SAGE Open Medical Case Reports

SAGE Open Medical Case Reports
JCMS Case Reports
Volume 8: 1–4

DOI: 10.1177/2050313X20919618
journals.sagepub.com/home/sco
SAGE

What lies downstream? A case of superior vena cava syndrome presenting in the dermatology clinic: A case report

Rochelle Tonkin[1,2] and Christopher Sladden[1,3]

Abstract

A 75-year-old female presented with a 1 year history of a firm, diffusely swollen, and erythematous facial plaque. She had preceding unsuccessful investigations and treatment for angioedema. Full-skin examination revealed multiple prominent varicosities on the chest and abdomen. Superior vena cava syndrome was suspected. Solid facial edema (Morbihan's syndrome) and various infiltrates included in the differential diagnosis were ruled out with a skin biopsy. Chest computed tomography confirmed a complete superior vena cava thrombosis. Extensive workup for associated malignancy has to date been unremarkable, and as yet an underlying cause remains to be found.

Keywords

Dermatology, solid facial edema, varicosities, superior vena cava syndrome

Introduction

Obstruction of the superior vena cava (SVC) that impedes blood flow leads to a constellation of signs and symptoms known as SVC syndrome.[1] Identifying SVC syndrome can be a challenge due to the variability in which it can present, especially when it presents to dermatology consultation with primary cutaneous findings. Key cutaneous manifestations of SVC syndrome include facial edema, flushing, and dilated chest veins.[1] The true diagnosis can be missed if these findings are not investigated appropriately and thoroughly. SVC syndrome can be a life-threatening condition, which can quickly deteriorate leading to a poor outcome if not recognized and treated in a timely manner. Despite SVC syndrome being an extremely rare cause of facial edema, which already has a broad differential, it should always be considered.

Case report

A 75-year-old female was referred to the outpatient dermatology clinic for management of angioedema. She presented with a diffusely swollen, firm, erythematous facial plaque that developed suddenly and persisted for 1 year. There were no other associated systemic symptoms or inciting event. There were mild fluctuations in the facial swelling, although it never fully resolved. She had undergone some initial investigations initiated by her family physician and allergologist, and was treated for angioedema without success. Multiple allergy tests were unremarkable. Chest X-ray had demonstrated normal heart size, unremarkable mediastinum, and no evidence of mediastinal mass or vascular dilation.

Past history included controlled type 2 diabetes and dyslipidemia. There was no previous history of malignancy or prior dermatology conditions. Medications included metformin, insulin, and atorvastatin. Family history included both parents with unspecified cancers. There were significant second-hand smoke exposures from both her parents and her partner.

Examination revealed a diffusely edematous facial plaque that was firm to palpation and associated with erythema that was continuous throughout including the nasolabial folds (Figure 1). On further evaluation, she was found to have prominent and dilated veins on the ventral chest and neck

[1]University Hospital of Northern British Columbia, Prince George, BC, Canada
[2]Faculty of Medicine, The University of British Columbia, Vancouver, BC, Canada
[3]Department of Dermatology & Skin Science, Faculty of Medicine, The University of British Columbia, Vancouver, BC, Canada

Corresponding author:
Christopher Sladden, 275 – 546 St Paul's St., Kamloops, BC V2E1S2, Canada.
Email: csladden@shaw.ca

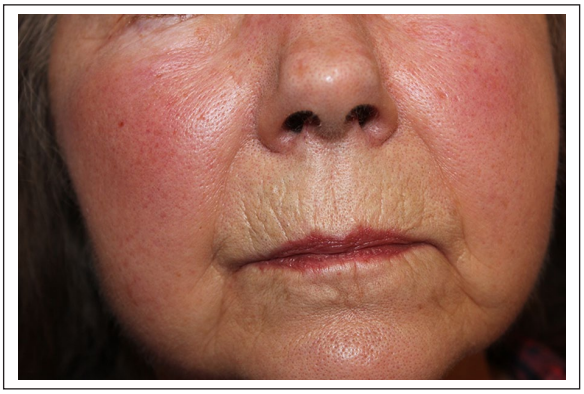

Figure 1. Initial presentation was a diffusely edematous and erythematous facial plaque.

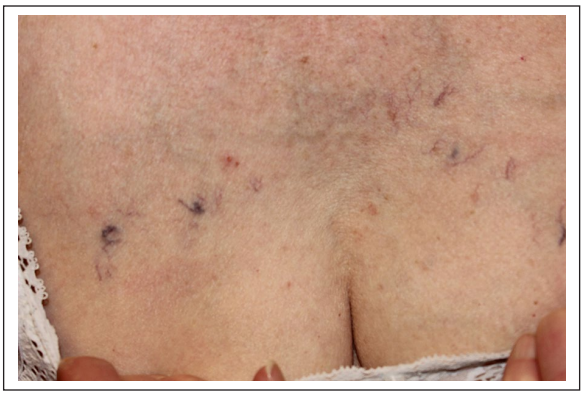

Figure 2. The patient's ventral chest, demonstrating prominent and dilated veins.

(Figure 2), as well as enlarged tortuous veins on her abdomen (Figure 3). There was no extremity edema or erythema.

Based on history and physical examination, a diagnosis of angioedema was unlikely. The differential diagnosis included solid facial edema (Morbihan's syndrome) and various infiltrates such as lupus, lymphoma, mucinosis, amyloidosis, myxedema, leprosy, and sarcoidosis. The prominent vascular findings taken in context with the facial features led to a vena cava syndrome being considered.

Biopsy of the facial skin reported benign solar elastosis, with no other significant pathologic abnormality, therefore helping to rule out Morbihan's syndrome and infiltrative etiologies. Blood work including antinuclear antibody with serum and urine protein electrophoresis was normal. An urgently ordered computed tomography (CT) of the chest revealed complete obstruction of SVC due to thrombosis, with multiple collateral vessels throughout chest and mediastinum (Figure 4(a) and (b)). There was no evidence of intraluminal airway mass, pulmonary parenchymal abnormality, nor pleural thickening or effusions. In addition, no axillary, mammary, or cardiophrenic lymphadenopathies were identified.

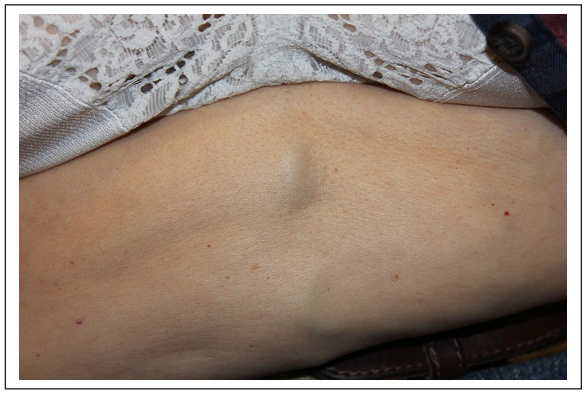

Figure 3. The patient's upper abdomen, showing enlarged tortuous veins.

The patient was admitted to hospital for anticoagulation therapy and workup to determine the thrombus etiology. Investigations did not uncover any malignancy or other cause for the thrombus. International normalized ratio (INR) was 1.0 and partial thromboplastin time of 24.9 (range 25–38) was within normal limits. The D-dimer level was elevated at 1.54 (normal range below 0.49). Within 1 month of being seen in the outpatient dermatology clinic, her condition had progressed to include a swollen left arm with pitting edema and the facial and neck swelling persisted. An ultrasound venous Doppler showed superficial thrombophlebitis of the basilic vein, but no evidence of deep vein thrombosis. Despite to date there being no known evidence of malignancy or intrathoracic mass, the former remains the most likely underlying cause. Radiology and thoracic surgery consultation agreed that an undiagnosed lung malignancy, including lymphoma, was the most likely etiology due to the patient's age and obstruction location. Covert neck or breast malignancy remains possibilities. Mass lesions such as esophageal cancer, thyroid goiter, Castleman's disease, lymphadenopathy secondary to cystic fibrosis, restrictive pericarditis, histoplasmosis, and syphilitic or tuberculotic aneurysm can rarely manifest as SVC syndrome. Repeat imaging and clinical exam helped exclude these mass lesions. The patient will be monitored longitudinally with further imaging and thoracic surgery follow-up.

Discussion

SVC syndrome can be a challenge to identify with primary cutaneous manifestations being the sole presenting features. The key signs and symptoms of SVC syndrome include the following: face or upper neck swelling, upper extremity swelling, dyspnea, cough, dilated chest veins, chest/shoulder pain, flushing/plethora, syncope/presyncope, headaches, and hoarseness.[1,2] In this case, the facial edema was the primary presentation, and only on full-skin examination were the more characteristic chest and abdominal varicosities discovered.

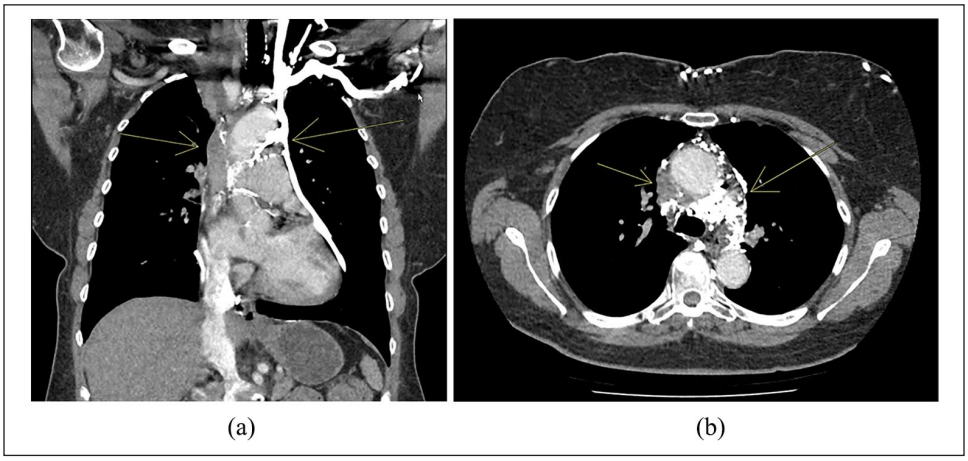

Figure 4. CT angiogram of the chest. (a) A coronal view and (b) an axial view, both show the occluded SVC (left-sided arrow on the patient's right) and the bright white opacified collaterals (right-sided arrow on the patient's left).

There are similar examples of SVC syndrome presenting to dermatology with primary cutaneous findings, including intermittent facial swelling,[3] periorbital edema,[4] persistent erythematous facial edema,[5] position-dependent periorbital edema,[6] and non-pitting infraorbital edema/rash on chest.[7]

There are cases of SVC syndrome similarly mistaken initially for angioedema,[3] as well as acute allergic reaction[8] and drug reaction or angioneurotic edema.[9] Given the extensive history of unremitting facial edema with no evident inciting event such as allergic exposure, this was extremely unlikely to be chronic angioedema.

The differential for facial edema is broad as it includes inflammatory, infiltrative, infectious, allergic, and autoimmune conditions. Although rare, SVC syndrome should always be included in the differential due to risk of negative outcomes such as severe edema, pleural effusions, and esophageal varices that can become life-threatening.[10,11] Our clinical suspicion for SVC syndrome was high due to the additional findings of truncal varicosities. However, it was still important to rule out primary cutaneous etiologies, especially those infiltrates which might be both the cause and effect such as lymphoma. Facial biopsy with essentially unremarkable findings was useful in this regard. CT imaging is the best modality for evaluating suspected SVC syndrome,[12] and it was key to confirming the diagnosis in this case.

SVC obstruction typically occurs due to a mass compressing the SVC or thrombosis occluding the vessel lumen.[13] For extrinsic mass compression, primary lung malignancy is statistically the most common cause, while lymphoma and metastatic breast cancer are also to be considered.[2] It is possible in this case that the obstructing SVC thrombus may have been a consequence of an infection, neoplastic process, or hypercoagulable state. Given the patient's age, malignancy causing a hypercoagulable state remains the most probable etiology.

Acknowledgements

The authors thank Dr Kristina Sharma, radiologist at the University Hospital of Northern British Columbia, for the images included.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship, and/or publication of this article.

Informed consent

The patient provided consent to publish her case and photographs.

References

1. Rice TW, Rodriguez RM and Light RW. The superior vena cava syndrome: clinical characteristics and evolving etiology. *Medicine (Baltimore)* 2006; 85(1): 37–42.
2. Cheng S. Superior vena cava syndrome: a contemporary review of a historic disease. *Cardiol Rev* 2009; 17(1): 16–23.
3. Pope C and Pillai SK. Intermittent facial swelling. *BMJ Case Rep* 2014; 2014: bcr2013202355.
4. Dyken JR, Pagano JP and Soong VY. Superior vena caval syndrome presenting as periorbital edema. *J Am Acad Dermatol* 1994; 31(2 Pt 1): 281–283.
5. Jansen T, Romiti R, Messer G, et al. Superior vena cava syndrome presenting as persistent erythematous oedema of the face. *Clin Exp Dermatol* 2000; 25(3): 198–200.
6. Nguyen AL, Belderbos H, van Harten JG, et al. Superior vena cava syndrome presenting as position-dependent periorbital oedema. *BMJ Case Rep* 2018; 2018: bcr-2018-225220.
7. Ratnarathorn M and Craig E. Cutaneous findings leading to a diagnosis of superior vena cava syndrome: a case report and review of the literature. *Dermatol Online J* 2011; 17(6): 4.

8. Yong ASW. Superior vena cava obstruction presenting as acute allergy. *Clin Exp Dermatol* 2013; 38(8): 955–956.
9. Angevine CD and Condemi JJ. The facial edema of vena cava syndrome mistaken for angioneurotic edema. *J Allergy* 1966; 38(5): 308–314.
10. Burgdorff T, Douwes KE, Bogenrieder T, et al. Superior vena cava syndrome: an important differential diagnosis in patients with facial edema. *Acta Derm Venereol* 2001; 81(3): 205–206.
11. Greenwell MW, Basye SL, Dhawan SS, et al. Dialysis catheter-induced superior vena cava syndrome and downhill esophageal varices. *Clin Nephrol* 2007; 67(5): 325–330.
12. Tatu WF, Winzelberg GG, Boller M, et al. Computed tomographic evaluation of compression of the superior vena cava and its tributaries. *Cardiovasc Intervent Radiol* 1985; 8(2): 89–99.
13. Parish JM, Marschke Rf Jr, Dines DE, et al. Etiologic considerations in superior vena cava syndrome. *Mayo Clin Proc* 1981; 56(7): 407–413.