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Endovascular stenting in malignant obstruction of superior vena cava

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ABSTRACT

INTRODUCTION: Superior vena cava syndrome (SVCS) is obstruction of blood flow through the SVC. It is a medical emergency and most often manifests in patients with a malignant disease process within the thorax. A patient with SVCS requires immediate diagnostic evaluation and therapy.
PRESENTATION OF CASE: A 33-years-old woman presented with complaints of dyspnoea and chest pain. Computer tomography revealed a large mass in the anterior mediastinum. This mass compressed surrounding structures. Stenting was indicated for early symptoms of SVCS. The diagnosis of Hodgkin's lymphoma (HL) was confirmed with biopsy. The patient's stage II HL has been subsequently treated with six cycles of chemotherapy with doxorubicin, bleomycin, vinblastine, dacarbazine (ABVD), followed by radiotherapy. Presently she is doing well.
DISCUSSION: Although lymphomas are a common cause of SVCS but almost always SVCS is caused by non-Hodgkin's lymphoma (NHL). HL despite its common presentation with mediastinal lymphadenopathy rarely causes SVCS.
CONCLUSION: Lymphomas are a common cause of SVCS in young age. HL may present as SVCS. Pathological confirmation of diagnosis should be done before initiating therapy while dealing with a case of SVCS. SVC stenting is effective and has few complications in patients with SVCS.

1. Introduction

The mediastinum is the cavity that separates the lungs from the rest of the chest [1]. More than two thirds of mediastinal tumors are benign, masses in the anterior compartment are more likely to be malignant [2]. Localizing symptoms are secondary to tumor invasion. Common localizing symptoms include respiratory compromise, dysphagia, paralysis of the limbs, diaphragm, and vocal cords, Horner syndrome, and superior vena cava syndrome (SVCS). Systemic symptoms are typically due to the release of excess hormones, antibodies or cytokines [3].

SVCS includes various clinical signs and symptoms due to external compression of the SVC itself or greater veins emptying into the SVC or the superior cavo-atrial junction, resulting in reduced blood flow. The assessment of a patient with SVCS must be prompt. The optimal treatment choice should be decided with an interdisciplinary consensus [4]. Although lymphomas are a common cause of SVCS but almost always SVCS is caused by non-Hodgkin's lymphoma (NHL). Hodgkin's lymphoma (HL) despite its common presentation with mediastinal lymphadenopathy rarely causes SVCS [5].

2. Presentation of case

A 33-years-old woman was hospitalized at the Department of Pulmonology and Phthisiology, Martin University Hospital with mediastinal mass finding on conventional chest radiography in October 2011. The patient was hospitalized on the recommendation of local pulmonologist, who ordered a chest X-ray for differential diagnosis of dyspnoea and chest pain. It revealed homogeneous dark area on the right chest side.

The patient reported a two-week history of progressive dyspnoea after exertion, dry irritating cough, and mild chest pain. She complained from night sweats but without fever. She also referred with a weight loss of 10 kg for the last 3 weeks. She had no experience with chest oppressions or heart palpitations but she suffered from occasional dizziness.

On physical examination, temperature was normal, blood pressure was 130/80 mm Hg in both arms, pulse rate is 96/min, and respiration rate is 26/min. Pulses were symmetric and equal in all extremities. There was marked facial oedema. The skin was pale, anicteric, with no signs of bleeding or peripheral cyanosis. Her neck veins were distended, prominent veins were present on whole of the chest wall and the flow in these veins was from above down-

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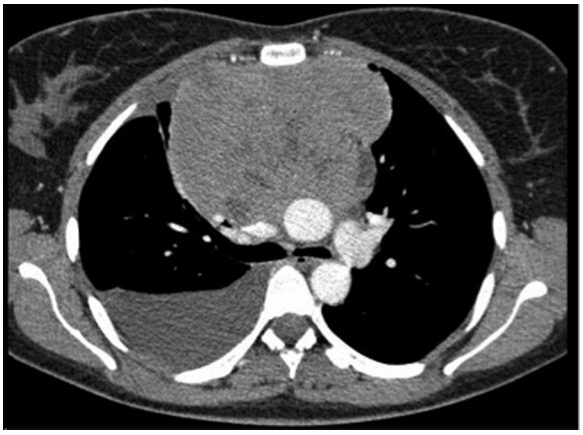

Fig. 1. Postcontrast CT images at the level of crossing of left brachiocephalic vein and arch of aorta showing a large homogeneously enhancing mass lesion in anterior mediastinum (October, 2011).

wards. On examination of respiratory system there was dullness on percussion over right side of chest at infraclavicular, suprascapular and interscapular areas, vocal resonance and vocal fremitus was also reduced in these areas and there were decreased intensity of breath sounds in these areas. Heartbeat was regular, without murmurs. The liver did not exceed the costal margin, the spleen was not felt and there was no mass in the abdomen. Peripheral lymphadenopathy was not present.

According to medical records, in June 2000, she underwent appendectomy. There was no other personal or family history. She had allergic contact dermatitis from nickel.

Based on classical clinical features of swelling over upper body, dilated veins over neck and chest, dyspnoea and dysphagia a provisional diagnosis of SVCS was made and she was investigated further.

2.1. Investigations

Laboratory tests on the day of admission revealed mild anaemia (haemoglobin level 11 g/dL). Total leucocyte count was 8500/mm^3 and differential leucocyte count was neutrophils 71% and lymphocytes 29%. The biochemical tests like liver and renal function tests were within normal limits. Bronchoscopy demonstrated stenosis of the right lower lobe bronchus. Segmental bronchi were also closed. Computed tomography (CT) scan revealed a mass in anterior mediastinum. This mass compressed the surrounding vascular structures (Fig. 1). Therefore, a thoracic surgeon was consulted. Anterior mediastinotomy was indicated. In early October, the operation was performed for tissue biopsy and pleural effusion. For early symptoms of the SVCS a stent was inserted into the SVC (Fig. 2). The procedure was performed with the patient under local anaesthesia (1% mesocain) in the angiographic suite (equipped with a fluoroscopic machine; Siemens, Munich, Germany) and was performed by an interventional team assisted by an anaesthesiologist for cardiocirculatory and airway support. Intubation was not necessary. Venography was performed to evaluate the location, length, and severity of the stenosis. The right common femoral vein was used as the approach site to access the SVC. After puncture of femoral vein, a 6-F sheath was inserted. The stenosis of the SVC was negotiated with a 5-F catheter and a 0.035-in hydrophilic guidewire that was 150 cm in length. The pressure gradient between the brachiocephalic vein or the internal jugular vein and the right atrium was measured before inserting the stent. Predilation was performed with a 6 mm (diameter) balloon catheter to determine stent size and to facilitate easy navigation of the stenosis and placement of the stent. We used the self-expanding nitinol Sinus-XL stent (OptiMed, Ettlingen, Germany). Heparin (4000 units) was administered intravenously prior to these procedures. Warfarin administration was continued for 18 months followed by aspirin administration (81 mg/day). The postoperative period passed without any complications. Histologic analysis revealed a Hodgkin's lymphoma (HL). Therefore, the patient was moved to the Department of Haematology and Transfusiology in order to start treatment and complet the staging. In the beginning of the first hospitalization the trephine biopsy excluded infiltration of bone marrow.

2.2. Treatment

The first cycle of ABVD immunochemotherapy was administered in early November. ABVD is an acronym for different chemotherapeutic drugs. "A" stands for adriamycin; "B" for bleomycin; "O" for vinblastine; and "D" for dacarbazine. All drugs were given intravenously on days 1 and 15 (doxorubicin 25 mg/m^2, bleomycin 10 mg/m^2, vinblastine 6 mg/m^2 and dacarbacine 375 mg/m^2). The treatment was repeated every 4 weeks.

2.3. Outcome and follow-up

After four cycles of chemotherapy CT showed regression of tumor, so it was decided to continue with the treatment (Fig. 3). The last (sixth) cycle of chemotherapy was administered in March 2012. Restaging CT examination revealed persistent tumor 6 × 5 × 3 cm in size. Based on this finding, external radiotherapy (ERT) in mediastinal region was indicated. The ERT consisted of a dose of 36 Gy in 6 fractions over.

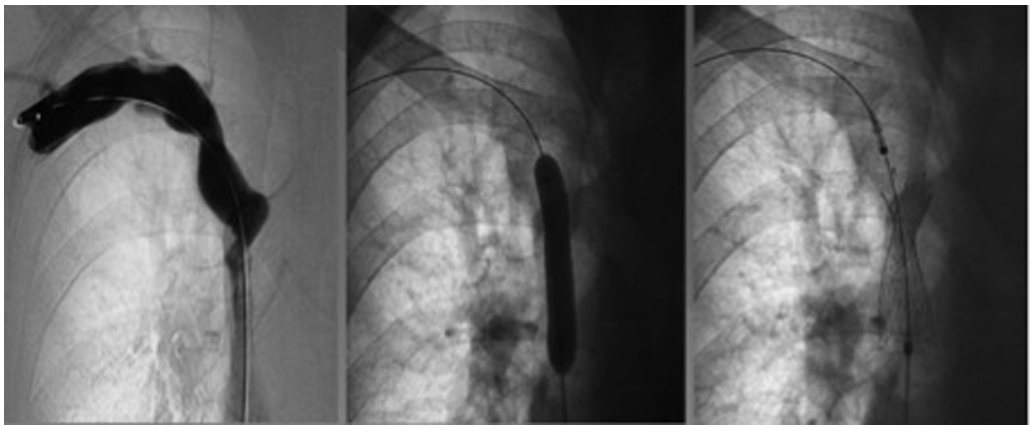

Fig. 2. Angiography shows SVC stenting in our patient.

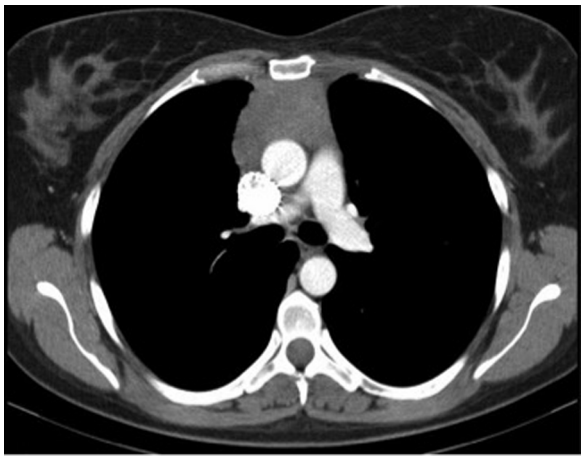

Fig. 3. Postcontrast CT images at the level of crossing of left brachiocephalic vein and arch of aorta showing regression of tumor mass (January, 2011).

Three months later, we performed positron emission tomography, which showed complete metabolic remission of the disease Fig. 4. At present the patient is in haematology outpatient care with no signs of relapse. In December 2013, she gave birth to healthy twins.

3. Discussion

SVCS is obstruction of blood flow through the SVC. It is a medical emergency and most often manifests in patients with a malignant disease process within the thorax. A patient with SVCS requires immediate diagnostic evaluation and therapy. Intrathoracic malignancies are responsible for 60–85% of SVCS cases. SVC obstruction is the presenting symptom of a previously undiagnosed tumor in up to 60% of these cases [6,7]. Non-small-cell lung cancer (NSCLC) is the most common malignant cause of SVCS, accounting for 50% of all cases, followed by small-cell lung cancer (SCLC) (25%) and NHL (10%) [6]. Lung cancer and NHL account for approximately 95% of all malignant SVCS cases [6]. In young age, malignant lymphoma is the leading cause of SVCS. HL involves the mediastinum more commonly than any other lymphoma but rarely causes SVCS [5].

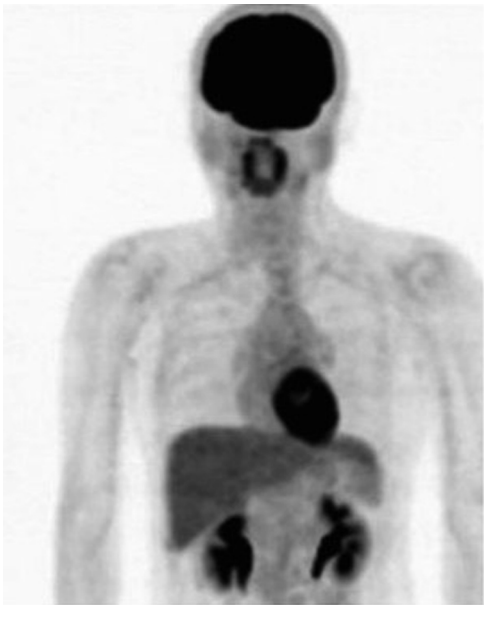

Fig. 4. Positron emission tomography showed complete metabolic remission of the disease (August, 2012).

The diagnosis of a large mediastinal mass can be made on a conventional chest radiograph. For a more detailed visualization of the SVC and its surrounding structures, a chest CT with intravenous contrast medium in the venous phase is recommended. Alternatively, magnetic resonance imaging (MRI) with MRI phlebocavography can be performed [8].

The underlying disease determines the treatment of SVCS, so the cornerstone of the treatment of thoracic malignancies is accurate diagnosis. It is desirable and most of the times possible to make a histological diagnosis of malignancy before going for treatment. Treatment without an established tissue diagnosis should be initiated only when the symptoms are rapidly progressive or multiple previous attempts to make a tissue diagnosis have failed [9]. In addition, cytological methods such as protected specimen brush, bronchial washing, or bronchoalveolar lavage tend to increase the diagnostic yield [9].

The primary goal in SVCS management is alleviation of symptoms and treatment of the underlying disease. Supportive measures like head end elevation, oxygen inhalation and diuretics are helpful in patients of SVCS due to any cause, dexamethasone also provides symptomatic relief by decreasing oedema and tumour burden in malignant causes of SVCS. In presence of acute thrombus thrombolysis is indicated [10]. Catheter induced SVCS can be managed with the help of anticoagulants, thrombolysis and endovascular treatment [11]. The placement of stent in vena cava superior can restore venous return with following prompt symptom relief [12]. There are still a few unanswered questions regarding interventional technique and medication before and after stent placement. Some authors recommend pre-dilatation of the vein before stent placement, and others perform primary stenting followed by post-dilatation [13,14]. The anticoagulation therapy is also controversial, and there is no consensus about it due to bleeding risk. Some recent results suggest that there are no differences between patients who received anticoagulation therapy and/or aspirin and patients who do not receive scheduled anticoagulation therapy in terms of stent thrombosis [15]. The majority of patients record complete or significant disappearance of head and upper extremity oedema, followed by reduction of headache, dyspnoea, and collateral venous circulation [15,16]. The systemic review from Rowell and Gleeson [17] is the biggest collected data which show symptom relief in 95% of the patients treated with stent placement and relapse in up to 11%. Complications after stent insertion are relatively rare and include stent fracture and migration (10%), pulmonary emboli, and local groin hematoma, but several fatal cases are reported after intervention with pericardial tamponade [18].

HL is a B cell-derived lymphoid malignancy with an incidence of 2–3/100000/year. Young adults aged 20–40 are most often affected [19]. Most patients of HL present with palpable lymphadenopathy that is non-tender, in most patients these lymph nodes are in the neck, supraclavicular area and axilla [20]. More than half the patients will have mediastinal adenopathy at diagnosis and this is sometimes the initial manifestation [20]. Despite this common occurrence of mediastinal involvement in HL SVCS rarely occurs [5]. One-third of the patients present with fever, night sweats and/or weight loss. Due to the development of highly active multiagent chemotherapy regimens such as ABVD as well as the optimization of radiotherapy field and doses, HL has become highly curable and about 80% of patients remain relapse-free after adequate first-line treatment.

4. Conclusion

SVCS is a medical emergency; most often manifests in patients with a malignant disease process within the thorax. HL involves the mediastinum more commonly than any other lymphoma but

rarely causes SVCS. Therefore, pathological confirmation of diagnosis should be done before initiating therapy while dealing with a case of SVCS. SVC stenting is effective treatment and has few complications in patients with SVCS.

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal. All identifying details have been removed.

Author contributions

J. Hamzik, J. Chudej, A. Dzian, J. Sokol, P. Kubisz performed all procedures, obtained the patient's written informed consent to publish the report, conducted the follow-up examinations, analyzed and interpreted the patient data, and wrote and edited the manuscript. J. Hamzik was a major contributor to reviewing and editing the manuscript. P. Kubisz contributed to the review and editing of the manuscript. All authors read and approved the final manuscript.

Conflict of interest

The authors declare that they have no competing interests.

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