When the stenting in the approach to superior vena cava syndrome allows radical treatment of non-small cell lung cancer at the diagnosis

Quando o stent *na abordagem do síndrome da veia cava superior permite o tratamento radical do cancro de pulmão de não pequenas células ao diagnóstico*

Inês Hilário Soldin[1,*], Ana Luísa Nunes[2], Paulo Dias[3], Fausto Sousa[4], Fernanda Estevinho[5]

[1] Interna de Formação Específica em Oncologia Médica, Instituto Português de Oncologia do Porto.
[2] Assistente Hospitalar de Medicina Interna, Hospital Pedro Hispano, Unidade Local de Saúde de Matosinhos.
[3] Assistente Hospitalar Graduado de Angiologia e Cirurgia Vascular, Centro Hospitalar de São João.
[4] Assistente Hospitalar de Radioncologia, Centro Hospitalar de São João.
[5] Assistente Hospitalar Graduada de Oncologia Médica, Hospital Pedro Hispano, Unidade Local de Saúde de Matosinhos.*
ineshsilva@gmail.com

RESUMO

O síndrome da veia cava superior é uma emergência oncológica. Em 15% dos casos causa risco imediato de vida. Quando é a forma de apresentação da doença oncológica e apresenta critérios de gravidade, a abordagem diagnóstica e terapêutica multidisciplinar é desafiante e urgente. Apresentamos o caso de um doente sem diagnóstico oncológico prévio, com síndrome da veia cava superior. Apresentava edema da face, região cervical e membros superiores, dispneia e alteração do estado mental. Realizada tomografia computadorizada de tórax, que evidenciou um síndrome da veia cava superior secundário a massa mediastínica. Considerando a gravidade da obstrução, ausência de diagnóstico histológico e de metastização no restante estudo imagiológico realizado no serviço de urgência, optou-se pela colocação de *stent* endovascular de urgência, com rápida melhoria clínica. Confirmou-se o diagnóstico de adenocarcinoma pulmonar estadio IIIA. Após resposta parcial a quimioradioterapia, iniciou terapêutica de consolidação com durvalumab. Sem recidiva seis meses após o diagnóstico.

Palavras-chave: Cancro do pulmão de não pequenas células; síndrome da veia cava superior; stent endovascular; radioterapia

ABSTRACT

Superior vena cava syndrome is an oncological emergency. It is life-threatening in 15% of the cases. When it is severe and the presentation form of malignancy, a multidisciplinary diagnostic and therapeutic approach is challenging and urgent. We present the case of a patient with no previous cancer that came to the emergency department due to superior vena cava syndrome. He had oedema of the face, cervical region and upper limbs, dyspnoea and altered mental status. Chest computed tomography showed a superior vena cava syndrome secondary to a mediastinal mass. Considering the severity of the obstruction, the absence of histological diagnosis and distant metastasis, an endovascular stent was placed urgently, with rapid clinical improvement. Subsequently, we obtained a diagnosis of stage IIIA lung adenocarcinoma. After partial response to chemoradiotherapy, the patient started consolidation therapy with durvalumab. Six months after the diagnosis, there is no relapse.

Key-words: Non-small cell lung cancer; superior vena cava syndrome; endovascular stent; radiotherapy

INTRODUCTION

Superior vena cava syndrome (SVCS) results from obstruction of blood flow through the superior vena cava (SVC). An intrathoracic malignancy is responsible for 60 to 85% of the cases. Lung cancer and non-Hodgkin lymphoma are the most frequent etiologies.[1-3] SVC obstruction is the presenting symptom of a previously undiagnosed tumour in up to 60% of the patients.[1] Early symptoms include cough, dyspnea, dysphagia, dysphonia, jugular vein distention, plethora, and arm, neck and face oedema. If not promptly treated, potentially fatal respiratory distress and cerebral oedema develop.[1-3]

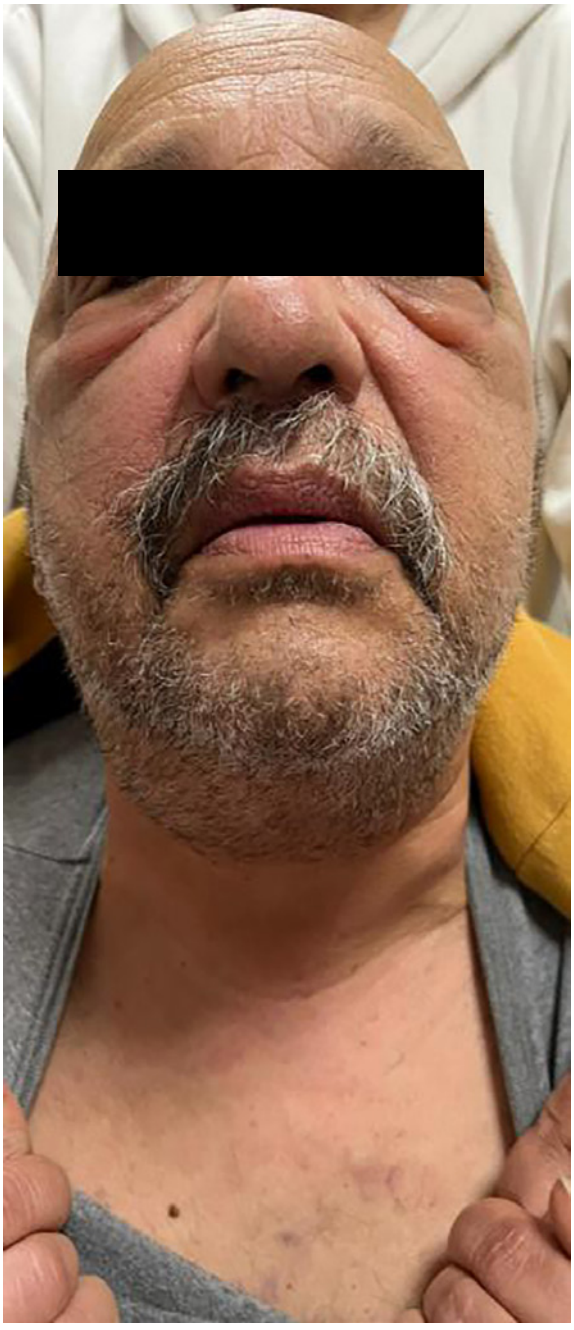
Contrast-enhanced computed tomography (CECT) is the initial diagnostic modality. The exam defines the extent of venous obstruction and collateral venous network and identifies potential thrombosis.[3] Concerning treatment, the primary goal is to alleviate symptoms. Secondary goals and intervention depend on the histology, staging and prognosis.[1,4] Treatment is guided by general recommendations since there are no evidence-based guidelines. The urgent stenting applies to severely symptomatic patients. Stenting will also be preferable when SVCS is the presentation form of an undiagnosed malignancy. Otherwise, radiotherapy (RT) could interfere with histological characterization, compromising subsequent treatment, and would not be effective in radioresistant tumours. Supportive treatment with glucocorticoids can be effective in steroid-responsive malignancies, such as lymphoma, or if the airway is compromised. However, it can hinder histological characterization. The use of diuretics is not consensual.[4-6]

CASE DESCRIPTION

Caucasian male patient, 66 years old, ex-smoker (55 pack-years), with ECOG 1 and a history of hypertension and obesity.

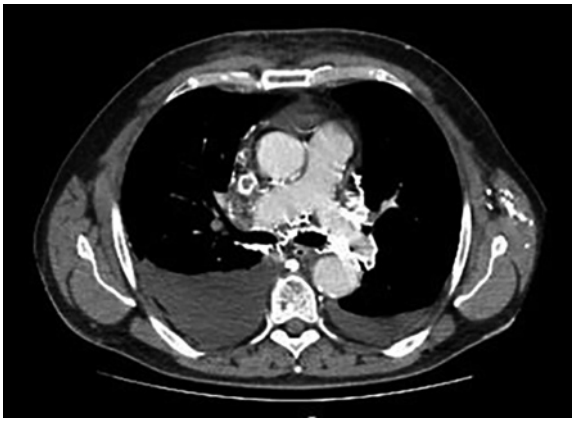
The patient presented to the emergency department due to worsening facial, neck and upper extremities oedema that started progressively for two months. Enlarged tortuous superficial bluish-

Figure 1. Facial plethora, periorbital, face and neck oedema; collateral vascular network on the chest wall



-coloured veins grew over the chest wall. He also complained of exercise dyspnoea, orthopnoea, psychomotor retardation, confusion, and sleepiness. On admission, a physical exam revealed non-pitting periorbital, face, neck and arms oedema, plethora, jugular vein distention and collateral circulation in the upper part of the trunk (Figure 1). He was hemodynamically stable, without respiratory distress or desaturation (SpO2 97%). Chest CECT scan revealed a mass with a 5,6 cm longest diameter located anteriorly to the trachea and on the right of the aorta, invading the SVC, that was almost completely obliterated (Figure 2). CT also evidenced a collateral venous network and a pleural effusion but no secondary lesions. Abdominal, pelvic and brain CECT scans excluded metastasis. Malignancy initially presenting as a grade 3 SVCS was assumed. The case was discussed by a multidisciplinary team involving medical and radiation oncology and vascular surgery physicians. Since the mass was not histologically characterized, endovascular treatment was the preferred option. Due to the severity of the

Figure 2. Obliteration of the superior vena cava, right brachiocephalic vein and proximal portion of the left brachiocephalic vein. Collateralization to the azygos system. Pleural effusion.



presentation, the procedure occurred in the first 24 hours. An angiogram was first performed demonstrating that occlusion extended from SVC to the brachiocephalic veins (Figure 2), causing extensive collateralization of azygos and hemiazygos veins, and excluding associated thrombosis. An Y-shaped stent deployment was chosen: three self-expanding nitinol stents were used from the SVC to the braquiocephalic veins in a kissing (or double barrel) technique (Figure 3). Due to the severity of SVC obstruction and the risk of stent thrombosis, the patient started low-molecular--weight heparin. In the first 24 hours, oedema has substantially diminished. Dyspnoea and orthopnoea improved in a few days.

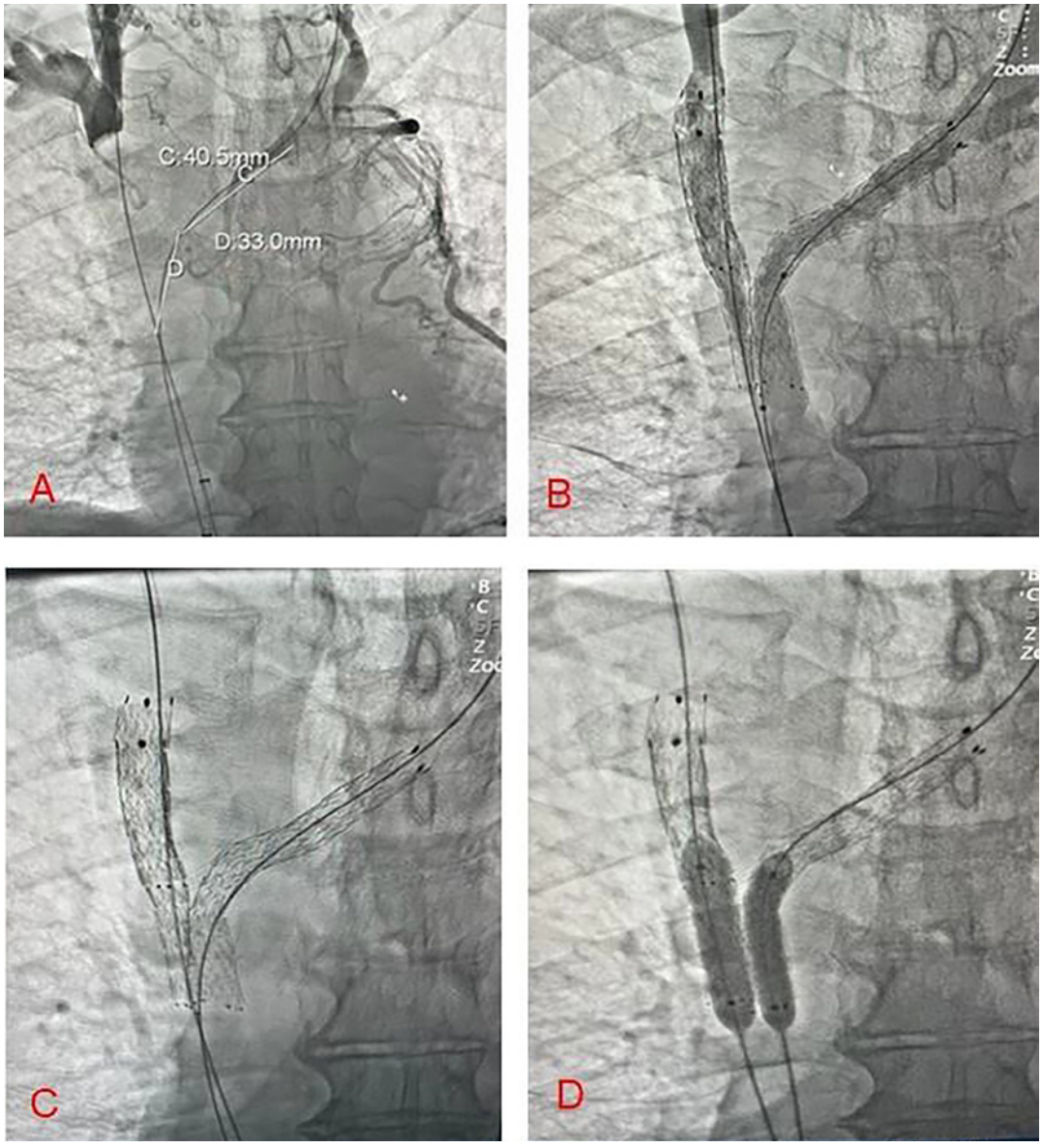
Posteriorly, an endobronchial ultrasound-guided transbronchial needle aspiration established the diagnosis of lung adenocarcinoma with PD-L1 expression of 15-20%. Next Generation Sequencing did not identify clinically significant mutations. Pleural effusion citology was negative for malignant cells. Normal positron-emission tomography completed staging. Cancer was staged as cT4N0M0 (stage IIIA). As a treatment, the patient received five cycles of radiosensitizing weekly chemotherapy with carboplatin and paclitaxel (full-dose), performed along with daily radiotherapy with curative intent (total dose of 66Gy/33fr, by photon therapy and volumetric modulated arc therapy). He obtained a partial response as the treatment's best response. After chemoradiation, he started durvalumab (10mg/kg Q2W) as a consolidation treatment. After six months, the patient is clinically stable, with an ECOG 1 and no relapse.

DISCUSSION

To stratify SVCS severity, Yu et al. proposed a grading system to stratify symptoms and to determine the urgency of intervention, facilitating communication between physicians. This system assesses the degree of oedema (laryngeal or cerebral) and hemodynamic status to categorize SVCS between life-threatening (grade 4), severe (grade 3), and non-life-threatening cases (grade 0-2).(7) Initial management depends on the underlying malignancy, the expected response to treatment and the severity of symptoms. A multidisciplinary team should decide the best treatment.(5) When malignancy initially presents as SVCS additional challenges have to be faced.

RT was formerly the gold standard for treating SVCS because lung cancer and lymphoma are characteristically radiosensitive. However, the efficacy of RT is about 80%, taking up to two weeks to improve symptoms. Furthermore, performing high-dose fractions (3-4Gy/day) will impede posterior chemoradiation with curative intent. Therefore, endovenous recanalization is nowadays considered the standard of care for patients with malignancy-related symptomatic SVCS. Few areas of venous disease provide a more satisfying experience for both the patient and the vascular specialist. Relief from severe, frequently incapacitating symptoms of venous congestion of the head and neck is almost instantaneous, and benefit is generally long lasting, as reported here. Additionally, stenting has a high technical success rate (95%-100%), efficacy rate (over 90%), as well as a low complication (less than 8%) and relapse rates (10.5% on average).[8] Besides RT, stenting does not interfere with subsequent treatment lines. Therefore, the absence of histological diagnosis determined the selected approach. In the absence of thrombosis, the decision of starting anticoagulation, which medication and duration of treatment after stenting depends on individual patient risk-benefit relationship.[3,6]

Figure 3. The extent of SVC occlusion in angiography (A); Double barrel stenting with self-expandable stents (Optimed Sinus Venous: 14x60 mm + 14x80 mm+ 14x40 mm) deployed as “kissing stents” in the SVC and both brachiocephalic veins (B, C, D).
In order to deploy stents, a left Internal jugular vein and right common femoral vein accesses were obtained, since it was not possible to cross the lesion via femoral access. Right internal jugular vein access and through-and-through (RIJV- RCFV) with GW Terumo stiff to the femoral access was used to overcome the SVC compression. Vessel was then prepared and a pre-dilatation with 10x40 mm balloon was performed. After stenting, a 10x80 balloon was used to redilate vessel. Final angiogram showed restored patency of both brachiocephalic veins and superior vena cava.



After stenting, our patient's treatment protocol was based on the phase III PACIFIC trial. The trial showed that consolidation durvalumab was associated with significant improvements in overall survival and progression-free survival, with manageable safety.

In conclusion, this clinical case illustrates how SVCS can be a true oncologic emergency and how essential is a multidisciplinary discussion. In this case, it allowed an endovascular approach in an urgent setting, not compromising subsequent treatment with chemoradiation and immunotherapy. Those treatments significantly improve morbidity and mortality in advanced-stage lung cancer.(4)

ORCID

Inês Hilário Soldin 0009-0008-5151-3259
Ana Luísa Nunes 0000-0002-5989-4219
Paulo Dias 0009-0009-8465-1620
Fausto Sousa 0000-0003-1756-9351
Fernanda Estevinho 0000-0002-2694-3993

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