Mediastinal cavernous hemangioma originating from the right superior vena cava. A case report

Luis Roberto Jimenez Hernandez M.D.
Rodrigo Daniel Urbina Ulloa M.D.
Julio César Cortes Carmona M.D.
Santiago Fajardo Arnaud M.D.

Mexico City, Mexico

Case Report

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Background: Mediastinal hemangiomas are extremely rare benign vascular neoplasms, accounting for less than 0.5% of all diagnosed mediastinal masses. Primary origin from or direct involvement of the walls of the right superior vena cava (SVC) is exceptionally uncommon. We present the case of a 31-year-old man with a 6-month history of recurrent upper respiratory tract infections. Chest radiography showed displacement of the tracheal midline, and noncontrast computed tomography demonstrated a large anterior mediastinal tumor arising from the right SVC. Complete surgical excision was performed through an open transthoracic approach to ensure safe vascular control. Intraoperative findings included abundant thymic fat, multiple dense fibrous adhesions, and a 5 × 6 cm tumor involving the superior jugular vein, right pleura, and pericardium, as well as periaortic neovascularization. Definitive histopathologic examination confirmed a cavernous hemangioma with dystrophic calcification arising in residual thymic and adipose tissue and showed positive surgical margins (R1 resection) in the specimens from the mediastinal tumor, the SVC extension, and the periaortic tissue. Because of the absence of a true fibrous capsule and the lesion's intimate fusion with the adventitia of the great vessels, achieving microscopically negative margins (R0) is highly challenging when preservation of vascular structural integrity is prioritized. Therefore, after an R1 resection, although the benign nature of the lesion precludes metastatic potential, strict long-term clinical and imaging surveillance with serial CT scans is required to monitor for possible local recurrence.

Keywords: Cavernous hemangioma, Anterior mediastinum, Superior vena cava, Surgical margins, Vascular control.

A 31-year-old man with a 6-month history of recurrent upper respiratory tract infections and no relevant medical history underwent chest radiography, which showed leftward displacement of the tracheal midline. Noncontrast chest computed tomography demonstrated an anterior mediastinal tumor in close relationship with the superior vena cava. The findings were considered compatible with a large hemangioma arising from the right superior vena cava, and a preoperative protocol was initiated to complete its resection.

Surgical findings

The following findings were documented: abundant thymic fat, multiple fibrous adhesions between the pericardium and aorta, the innominate vein and right pleura, and the jugular vein and right pleura; a 5 × 6 cm mass involving the superior jugular vein, right pleura, and pericardium; and periaortic neovascularization. A 28-Fr retrosternal drain and a 24-Fr cardiorespiral retrocardiac drain were placed.

Histopathologic diagnosis

1. Biopsy specimen submitted as thymic fat: adipose tissue and residual thymic tissue with dystrophic calcification, without evidence of malignancy.
2. Biopsy specimen submitted as mediastinal tumor: adipose tissue and residual thymic tissue with dystrophic calcification and cavernous hemangioma involving the surgical margins.
3. Biopsy specimen submitted as extension of the superior vena cava tumor: ADIPOSE TISSUE WITH CAVERNOUS HEMANGIOMA involving the surgical margins.
4. Biopsy specimen submitted as periaortic tissue: fibroadipose tissue with cavernous hemangioma involving the surgical margins.

Discussion

Mediastinal hemangiomas are extremely uncommon benign vascular neoplasms, accounting for less than 0.5% of all masses diagnosed in the mediastinum

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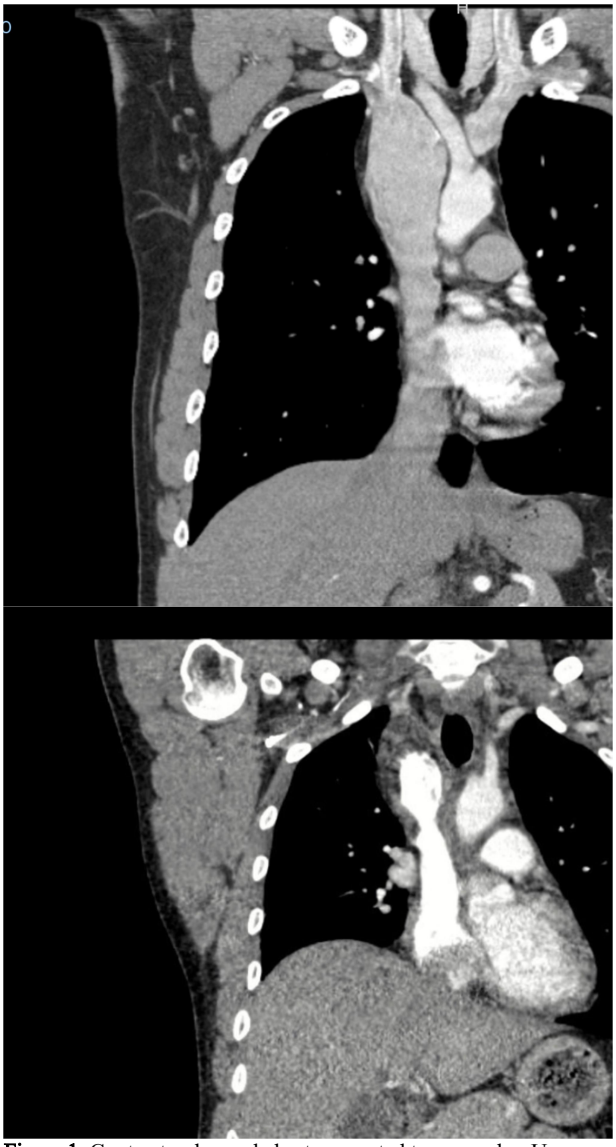

Figure 1. Contrast-enhanced chest computed tomography. Upper: Preoperative coronal CT. Lower: Postoperative coronal CT.

[1, 6]. Histologically, cavernous hemangioma is one of the variants reported within this spectrum of vascular anomalies, together with other subtypes occurring in the mediastinum, including capillary hemangioma, hemolymphangioma, cystic hemangioma, infantile hemangioma, and the recently characterized anastomosing hemangioma [2, 3, 5, 8, 9]. Although most of these lesions develop within the anterior mediastinal compartment [7], primary origin from or direct involvement of the walls of major venous structures, such as the right superior vena cava (SVC), is exceptionally uncommon. The medical literature more frequently describes cases located in the posterior mediastinum or mimicking masses in other cavities [1, 2, 3].

The clinical presentation of mediastinal hemangiomas is directly influenced by the size of the mass, its location, and its anatomic relationship with

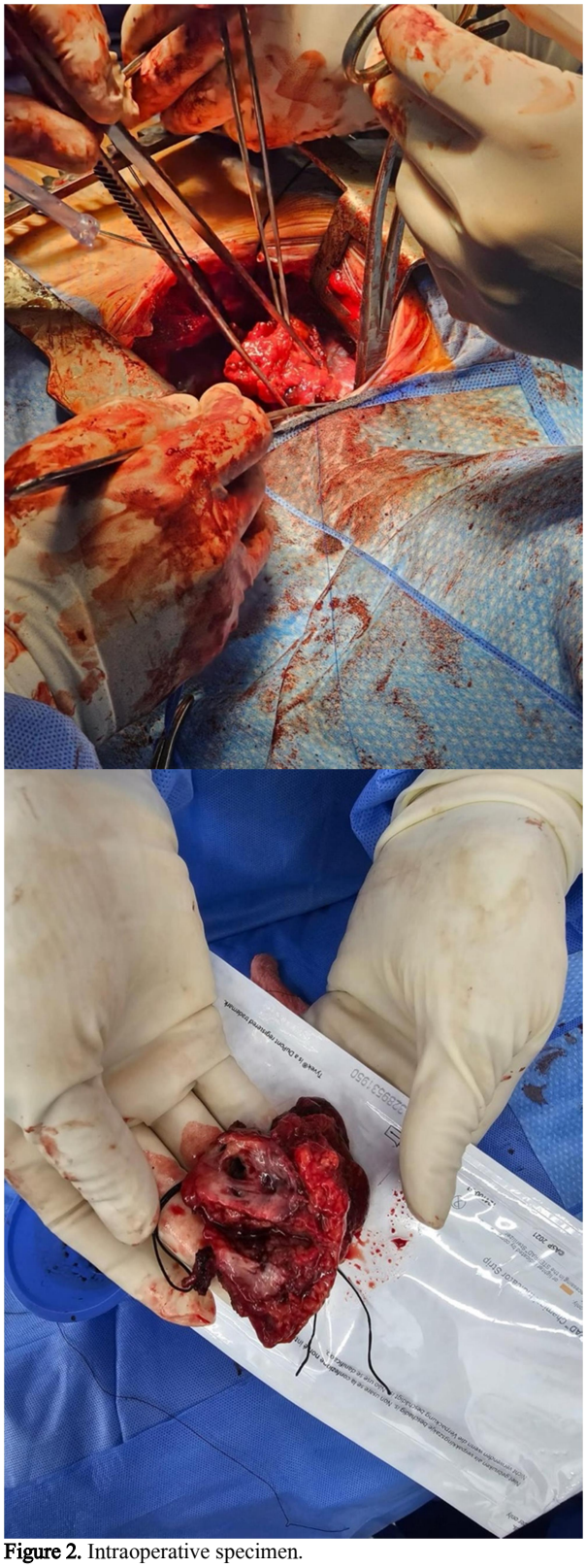

Figure 2. Intraoperative specimen.

adjacent structures [1, 5]. A substantial proportion of patients are asymptomatic, and the tumor is identified incidentally on imaging studies obtained for other reasons [6, 8]. By contrast, symptomatic patients usually present with findings related to mass effect or

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local compression of the airway, osseous, neural, or vascular structures, including chest pain, dyspnea, or even severe compressive syndromes such as spinal cord compression in posterior lesions [5, 6, 7]. In the present case, the patient had an atypical presentation consisting of recurrent upper respiratory tract infections over a 6-month period. This presentation highlights the initially nonspecific nature of respiratory symptoms in young adults, which frequently delays suspicion of a space-occupying mediastinal mass.

Imaging studies play a crucial role in the preoperative diagnostic algorithm. Initial chest radiography may demonstrate indirect signs of a space-occupying lesion. In our patient, it showed marked leftward displacement of the tracheal midline caused by the tumor volume. Chest computed tomography (CT) is a fundamental tool for the anatomic characterization of these lesions because it allows delineation of tumor extent, assessment of a possible vascular origin, and definition of the relationship with adjacent vessels [1, 6]. In this case, noncontrast CT provided findings highly compatible with a large hemangioma arising from the right superior vena cava, thereby facilitating advance planning of the surgical approach.

The definitive treatment of choice for mediastinal hemangiomas is complete surgical excision, with the goals of relieving symptoms, obtaining histologic confirmation, and preventing potentially serious complications such as irreversible compression, intratumoral hemorrhage, or spontaneous rupture [5, 6, 8]. Nevertheless, the operation is technically complex because of the highly vascular nature of the tissue and its marked tendency to induce a local fibrous adhesive response. The intraoperative findings documented in this case included abundant thymic fat, multiple dense fibrous adhesions between the pericardium and aorta, the innominate vein and right pleura, and the jugular vein and right pleura; a 5 × 6 cm mass involving the superior jugular vein and pericardium; and periaortic neovascularization. These findings reflect the locally adherent and infiltrative behavior that complicates dissection from these vascular structures [1]. Although some authors have proposed a thoracoscopic approach for selected, well-circumscribed lesions [7], the open transthoracic approach used in this case provided optimal exposure for safe vascular control of the great vessels. This was complemented by the strategic placement of mediastinal and retrocardiac drains—a 28-Fr retrosternal drain and a 24-Fr cardiorespiratory drain—to monitor output during the immediate postoperative period.

Definitive histopathologic examination is the only method capable of confirming the diagnosis and classifying the specific vascular subtype, thereby differentiating it from lymphangiomas, neurofibromas, and other mediastinal neoplasms [2, 3]. Macroscopically, the resected surgical specimens were lobulated, with bright red surfaces and a soft consistency, as well as areas with an adipose appearance. These findings are consistent with the classic morphologic descriptions of cavernous and cystic hemangiomas [8]. Microscopically, this entity is defined by a proliferation of markedly dilated vascular channels lined by a monolayer of mature endothelial cells without atypia and separated by connective and adipose tissue septa [1]. A relevant microscopic finding in this patient was the presence of dystrophic calcification in the tumor and residual thymic tissue. This phenomenon results from prolonged circulatory stasis, organized thrombosis, and subsequent deposition of calcium salts, or phleboliths, within the cavernous dilations of the tumor, a characteristic feature well documented in mediastinal vascular pathology [6].

Finally, a critical aspect of this case is the histopathologic report of “involvement of the surgical margins” in the specimens corresponding to the mediastinal tumor, the superior vena cava extension, and the periaortic tissue. The scientific literature indicates that, because many mediastinal hemangiomas lack a true fibrous capsule and are intimately fused with or continuous with the adventitia of the great vessels or other mediastinal structures, achieving completely negative microscopic margins (R0 resection) is extremely challenging and may occasionally be unfeasible when preservation of the structural integrity of the SVC, aorta, or jugular axis is prioritized [1, 9]. Although cavernous hemangioma is a benign lesion without metastatic potential, involvement of the surgical margins (R1 resection) increases the possibility of persistent disease or local tumor recurrence. Therefore, analysis of this clinical case and the current scientific evidence support the strict need for long-term clinical and imaging surveillance with serial computed tomography scans to closely monitor the patient’s clinical course.

Conclusion

Cavernous hemangioma arising from the right superior vena cava is an exceptionally rare mediastinal vascular neoplasm. Complete surgical excision may be technically challenging because of the lesion’s hypervascularity, dense fibrous adhesions, and intimate relationship with the major mediastinal vessels. Histopathologic examination is required for definitive diagnosis. When microscopically negative margins cannot be achieved without compromising vascular integrity, long-term clinical and imaging surveillance is warranted because of the risk of local persistence or recurrence after R1 resection.

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Rodrigo Daniel Urbina Ulloa
Department of General Surgery
Hospital Regional "Lic. Adolfo López Mateos,"
ISSSTE
Mexico City, Mexico.

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