosis of acute dyspnea and thoracic pain in young patients is essential for effective and early management. We present the case of a young woman with a huge, spontaneously ruptured vena azygos saccular aneurysm, successfully repaired via median sternotomy under cardiopulmonary bypass.

Introduction

Azygos vein and superior vena cava (SVC) aneurysms are extremely rare phenomena, with fewer than 40 cases reported in the indexed English literature.¹ In 1963, Abbott and Leigh introduced their classification of cava aneurysms and divided them into four groups (congenital, acquired, pseudoaneurysm, and arteriovenous aneurysm).² They can be solitary or coexist with cystic hygromas and angiomas (because of the similar embryologic origins of the venous and lymphatic systems). The cause is usually unknown.³ An azygos vein aneurysm's rupture is a very rare but catastrophic complication. Acute dyspnea and thoracic pain in young patients must be differentially diagnosed to ensure its early and effective management. In this report, we describe a case of a young woman with a huge spontaneously ruptured vena azygos saccular aneurysm successfully corrected via median sternotomy under cardiopulmonary bypass.

Clinical Summary

A 33-year-old woman with no medical history was admitted suffering from acute, severe right-sided chest pain and

progressive dyspnea, which she experienced after lifting her 2-year-old son. She denied any history of chest trauma, thoracic surgery, or central venous access. Physical examination revealed no significant findings. There was no clinical or laboratory evidence of acute infection. She had undergone chest radiography that showed a new right-sided pleural effusion (► **Fig. 1**). Her transthoracic echocardiography was unremarkable, showing good systolic biventricular function and no intracardiac abnormalities. After chest tube insertion and removing 700 mL of hemorrhagic fluid, the effusion regressed completely and her right lung was successfully re-expanded. The patient remained hemodynamically stable without any hemoglobin drop. Her 3D chest computed tomography (CT) revealed a saccular azygos vein aneurysm measuring 7.7 cm × 5.8 cm (transverse × craniocaudal) without thrombus (► **Fig. 2**). This mass was constricting the right lung. There was no evidence of arterial inflow. The mass was in contact with the anterolateral chest cavity wall and adjoining the superior mediastinum.

After diagnosing a huge, complicated azygos vein aneurysm, we considered open surgery after discussion among our heart team 1 week after her hospitalization. During surgery, femoral arterial (17 cm) and venous (21 cm) cannulation was

received
February 26, 2023
accepted after revision
March 9, 2023
accepted manuscript online
April 12, 2023

DOI <https://doi.org/10.1055/a-2072-0285>.
ISSN 2194-7635.

© 2023. The Author(s).

This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)

Georg Thieme Verlag KG, Rüdigerstraße 14, 70469 Stuttgart, Germany

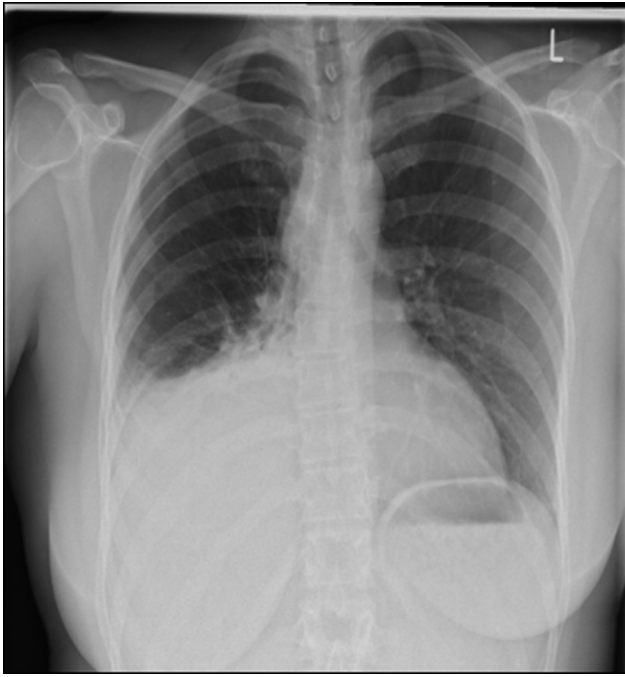


Fig. 1 Chest X-ray illustrates new right-sided pleural effusion.

established to be prepared for the aneurysm's accidental rupture during sternotomy. After opening the pleural cavity, we identified a giant vein aneurysm 15 cm × 7.5 cm in size. Its maximum extension was cranial at the azygos confluence to the SVC. The right brachiocephalic and right subclavian veins were of normal caliber.

After dissection of the aneurysm from the right phrenic nerve and surrounding tissue, we performed an additional venous cannulation (15 cm) at the left brachiocephalic vein with additional snaring. The right brachiocephalic vein and SVC (proximal) were also snared (►Fig. 3). A "beating heart" perfusion and hypothermia at 24°C were established. The distal azygos vein was ligated. The aneurysmal wall was opened longitudinally (►Fig. 4A). The SVC wall structure, especially at the posterior circumference, was as translucent as a pane of glass. This thin-walled aneurysmal posterior wall with multiple pretracheal veins' confluences was subsequently resected and closed with 7-0 Prolene in interrupted fashion (►Fig. 4B, C). The resulting ventral wall defect was augmented with a fresh autologous pericardial patch (►Fig. 5A). The outer surface of the reconstructed SVC was "wrapped" with an 18-mm-long acellular pericardial patch (Supple Peri-Guard, Lamed GmbH, Germany) to prevent the potential formation of pseudoaneurysms or a rupture relapse (►Fig. 5B).

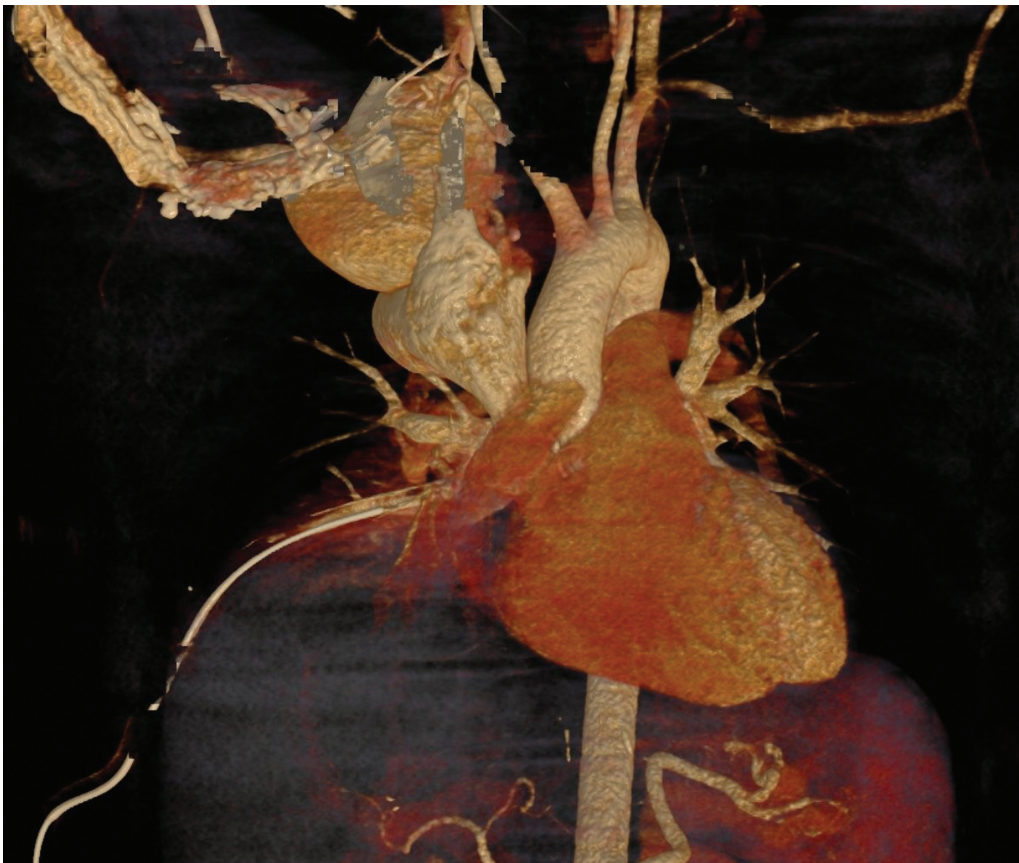


Fig. 2 Preoperative 3D chest CT angiography.

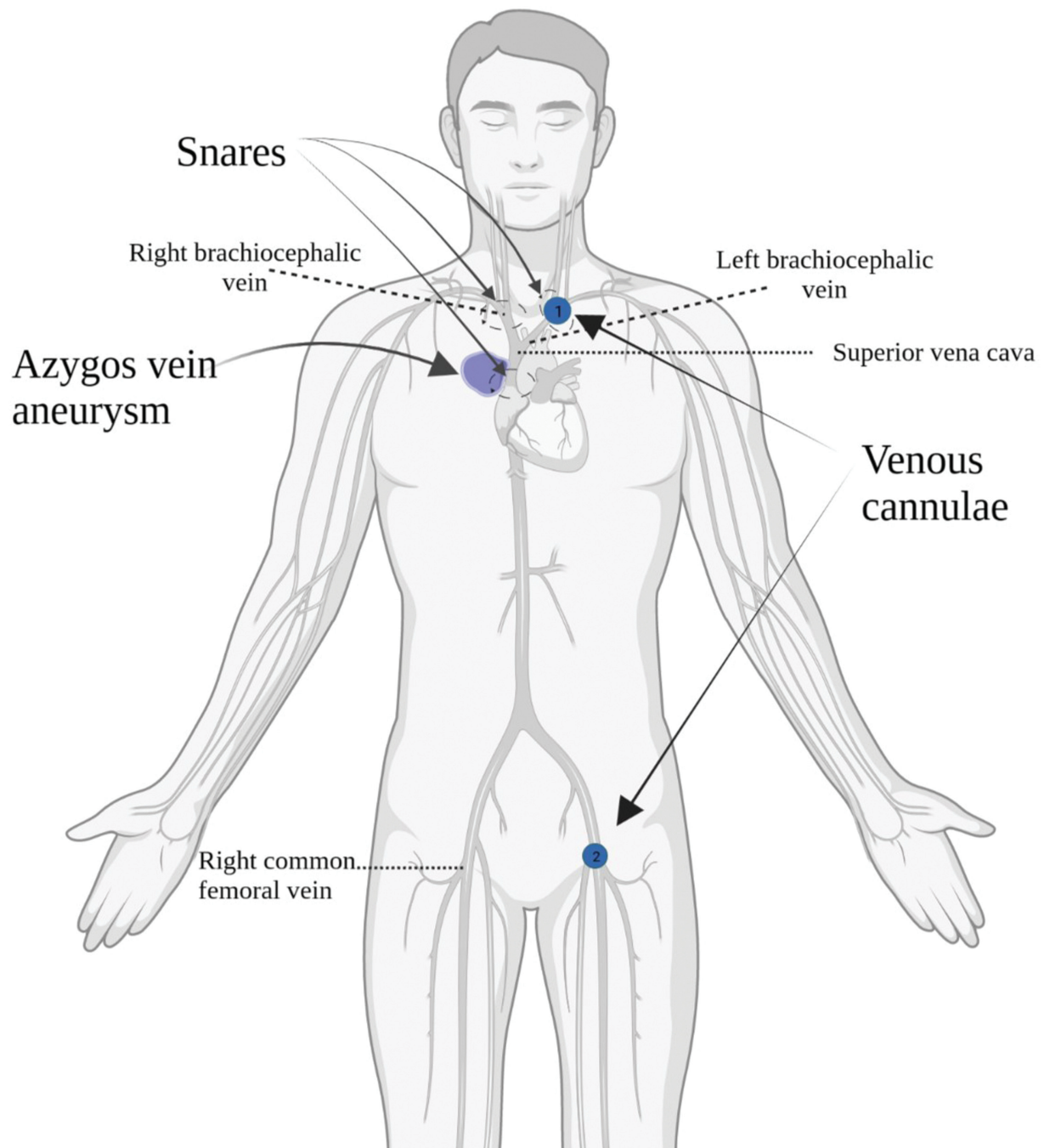


Fig. 3 Schematic drawing of the cannulation and the aneurysm topography.

Aspirin was started on the first postoperative day. CT at discharge showed a completely eliminated aneurysm and no evidence of relevant venous stenosis (►Fig. 6). After an initially unremarkable course, the patient developed supraventricular tachycardia, which was successfully converted to a sinus rhythm under medical therapy. She also developed a pulmonary disorder 2 weeks after surgery, attributable to atelectasis and an onset of pneumonia. After antibiotic therapy and intensive respiratory training, these resolved completely, and she was symptom-free at the time of discharge, which occurred 21 days after her admission. We prescribed 100-mg aspirin for the 6 months until her

next ambulatory follow-up. If no thrombosis or relevant stenosis is detected on the follow-up CT scan, we will discontinue aspirin. We recommend CT angiography 6, 12, and 24 months after surgery.

Discussion

SVC and azygos vein aneurysms are extremely rare phenomena, as fewer than 40 cases have been reported in the indexed English literature.¹ Detailed diagnostics include chest CT or magnetic resonance imaging (MRI) with angiography. The more common fusiform aneurysms are usually detected

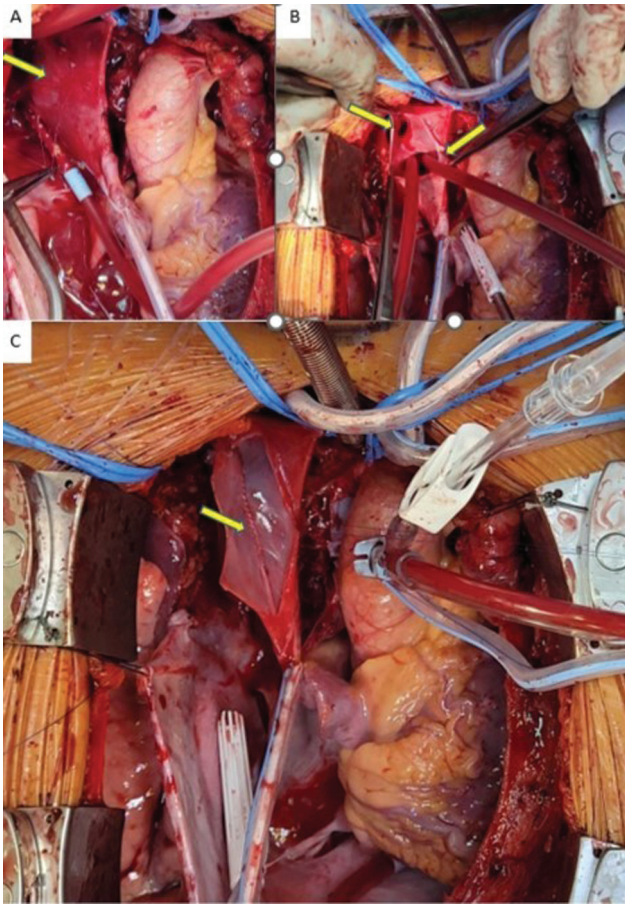


Fig. 4 Dissection of the aneurysm from the right phrenic nerve and the surrounding tissue (A), SVC posterior wall with multiple pretracheal vein's confluences. The aneurysm is depicted with a yellow arrow (B), and closure of SVC posterior wall after resection of pathological confluences (C).

incidentally, portend a benign prognosis, and are managed conservatively. Conversely, saccular aneurysms are often postinterventional or postinflammatory, carry a higher risk for rupture or thromboembolism, and usually require surgical resection.⁴ The rapid progression of such aneurysms can trigger thrombosis as well as pulmonary thromboembolism, and consequently cause acute right ventricular failure.⁵

Early surgical resection to prevent rupture is recommended for saccular aneurysms with a diameter of >40 mm, as well for symptomatic and complicated aneurysms.^{4,6} Cardiopulmonary bypass should be considered not only when a thrombus formation is detected, but also in case of calcification in the aneurysm wall. It is also safer for the patient and makes the surgeon more confident.⁴

The alternative to open surgery if the aneurysm's entrance measures less than 40 mm is an endovascular approach. This involves injecting thrombin into the aneurysm sack or coil occlusion of the aneurysm neck. However, most surgeons have little experience with these methods.⁷

After reviewing the literature, we found just one case report of a ruptured inferior vena cava aneurysm, and none of ruptured SVC or azygos vein aneurysm.⁸

After resecting her aneurysm and reconstructing the SVC, postoperative imaging showed a good surgical outcome without relevant stenosis and a completely eliminated aneurysmal sack, followed by a smooth recovery until discharge.

Conclusion

Azygos vein and SVC aneurysms are extremely rare and catastrophic phenomena. Young patients suffering from acute dyspnea and thoracic pain need to undergo careful differential diagnosis to ensure this high-risk anomaly is managed early

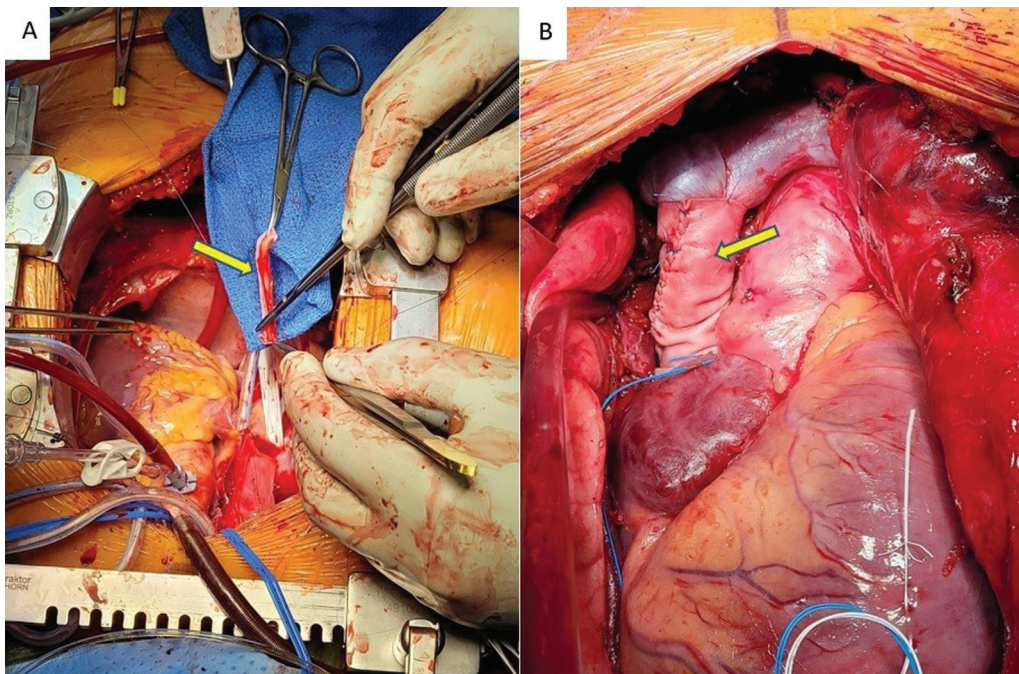


Fig. 5 Augmentation of SVC anterior wall with autologous pericardial patch (depicted with a yellow arrow) (A) and SVC "wrapping" with acellular pericardial patch (depicted with a yellow arrow) (B).



Fig. 6 Postoperative 3D chest CT angiography.

and effectively. Although surgical intervention is the standard treatment for >4-cm saccular azygos vein and SVC aneurysms and for any complicated aneurysm, conservative treatment can be an acceptable option in certain cases. We recommend caution and discretion when attempting to apply how we managed this patient to those with other venous aneurysms.

Funding

None.

Conflict of Interest

None declared.

References

- 1 Sharma R, Ravi M, Unni TG. Primary fusiform superior vena cava aneurysm. *Cardiol Res* 2017;8(04):176–178
- 2 Abbott OA, Leigh TF. Aneurysmal dilatations of the superior vena caval system. *Ann Surg* 1964;159(06):858–872
- 3 Pasic M, Schöpke W, Vogt P, von Segesser L, Schneider J, Turina M. Aneurysm of the superior mediastinal veins. *J Vasc Surg* 1995;21(03):505–509
- 4 Soares Souza LV, Souza AS, Morales MM, Marchiori E. Superior vena cava aneurysm: an unusual mediastinal mass. *Eur J Cardiothorac Surg* 2021;59(01):276–277
- 5 Oh SG, Kim KH, Seon HJ, et al. Unusual cause of acute right ventricular dysfunction: rapid progression of superior vena cava aneurysm complicated by thrombosis and pulmonary thromboembolism. *J Korean Med Sci* 2011;26(05):690–693
- 6 Varma PK, Dharan BS, Ramachandran P, Neelakandhan KS. Superior vena caval aneurysm. *Interact Cardiovasc Thorac Surg* 2003;2(03):331–333
- 7 Griviau L, Chevallier O, Favelier S, Pottecher P, Gehin S, Loffroy R. Endovascular management of a large aneurysm of the superior vena cava involving internal thoracic vein with remodeling technique. *Quant Imaging Med Surg* 2016;6(03):315–317
- 8 Momeni M, Momeni F. Ruptured inferior vena cava aneurysm in the setting of mural vascular malformation: a case report. *J Clin Ultrasound* 2019;47(07):423–425