Superior vena cava syndrome presenting as position-dependent periorbital oedema

A L Nguyen,1 H Belderbos,2 J G van Harten,2 L Wijne3

DESCRIPTION
A 54-year-old woman presented to our dermatology clinic with a 2 months history of periorbital oedema, more prominent after lying down in horizontal position. Her face and throat felt swollen. She experienced dyspnoea on exertion, fatigue and had lost 6 kg of weight. Treatment with antihistamines and oral prednisone were ineffective. She smoked one pack of tobacco per week. Medical history and family history were unremarkable. She did not use any medications. Physical examination revealed prominent periorbital oedema and facial swelling (figure 1). Chest radiograph revealed a pathologically enlarged right hilum, broadened mediastinum and modest pleural effusion (figure 2). Diagnosis of superior vena cava syndrome (SVCS) was made and the patient was referred to the pulmonologist.

Five days later, she presented at the emergency department with haemoptysis and dyspnoea. Physical examination revealed additional distended superficial veins of anterior chest wall and decreased breath sounds over right lung area. CT scan demonstrated a large process in the right hilum with extensive mediastinal lymphadenopathy, almost completely compressing the superior vena cava, with partial atelectasis of right lower lung lobe and extensive pleural effusion (figure 3). Histological biopsy obtained by bronchoscopy revealed small cell lung cancer (SCLC). Positron emission tomography scan was suspect for left adrenal gland metastasis. SVCS was caused by stage IV SCLC, extensive disease. The patient passed away after several lines of therapy (chemotherapy, radiotherapy and study medication) due to progressive adrenal metastasis and symptomatic brain metastasis.

SVCS consists of symptoms and signs resulting from partial or complete obstruction of blood flow within the superior vena cava. Most cases are caused by underlying intrathoracic malignancy, of which up to 95% are due to lung cancer or non-Hodgkin’s lymphoma.1 2 This syndrome should be considered in patients presenting with periorbital or facial oedema, aggravated in horizontal position. Other important clinical clues for SVCS include distension of neck and chest wall veins, dyspnoea on exertion, coughing and arm swelling.1 2 Also, the Pemberton’s sign is indicative of SVCS when facial congestion and cyanosis occur by elevating both arms.3 Diagnosis should be confirmed by CT scan.

Figure 1 Prominent periorbital oedema and facial swelling after lying down the night before as a presenting symptom of superior vena cava syndrome.

Figure 2 Chest radiography showing a broadened mediastinum with shading of the right hilus (green arrow) and pleural effusion (blue arrow).

Figure 3 Chest CT demonstrating demonstrated a large infiltrative tumour in the right lower lung lobe (green arrow), almost completely compressing the superior vena cava (red arrow), with extensive lymphadenopathy and pleural effusion (blue arrow).

1Department of Dermatology, Leids Universitair Medisch Centrum, Leiden, The Netherlands
2Department of Pulmonology, Amphia Ziekenhuis, Breda, The Netherlands
3Department of Dermatology, Amphia Ziekenhuis, Breda, The Netherlands

Correspondence to L Wijne, lwijne@amphia.nl

Accepted 18 May 2018
Images in...

Learning points

► Superior vena cava syndrome (SVCS) should be considered in patients with periorbital or facial oedema, especially when aggravated in horizontal position.
► Early diagnosis of SVCS is crucial in order to alleviate symptoms of obstruction and initiate immediate treatment to prevent life-threatening complications.

Contributors ALN and LW discussed the reporting and design of the manuscript. ALN and LW drafted the manuscript. ALN, HB, JvH and LW critically revised the manuscript for improved intellectual content. ALN, HB, JvH and LW read and gave final approval for the submitted manuscript. ALN, HB, JvH and LW cared for the patient.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent Next of kin consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Author note Informed consent was obtained from the patient (oral approval) and the husband of the patient (signed approval).

References


Copyright 2018 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit http://group.bmj.com/group/rights-licensing/permissions.

BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:
► Submit as many cases as you like
► Enjoy fast sympathetic peer review and rapid publication of accepted articles
► Access all the published articles
► Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow