

Giant teratocarcinoma of anterosuperior mediastinum

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ABSTRACT

Germ cell tumors are not so frequent; among them, teratomas are most common in the mediastinal location. Teratomas in young men are usually malignant (teratocarcinoma), with a gender ratio of 13.5:1, and an average age of 26 years. We describe the evolution of a 19-year-old man with a mediastinal tumor, with pulmonary metastases and intracardiac infiltration, successfully removed; whose most frequent symptoms were: dyspnea, retrosternal pain, fever, cough, weight loss and superior vena cava syndrome. The diagnosis was made by chest x-ray and computed tomography. Total removal of the tumor and metastases was achieved, as well as the intracardiac tumor, with tricuspid valve repair. The patient had a satisfactory outcome during the first follow-up year.

Keywords: Mediastinal neoplasms, Germ cells, Gonadal tissue neoplasms, Ex-tragonadal germ cell tumor

Teratocarcinoma gigante de mediastino anterosuperior

RESUMEN

Los tumores de células germinales no son tan frecuentes, dentro de ellos se encuentra el teratoma como el más usual en ubicación mediastinal, es más habitual en hombres jóvenes cuando es maligno (teratocarcinoma), con una relación por género de 13,5:1, y una edad promedio de 26 años. Se describe la evolución de un hombre de 19 años de edad con un tumor mediastinal, con metástasis pulmonares e infiltración intracardiaca, resecado satisfactoriamente, cuyos síntomas más frecuentes fueron: disnea, dolor retroesternal, fiebre, tos, pérdida de peso y síndrome de vena cava superior. El diagnóstico se realizó por radiografía de tórax y tomografía axial computarizada. Se logró realizar la excéresis total del tumor y las metástasis, así como la tumoración intracardiaca, con reparación de la válvula tricúspide. El paciente ha tenido una evolución satisfactoria durante el primer año de seguimiento.

Palabras clave: Neoplasias del mediastino, Células germinativas, Neoplasias de tejido gonadal, Tumor de células germinales extragonadal

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INTRODUCCIÓN

Mediastinum tumors are not so frequent, with an incidence of 1/100000. Anterior mediastinum tumors are the most common and within them thy-

mus, followed by lymphomas and germ cells tumors, which represent 10 to 15% in adults. Teratomas are most commonly found in young men in the second decade of life. Malignant teratomas are most commonly diagnosed in males. They are derived from cells of the three embryonic layers and are foreign to the area where they form. They have been known to contain tissue such as hairs, nails, teeth, skin, pancreatic tissue, among others. They may be asymptomatic or (if growth is sustained) present with compression symptoms. They originate due to failure in the migration of these cells during pregnancy^{1,2}.

Symptoms vary depending on the size, location and type of tumor; thus, it may constitute a finding on the chest radiograph. Symptoms may include: chest pain, cough, dyspnea, recurrent respiratory infections and dysphagia. General symptoms may be fever, weight loss and general malaise. Some patients present with gynecomastia, due to the production of human beta-gonadotropin^{3,4}.

Germ cell tumors are not as frequent (including mediastinal teratoma) which is most commonly diagnosed in men with a 90% incidence; commonly malignant and symptomatic (teratocarcinoma). It is mostly seen in young people with a gender ratio of 13.5:1, and an average age of 26 years³.

CASE REPORT

We present the case of a 19-year-old male, apparently healthy, who began with mild fever in the morning, cough and weight loss, exacerbated in the last month before admission. The chest radiograph (**Figure 1**) showed substantial mediastinal enlargement, extended to the apex of the right lung. Computed tomography showed metastatic lesions in both lungs. Since it was impossible to perform a biopsy, we decided to begin neoadjuvant therapy. Persistent fever appeared and antibiotic therapy was started. The patient continued with an unfavorable outcome: anorexia, noticeable asthenia, progressive deterioration and presence of edema, more evident in the right upper limb.

One week later he had an episode of atrial fibrillation with rapid ventricular response. A transthoracic



Figure 1. Preoperative posteroanterior chest radiograph.

echocardiogram was performed which detected a bulk in the right atrium, showing a tricuspid valve tumor image, suggestive of vegetation. A priori analysis with a cardiovascular surgery team was conducted for high surgical and technical risks when accessing the heart and surgical intervention was jointly accepted; with a preoperative diagnosis of malignant mediastinal tumor with pulmonary metastasis and possible tricuspid valve infective endocarditis (versus infiltration or metastasis), in an immunosuppressed patient with persistent fever.

For better care, joint action was required with thoracic surgery for the pulmonary region. A large-scale surgery was then planned to, at first, perform a total resection of the giant mediastinal tumor and pulmonary metastases (**Figure 2**) and, subsequently during the same surgical procedure (with extracorporeal circulation) removal of the right atrial tumor (**Figura 3**).

Surgery was started with a multidisciplinary team of cardiac and thoracic surgeons, they approached the mediastinum through median sternotomy and a large anterior mediastinal mass was found. Its dissection was begun for exeresis and it was possible to separate all neighboring structures, the bulk had greatly extended towards the right pulmonary vertex without infiltrating it, coming from the anterior mediastinum. Tumor removal was challenging as it was

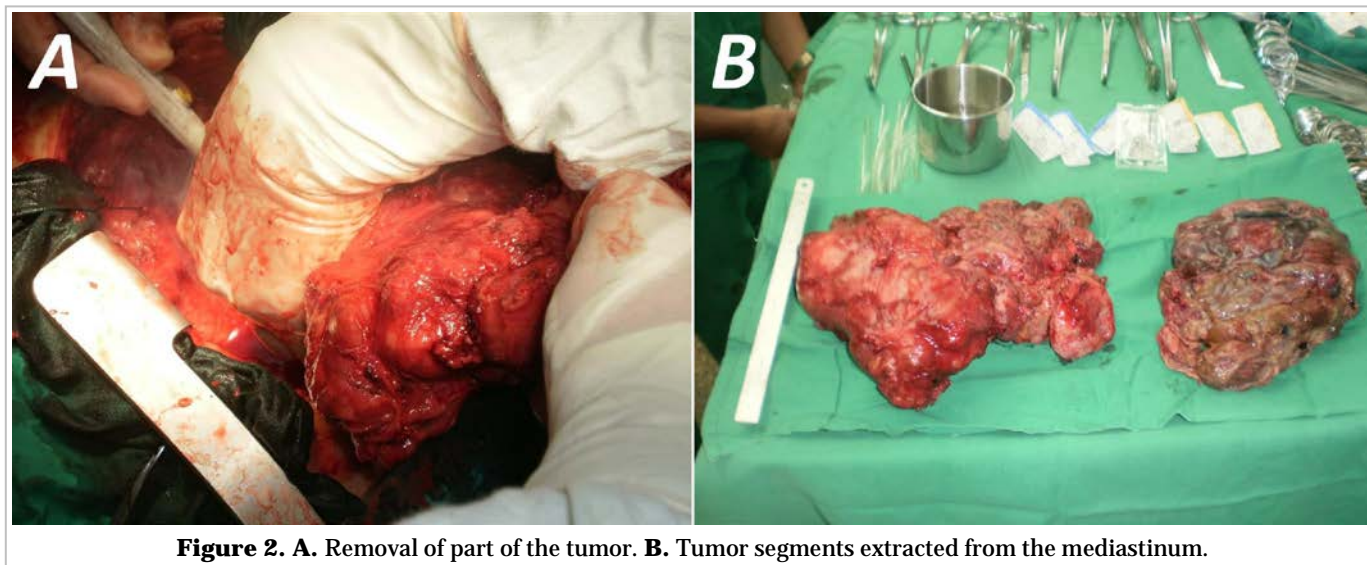


Figure 2. A. Removal of part of the tumor. B. Tumor segments extracted from the mediastinum.

attached to the superior cava. Due to injury it was necessary to suture the vessel. Both lungs were thoroughly examined where multiple metastatic nodules were found and resected; the largest ones in the left base and right vertex. Subsequently, arterial cannulation (ascending aorta, cannula 24) and venous cannulation (bicava, through the right atrium) was performed to guarantee total cardiopulmonary bypass. The right atrium was opened where the mass affecting the anterior tricuspid veil was found, the tumor was extracted as a “bouquet” and, finally, a 2 cm diameter cyst; for total removal, it was essential to resect its base in the affected veil, which motivated valve repair, with the consequent need for normothermic cardiac arrest. Subsequently, anoxic cardiac arrest and cardiopulmonary bypass, decannulation and layered closure were carried out, following careful hemostasis.

The proposed goal was achieved from the surgical perspective, in addition to obtaining the histological diagnosis, which reported: highly malignant mediastinal teratocarcinoma with intracardiac infiltration and tricuspid valvular intake; in addition, pulmonary metastases in the resected nodules. Chemotherapy was started one year later

under treatment regimen by clinical trials; the patient remains asymptomatic with full weight recovery and incorporation to social life.

COMMENTS

Mediastinal germ cell tumors are rare¹⁻³, but when they occur, they are a major surgical challenge and

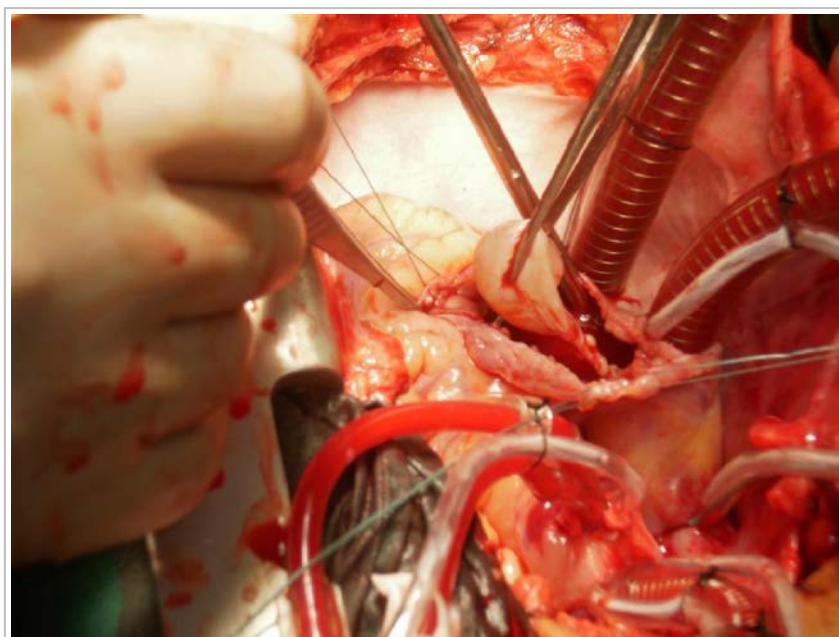


Figure 3. Excision of intracardiac tumor.

often require interdisciplinary management. Many of them, such as teratomas or teratocarcinomas, require surgery and subsequent chemotherapy^{5,8}.

Our case is challenging due to his age and medical condition which forced us into taking faster decisions as he had no histological diagnosis due to the location of the tumor and the danger of injuring important structures when taking a sample for biopsy. Surgical treatment-decision was collectively discussed as well as the diagnosis of infectious endocarditis, suggested in view of persistent febrile symptoms and immunosuppression. But medicine requires general agreement and the logical thing to do was to think about a cardiac infiltration of the mediastinal tumor. Factors such as presence of a very advanced stage disease with pulmonary metastasis and cardiac infiltration affect prognosis in our patient. Surgery was challenging due to its magnitude and high risk. It involved collectively handling many organs (one of them vital) with few reports in literature suggesting this type of combined pulmonary and cardiac operation.

The diagnosis of this type of tumors can be made by various radiological examinations in which a widening of the mediastinum or a large tumor can be seen depending on the time of the study.

Biopsies should be obtained to define the histological strain and its treatment, since surgery is likely to be successful for most men. Teratomas are usually surgical at an early stage, while the rest of them, although may present with an invasive pattern, have a good prognosis with the use of chemotherapy^{5,7}.

In some cases, subsequent surgical resection is recommended for residual mass, where a necrotic tumor proves to be a good prognosis. Surgical removal is the treatment of choice for benign tumors, whether technically resectable, normally have a favorable outcome. In malignant patients, apparent complete resection does not guarantee cure. Therefore, adjuvant chemotherapy should be considered, although there might be recurrences^{8,9}.

The treatment of this type of patients is multidis-

ciplinary and requires, among others, the cooperation of specialists in pulmonology, thoracic surgery and oncology³.

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