Superior Vena Cava Syndrome Caused by Retrosternal Goiter: A Case Report

Sandra Doko Barišić[1], Marija Pastorčić Grgić[2], Mirta Zekan Vučetić[3], Tiha Vučemilo[4] and Dubravka Krleža Supić[5]

[1] Division of Head and Neck Surgery, Department of Surgical Oncology, University Hospital for Tumors, Sestre milosrdnice University Hospital Center, Zagreb, Croatia;
[2] Department of Otolaryngology, Head and Neck Surgery, Zagreb University Hospital Center, Zagreb, Croatia;
[3] Department of Interventional and Diagnostic Radiology, Merkur University Hospital, Zagreb, Croatia;
[4] Department of Transfusion and Regenerative Medicine, University Hospital for Tumors, Sestre milosrdnice University Hospital Center, Zagreb, Croatia;
[5] Department of Anaesthesia, Connolly Hospital, Blanchardstown, Dublin, Ireland

CORRESPONDENCE TO Sandra Doko Barišić, Division of Head and Neck Surgery, Department of Surgical Oncology, University Hospital for Tumors, Sestre milosrdnice University Hospital Center, Ilica 197, HR-10000 Zagreb, Croatia
sandra.doko@gmail.com

RECEIVED December 31, 2022
ACCEPTED October 9, 2024

DOI 10.20471/acc.2026.65.02.17

SUMMARY

Superior vena cava (SVC) syndrome develops because of intravascular thrombosis or extrinsic compression of the SVC. Current literature shows that 90%-97% of SVC syndrome cases are due to malignant mediastinal disease, and a small proportion of the cases develop because of benign conditions. The classic symptoms include dyspnea, cough, headache, facial and arm swelling. We present a rare case of SVC syndrome caused by retrosternal goiter in a 77-year-old woman. After confirming the diagnosis, total thyroidectomy was performed, resulting in complete remission of compressive symptoms. Multidisciplinary management is necessary to avoid complications in the treatment of high-risk patients with SVC syndrome.

KEYWORDS

Superior vena cava syndrome; *Retrosternal goiter*; *Case report*

Introduction

The first case of superior vena cava (SVC) syndrome was described by William Hunter in 1957, in a patient who died of aortic aneurysm as a complication of syphilis[1]. SVC is responsible for returning venous blood from the head, neck, upper mediastinum and arms to the right atrium. The right and left brachiocephalic veins converge to form the SVC at the level of the right first costal cartilage. The SVC is surrounded by rigid structures, i.e., sternum, trachea, right bronchus, aorta, perihilar and paratracheal lymph nodes. It is easily compressible due to its thin wall and low intravascular pressure. The causes of obstruction include extrinsic compression or thrombosis of the vein[2]. Existing literature shows that 90%-97%

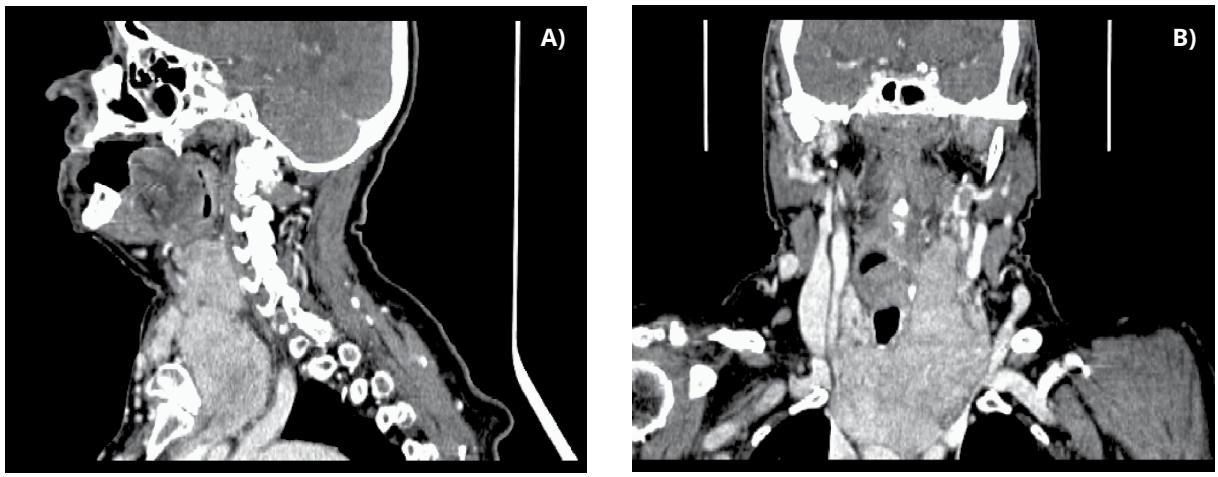

Fig. 1. Computed tomography finding of thyroid goiter extending retrosternally: sagittal plane (A); coronal plane (B).

of SVC syndrome cases are due to malignant conditions[3,4], with around 70% attributed to advanced lung cancer[5,6], 12% to lymphoma, and 9% to metastatic cancer[7]. A small proportion of SVC syndrome cases develop as a result of benign conditions[8]. We report a rare case of SVC syndrome caused by multinodular retrosternal goiter. This case report is presented in concordance with CARE guidelines for case reports[9].

Case report

A 77-year-old woman with a history of hypertension and chronic obstructive pulmonary disease was referred from another institution, where she presented with dyspnea. She reported noticing a slight painless swelling on the left side of the neck two weeks before. She underwent left hemithyroidectomy ten years before and histopathology confirmed nodular goiter. There was no history of prolonged immobilization or central venous catheter insertion. Clinical examination did not reveal any feature of deep vein thrombosis (DVT). Shortness of breath occurred with high respiratory rate (40/min) and extensive use of accessory respiratory muscles. Neck veins were distended.

After admission to the local hospital, a computed tomography (CT) scan and CT angiography were performed, and clinical diagnosis was SVC thrombosis. A second CT scan performed in our institution showed marked multinodular thyroid goiter that extended retrosternally and into the left mediastinum to the aortic arch (Fig. 1). The left

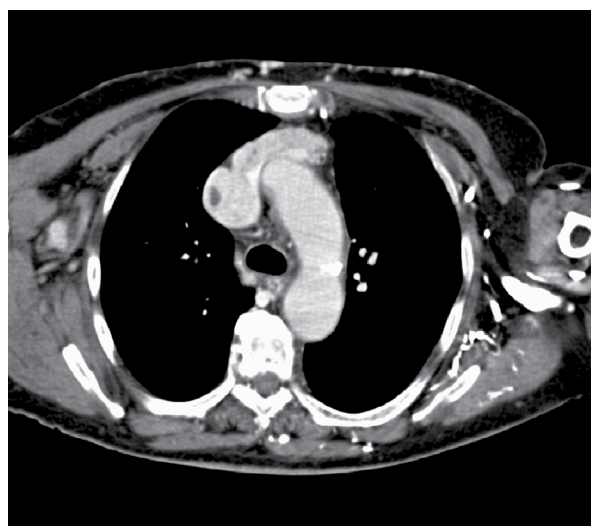

Fig. 2. Computed tomography finding of superior vena cava thrombosis.

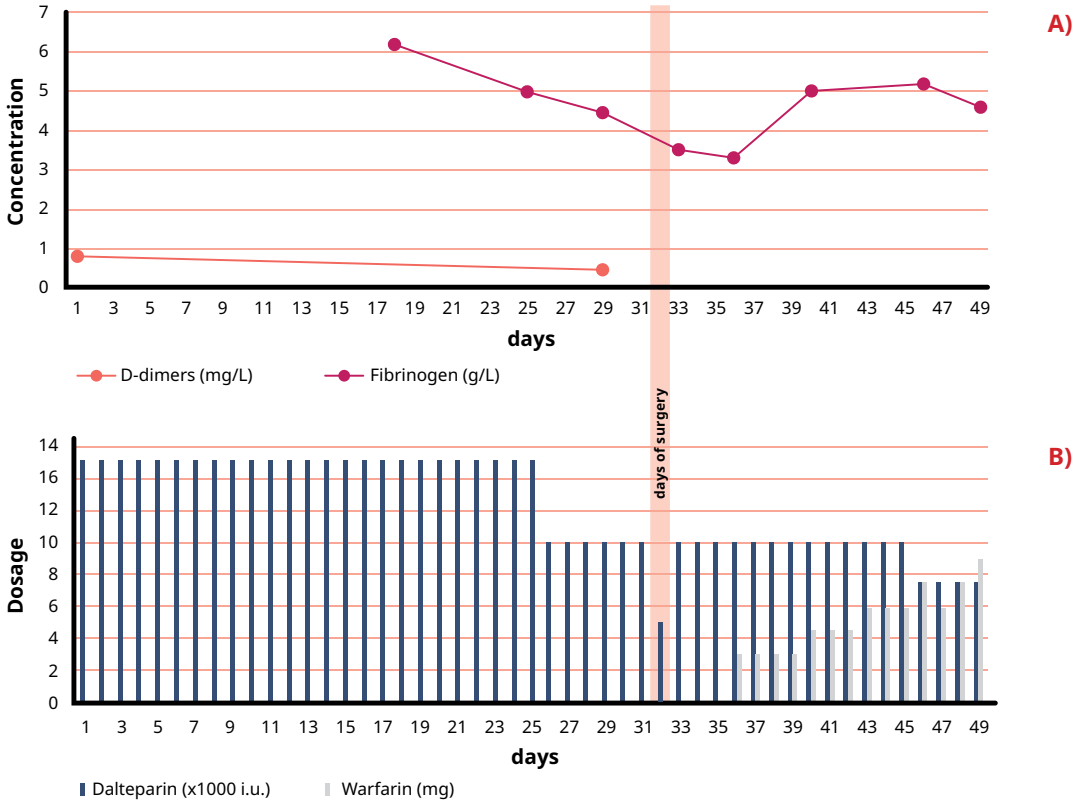

Fig. 3. Laboratory values (A) and therapy (B).

brachiocephalic vein was displaced in front of the enlarged left thyroid lobe and compressed. The scan also showed thrombosis of the left brachiocephalic vein and SVC (Fig. 2). The trachea was displaced to the right and slightly narrowed. Fine needle aspiration cytology showed follicular tumor. A battery of tests was done to exclude other causes of blood hypercoagulability. Prothrombin time, activated partial thromboplastin time, thrombin time, and international normalized ratio were within the normal ranges. Laboratory tests showed increased D-dimer and fibrinogen levels as expected in large vein thrombosis. The patient had elevated D-dimer level of 0.82 mg/L on day 1 and 0.48 mg/L on day 29 of the disease (normal range 0-0.55 mg/L). Fibrinogen level was elevated up to 6.7 g/L (normal range 1.8-3.5 g/L) on day 18 from symptom onset, with a tendency to normalization prior to surgery and slight elevation after surgery.

The patient was treated with therapeutic doses of low molecular weight heparin dalteparin (15000 IU once daily, sc), considering its efficacy in settings of acute DVT, and methylprednisolone (80 mg, once daily, iv), resulting in improvement of the symptoms. Figure 3 shows changing levels of D-dimer and fibrinogen, as well as of the anticoagulant therapy applied.

Total thyroidectomy *via* a neck approach was successfully performed 32 days after the first symptoms of SVC syndrome. Intraoperative findings correlated with CT scans, showing a large left thyroid lobe with retrosternal extension (Fig. 4). In the postoperative period, the patient developed transient hypocalcemia and left vocal cord paresis.

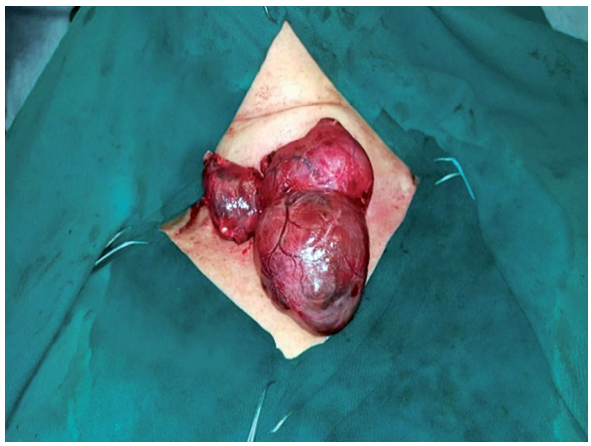

Fig. 4. Total thyroidectomy specimen with enlarged left thyroid lobe.

Histopathologic analysis indicated follicular adenoma and nodular goiter. The patient was discharged on postoperative day 18 with a plan for 2-monthly follow-up. Two years after the surgery, the patient was well, asymptomatic, and with no signs of malignant disease. Follow-up CT scan showed persistent thrombus in SVC.

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. Informed consent was obtained from the patient. Ethics approval was not required for this article, according to the Sestre milosrdnice University Hospital Center Ethics Committee.

Discussion

The SVC syndrome is a critical condition in which an intrathoracic mass causes extrinsic compression or thrombosis of SVC[2]. Due to the limited space of the thoracic cage, intrathoracic goiters can cause adjacent structure compression more frequently than cervical goiters. Differential diagnosis of the cause of SVC syndrome includes infectious and benign diseases, as well as malignant tumors. Nowadays, benign conditions are a rare cause of SVC syndrome. Thrombosis of the SVC related to the presence of indwelling intravascular devices is infrequent in spite of the increasing use of central venous catheters. Up to 50% of SVC syndrome cases not due to malignancy are attributable to fibrosing mediastinitis, as a result of a prior infection with *Histoplasma capsulatum*[8]. Among all the benign causes, retrosternal goiter is exceedingly rare. There are few isolated case reports of SVC syndrome secondary to retrosternal goiter[7,10]. In this case, the patient had a previously undiagnosed recurrence of multinodular retrosternal goiter, which eventually caused compression and obstruction of the left brachiocephalic vein and SVC.

The most common symptoms of SVC syndrome are dyspnea, cough, headache, facial and arm swelling. However, a patient can be asymptomatic in 10% of cases[11]. This patient had a prominent symptom of dyspnea, which could easily be misdiagnosed as exacerbation of the patient's chronic obstructive pulmonary disease. Additionally, extensive distension of neck veins was found on physical examination, which probably led to proper diagnostic protocol. According to the literature, both symptoms occur in more than half cases of SVC syndrome[12]. Yu *et al.*[12] proposed a classification system and algorithm for the management. Our patient would be classified as a mild to moderate case (grade I-II).

Based on diagnostic work-up, the underlying cause of SVC syndrome in this patient was benign and treatment plan was with curative intent[13]. The management of DVT depends largely on the etiology; however, in the absence of a contraindication, the cornerstone of treatment is anticoagulation[14]. Although the majority of clinicians first advise thrombolytic therapy followed by parenteral heparin therapy, we opted for a less aggressive management.

Therefore, we started therapy with a therapeutic dose of low molecular weight heparin, considering its efficacy in settings of acute DVT[14]. Anticoagulation should be continued for at least 3 months with either low molecular weight heparin, vitamin K antagonists, or direct oral anticoagulants[14]. In our case, dalteparin was applied for 49 days, gradually reducing the dose, and warfarin was added on postoperative day 5.

There are several clinically relevant complications resulting from upper extremity DVT, as follows: symptomatic pulmonary embolism, recurrence, and post thrombotic syndrome[15]. Upper extremity DVT has a lower risk of pulmonary embolism (up to 10%) compared to lower extremity DVT (15%-30% of symptomatic pulmonary embolism)[15-17]. The post-thrombotic syndrome which combines upper extremity pain and swelling has been seen in up to 13% of patients[14].

Timing of surgical decompression is controversial; if necessary, surgical evaluation is usually performed within 3 months[14].

Therapeutic-dose anticoagulation can lead to unacceptable risk of bleeding complications in surgical patients[18,19]. Considering the risks of DVT and therapeutic-dose anticoagulation, surgery was planned once the D-dimer and fibrinogen values were back in the normal range. Furthermore, dalteparin dose was reduced on the day of surgery.

In conclusion, it is important to consider a possibility of SVC syndrome in a patient with a thyroid goiter. Multidisciplinary management is necessary to avoid complications in the management of a high-risk patient with SVC syndrome. ■

References

1. Hunter W. The history of an aneurysm of the aorta, with some remarks on aneurysms in general. Med Observ Inq. 1757;1:323-57.
2. Ingle SA, Karnik ND, Pradhan NN, Ranadive NU, Dhurde A, Kotalwar M. Papillary carcinoma of thyroid presenting as posterior mediastinal mass with superior vena cava syndrome. J Assoc Physicians India. 2004;52:1004-5.
3. Wilson LD, Detterbeck FC, Yahalom J. Clinical practice. Superior vena cava syndrome with malignant causes. N Engl J Med. 2007;356:1862-9. doi: 10.1056/NEJMcp067190.
4. Padmanabhan H. Superior vena cava syndrome: a presentation of anaplastic thyroid carcinoma. J Clin Oncol. 2010;28:151-4. doi: 10.1200/JCO.2009.24.8740.
5. Flounders JA. Superior vena cava syndrome. Oncol Nurs Forum. 2003;30:84-8. doi: 10.1188/03.ONF.E84-E90.
6. Ahmann FR. A reassessment of the clinical implications of the superior vena caval syndrome. J Clin Oncol. 1984;2:961-9. doi: 10.1200/JCO.1984.2.8.961.
7. Promisloff RA, Pervall-Phillips GP. Superior vena cava syndrome caused by a benign retrosternal multinodular goiter. J Am Osteopath Assoc. 1997;97:409-10.
8. Lepper PM, Ott SR, Hoppe H, Schumann C, Stammberger U, Bugalho A, *et al.* Superior vena cava syndrome in thoracic malignancies. Respir Care. 2011;56:653-66. doi: 10.4187/respcare.00947.
9. Riley DS, Barber MS, Kienle GS, Aronson JK, von Schoen-Angerer T, Tugwell P, *et al.* CARE guidelines for case reports: explanation and elaboration document. J Clin Epidemiol. 2017;89:218-35. doi: 10.1016/j.jclinepi.2017.04.026.
10. Polat Y, Altinsoy HB, HT Polat, Kanat BH, Sözen S, Dal MB. Superior vena cava syndrome caused by a benign intrathoracic goiter. Arch Clin Exp Surg. 2016;5:242-5. doi:10.5455/aces.20150508103342
11. Holliday EB, Hampton DA, Thomas CR Jr, Rana S. Prognostic value of the Yale grading system for superior vena cava syndrome. Ann Am Thorac Soc. 2016;13:1862□5. doi: 10.1513/AnnalsATS.201605-412LE.
12. Yu JB, Wilson LD, Detterbeck FC. Superior vena cava syndrome: a proposed classification system

and algorithm for management. J Thorac Oncol. 2008;3:811-4. doi: 10.1097/JTO.0b013e3181804791.
13. Klein-Weigel PF, Elitok S, Ruttloff A, Reinhold S, Nielitz J, Steindl J, *et al.* Superior vena cava syndrome. Vasa. 2020;6:437-48. doi: 10.1024/0301-1526/a000908.
14. Kearon C, Akl EA, Ornelas J, *et al.* Antithrombotic therapy for VTE disease: CHEST guideline and expert panel report. Chest. 2016;149:315-52. doi: 10.1016/j.chest.2015.11.026.
15. Engelberger RP, Kucher N. Management of deep vein thrombosis of the upper extremity. Circulation. 2012;126:768-73. doi: 10.1161/CIRCULATIONAHA.111.051276.
16. Munoz FJ, Mismetti P, Poggio R, Valle R, Barrón M, *et al.* Clinical outcome of patients with upper-extremity deep vein thrombosis: results from the RIETE Registry. Chest. 2008;133:143-8. doi: 10.1378/chest.07-1432.
17. Lee JA, Zierler BK, Zierler RE. The risk factors and clinical outcomes of upper extremity deep vein thrombosis. Vasc Endovascular Surg. 2012;46:139-44. doi: 10.1177/1538574411432145.
18. Muntz JE, Michota FA. Prevention and management of venous thromboembolism in the surgical patient: options by surgery type and individual patient risk factors. Am J Surg. 2010;199(1 Suppl):S11-20. doi: 10.1016/j.amjsurg.2009.10.007.
19. Muntz J, Scott DA, Lloyd A, Egger M. Major bleeding rates after prophylaxis against venous thromboembolism: systematic review, meta-analysis, and cost implications. Int J Technol Assess Health Care. 2004;4:405-14.

SAŽETAK

Sindrom gornje šuplje vene uzrokovan retrosternalnom strumom: prikaz slučaja

Sandra Doko Barišić, Marija Pastorčić Grgić, Mirta Zekan Vučetić, Tiha Vučemilo i Dubravka Krleža Supić

Sindrom gornje šuplje vene je kliničko stanje koje nastaje kao posljedica intravaskularne tromboze ili vanjske kompresije gornje šuplje vene. Prema dostupnoj literaturi, u 90%-97% slučajeva trombozu uzrukuje zloćudna bolest u medijastinumu, dok su dobroćudna stanja iznimno rijedak uzrok. Tipični simptomi su zaduha, kašalj, glavobolja i oteklina glave i ruku. Prikazujemo rijedak slučaj sindroma gornje šuplje vene koji je uzrokovan retrosternalnom strumom kod 77-godišnje bolesnice. Nakon potvrde dijagnoze učinjena je totalna tireoidektomija, što je dovelo do potpunog povlačenja kompresivnih simptoma. U liječenju sindroma gornje šuplje vene kod ovakvih visokorizičnih bolesnika iznimno je važan multidisciplinarni pristup.

KLJUČNE RIJEČI

Sindrom gornje šuplje vene; *Retrosternalna struma*; *Prikaz slučaja*