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Superior vena cava syndrome due to a leiomyosarcoma of the anterior mediastinum: A case report and literature overview

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ABSTRACT

INTRODUCTION: Leiomyosarcomas are an infrequent cause of malignant superior vena cava syndrome (VCS).
PRESENTATION OF CASE: A 51-year old male patient was admitted for a three-day history of dyspnoea, dysphagia and erythema of the head and neck. Computed tomography and magnetic resonance imaging showed a lesion arising on the anterior mediastinum, which was in close proximity with a thrombus in the superior vena cava. Surgical excision was performed, including open resection of the primary tumour and an atrio-innominate vein bypass with 8-mm polytetrafluoroethylene (PTFE). Histology confirmed a leiomyosarcoma and postoperative radiotherapy sessions were performed. Due to evidence of enlargement of the thrombus, a second intervention was undertaken. In this procedure, a remainder of the primary tumour was resected and the superior vena cava reconstructed with an autologous pericardium patch. The patient recovered satisfactorily and was discharged on the seventh postoperative day, with no evidence for relapse after 10 months of follow-up.
DISCUSSION: Leiomyosarcomas comprise less than 2% of the tumours of the mediastinum and are a rare cause of paraneoplastic VCS. Male patients in their sixties are most commonly affected. Relapses seem to be common, and thus a careful follow-up is often recommended.
CONCLUSION: In spite of the limited data on the management of thoracic leiomyosarcomas, surgery is currently considered the mainstay of treatment.

1. Introduction

Soft tissue sarcomas are a heterogenous group of mesenchymatic tumours. Current estimations indicate that they constitute less than 1% of all neoplasms [1,2]. They can occur in various locations, but the most frequent ones are the lower extremities, thorax, and the head and neck region [3].

Amongst the sarcomas of the mediastinum, the rare leiomyosarcoma is characterised by the differentiation of smooth muscle cells [4]. Leiomyosarcomas preferentially occur in the abdominal cavity and retroperitoneum [1]. Those that arise in the mediastinum originate from thoracic organs, such as the atrium, oesophagus and large blood vessels [3].

When associated with malignancies, the superior vena cava syndrome (VCS) is most often associated with lung carcinomas, lymphomas and distant-organ metastases [5]. We present a case of VCS due to a leiomyosarcoma in the anterior mediastinum.

2. Presentation of case

A 51-year old male, without any known comorbidities, attended the emergency department due to a 3-day history of worsening dyspnoea, dysphagia, oedema and erythema of the head and neck. The physical examination showed significant oedema, facial erythema and engorgement of the jugular veins, without any evidence of haemodynamic instability.

Routine laboratory tests were unremarkable. A computed tomography (CT) scan of the thorax revealed a mass lesion in the upper anterior mediastinum, with a near-complete thrombosis of the superior vena cava (SVC). Images also showed signs compatible with a partial thrombosis of the brachiocephalic trunk (Fig. 1).

The patient was admitted to an intermediate care unit. Vascular magnetic resonance imaging (MRI) confirmed the findings seen in the CT and allowed the diagnosis of a paraneoplastic thrombosis of the SVC to be made. A course of non-fractionated heparin

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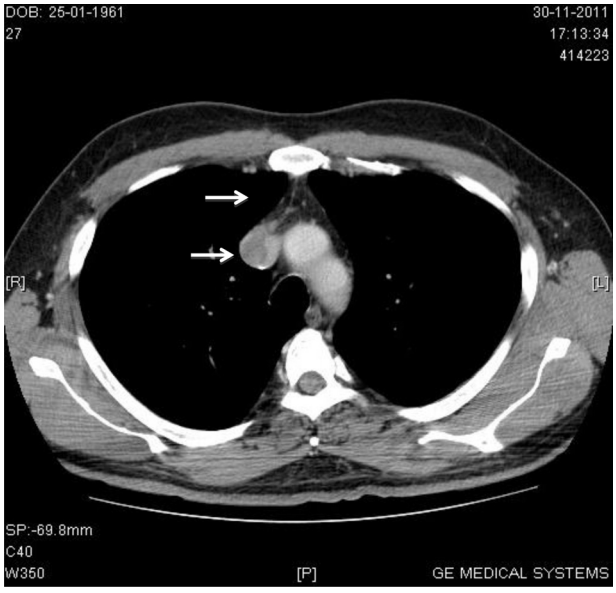

Fig. 1. CT Scan showing a tumorous lesion affecting the upper anterior mediastinum (upper arrow). Luminal occupation of the superior vena cava is also shown (lower arrow).

was started, which reduced the symptoms only partially, and thus surgical exploration was recommended. The procedure aimed at resecting the main tumour, obtaining biopsies and performing a brachiocephalic vein (or innominate vein) derivation to the atrium.

The surgical approach was performed in several stages. First, an opening of the sternum and pericardium was undertaken to allow the dissection of the upper mediastinum. Then, an en-bloc resection of tissue was performed and followed by the loosening of the innominate vein. An intraoperative biopsy identified the tissue as an involute thymus. A round, hard, wood-like structure of 3 by 4 cm was found next to the upper right corner of the thymus, with several dilated veins with clear signs of thrombosis in its close proximity. The lumen of these veins contained a tumour-like tissue that infiltrated the SVC, which had a similar rounded lesion occupying its entire lumen as well. A rapid biopsy showed a fibrous neoplasm made of fusiform and atypical cells with central fibrosis. Complete resection of the lesion was then performed and the innominate vein anastomosed to the right atrium through the atria.

The patient had a favourable course after surgery and recovered satisfactorily. A definitive histological examination of the surgical specimen showed skeletal muscle and a neoplasm made of fusiform cells, which was compatible with an advanced leiomyosarcoma. Radiation therapy was suggested, and thus 50.4 Gy of tridimensional radiation on the surgical site, mediastinum and SVC were administered over five and a half weeks.

Once the RT sessions were concluded, a follow-up MRI revealed a modest increase in the original thrombus, and thus a second intervention was scheduled.

Upon opening of the sternum, significant fibrosis and tissue adherences to the mediastinal wall were seen and removed with some difficulty. A transversal section at the level of the sternomanubrial joint was performed. The SVC was proximally and distally occluded, and a ligature of the azygos vein was conducted. Formal exploration of the cava was undertaken after this step. Inside, originating from the innominate vein junction, a white, smooth and seemingly necrotic tumour of 3.5 by 3 cm was found. It was firmly attached to the lateral wall of the cava. The lesion was resected along with the medial wall of the cava and a pericardium patch was used for its subsequent repair (Fig. 2).

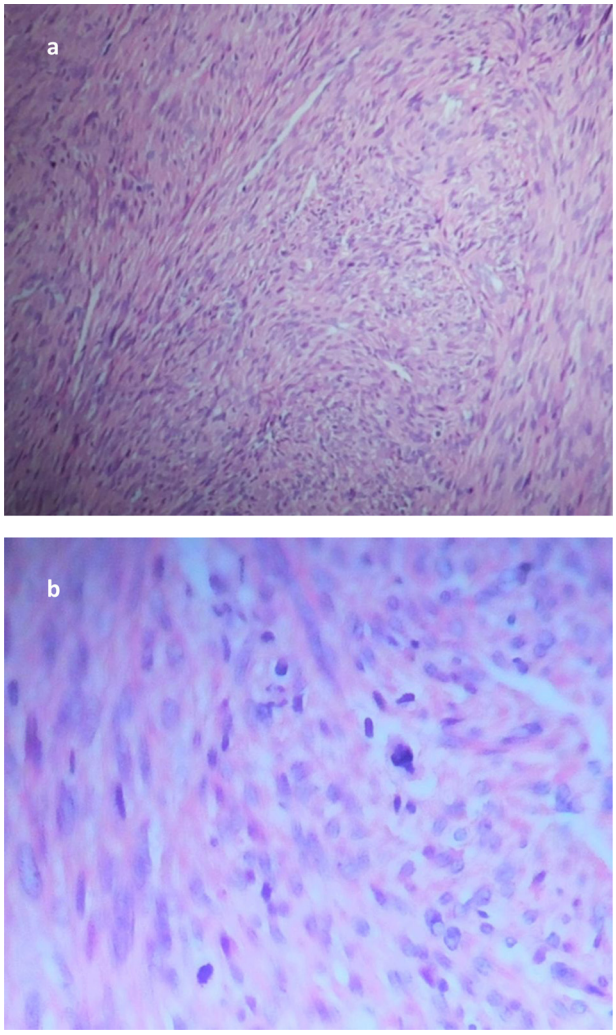

Fig. 2. Microphotography of the neoplasm (Haematoxylin–Eosin, 10× and 40×), showing a tumour comprised of fusiform cells (2a) with an abundant cytoplasm, markedly pleomorphic nuclei and some atypical mitoses (2b).

The patient was discharged seven days after the procedure. A definitive histological examination showed an extensively necrotic tumour comprised of fusiform cells and nested within the vessel walls. Immunohistochemical analyses showed a strong positivity for vimentin and actin, thus confirming the diagnosis of a high-grade leimyosarcoma.

3. Discussion

We present the case of a 51-year old man who presented with a VCS due to a leiomyosarcoma of the mediastinum. As shown by previous reports [6,8–10], the initial clinical presentation is heterogenous and nonspecific. Cough, dyspnoea, chest pain and fever have often been cited as initial manifestations [2,6]. Neurologic and haemodynamic alterations, including syncope, confusion and blurred vision can constitute presenting symptoms as well. These neoplasms may attain significant sizes and display mass-related clinical features. Among these, evidence of vascular obstructions, such as aortic compression, thrombosis or VCS with facial and neck oedema, have been previously reported [7,9,11]. However, it should be pointed out that amongst the malignant causes of VCS, leiomyosarcomas are considered extremely rare, with lung cancer or lymphomas representing far more likely explanations for the

syndrome. Less than 30 cases of primary leiomyosarcoma of the anterior mediastinum are currently available in the literature [2,6].

Plenty of controversy exists regarding the origin of this neoplasm. It has been suggested that leiomyosarcomas could arise from the walls of small vessels located within soft tissues of the mediastinum. Alternatively, these tumours could originate from heterotrophic smooth muscle cells derived from the oesophagus, mediastinal veins or a displaced spleen [10]. Histologically, leiomyosarcomas often show features that resemble thymomas, thymic carcinomas, malignant schwannomas, malignant histiocytomas and other solitary fibrous tumours of the mediastinum [12]. Therefore, the use of immunohistological staining, such as antibodies for smooth muscle actin, desmin and vimentin, may be required for the proper identification of these neoplasms.

Because of this neoplasm's rarity, clinical features have been difficult to ascertain. Mean age at diagnosis seems to be around the fifth or sixth decade of life, and a male preponderance has been described [2]. In 1994, Moran et al. [8] presented data from 10 patients diagnosed with mediastinal leiomyosarcomas. In this seminal case series, the median age was 56 years (range 21–76 years), and a male to female ratio of 7:3 was observed. The only treatment was surgical excision, and recurrences were usually observed 2–3 years after diagnosis. Five-year survival rates of 15–20% have been reported only among patients that received complete surgical excision [13]. Although no definitive conclusions can be drawn from case series, these data have traditionally been used to establish surgery as the treatment of choice for these patients. This contradicts a common conception that mediastinal neoplasms that show superior vena cava infiltration are not candidates for surgical resection [14,15].

The surgical approach depends on the degree of tumorous invasion of vascular and adjacent structures. In cases where only a small segment of the vessel is affected, it is possible to partially occlude the vessel and then perform resection of the neoplasm. However, if the resection is expected to be large, and autologous pericardium patch might be necessary to correct the defect. In extreme cases, prosthetics might be required. While there are no firm recommendations regarding specific techniques, it seems that defects of less than 50% of the vena cava's circumference can be effectively repaired with conservative approaches, such as clamping and the use of pericardium patches. For defects that exceed this magnitude, a thorough resection of the cava with prosthetic replacement is commonly suggested [16,17]. There are multiple options when selecting prosthetic materials, but PTFE is the most frequently used. Smaller devices (8–10 mm) are usually preferred to optimise perfusion [18], but because of the extensive damage that was seen in this case, a bypass from the atrium to the innominate vein was performed prior to tumour resection during a second surgery.

An important consideration is to ensure adequate brain and systemic oxygenation. Clamping will have different effects depending on whether the cava is occluded and the presence of collateral veins. Consequences can be dire if the cava is not occluded or if there's insufficient development of collaterals. Another further consideration in this surgery is the decrease of cardiac volumes, which often results in a decrease of the systemic blood pressure accompanied by an increase in cerebral venous pressure and consequent brain oedema. In these cases, the bypass allows constant venous return to the right atrium, thus preventing the development of neurological complications. The procedure also eliminates the time constraints imposed by clamping of the superior vena cava and helps making surgical repairs easier.

4. Conclusions

Mediastinal leiomyosarcomas are exceedingly rare neoplasms. Clinical presentation is heterogeneous and dependant on size and site of presentation. Patients with a localised tumour might benefit from surgical excision and reconstruction. Current surgical approaches depend on the neoplasm's size and the invasion of neighbouring structures. Due to the special considerations entailed in this kind of procedure, the decision to perform surgery must be tailored to the needs of each particular patient.

Conflict of interest

None to disclose

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Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Eduardo Labarca, Alvaro Zapico and Mario Santamarina gathered the data and drafted the manuscript. Dr. Rios and Dr. Martinez wrote the final revision of the report. All authors reviewed the manuscript and agreed on the contents prior to its submission.

Key learning points

- Leiomyosarcomas are rare causes of superior vena cava syndrome.
- Surgery is the treatment of choice.
- Specific approaches depend on the neoplasm's size and invasion of neighbouring tissues.

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