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CASE REPORT

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Giant Mediastinal Teratoma. Case Report and Literature Review

Teratoma gigante mediastinal. Reporte de caso y revisión bibliográfica

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ABSTRACT

Mature mediastinal teratoma is the most common thoracic germ cell tumor. No specific cause has been detected, although it has been linked to some risk factors, such as maternal exposures to harmful inhalational environmental agents, maternal nutritional deficiencies, cryptorchidism, hypospadias, hemolytic anemia, among others. It is usually a rare entity, without having its prevalence clearly described, it can reach 3 % of thoracic pathology, being the 10 % of the histological lineage detected in the mediastinal masses. Although some authors have not described differences in the prevalence between males and females, others have detected a clear majority in males, reaching a ratio of 13 to 1. Mature teratoma usually lacks malignant cells and usually has an excellent prognosis after complete surgical excision of the tumor. However, some teratomas may have atypical cells among the diversity of tissues that compose it, which makes it imperative a directed and exhaustive search by the pathologist, to detect this tissue in the tumor mass, since this would imply a worse prognosis and the need to use adjuvant chemotherapy and a follow-up of tumor extension, thus improving the survival of these patients. Next, a case of mature teratoma of the chest is described and a literature review is performed.

Key word: Giant cell tumor, Teratoma, Mediastinum

RESUMEN

El teratoma maduro mediastínico es el tumor de células germinales torácico más frecuente. No se ha detectado una causa específica, aunque se ha relacionado con algunos factores de riesgo, como exposiciones de la madre a agentes nocivos ambientales inhalatorios, deficiencias nutricionales maternas, criptorquidia, hipospadias, anemia hemolítica, entre otros. Suele ser una entidad poco frecuente; sin tener claramente descrita su prevalencia, puede alcanzar el 3 % de la patología torácica y el 10 % de la estirpe histológica es detectada en las masas mediastinales. Si bien algunos autores no han descrito diferencias en la prevalencia entre varones y mujeres, otros han detectado una clara mayoría en la afectación a varones, que alcanza una relación incluso de 13 a 1. El teratoma maduro suele carecer de células malignas y suele tener un excelente

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pronóstico tras la exéresis quirúrgica completa del tumor. Sin embargo, algunos teratomas pueden tener células atípicas entre la diversidad de tejidos que lo componen, lo que hace imperiosa una búsqueda dirigida y exhaustiva por parte del patólogo, para detectar dicho tejido en la masa tumoral, pues ello implicaría un peor pronóstico y la necesidad de utilizar quimioterapia coadyuvante y un seguimiento de extensión tumoral, lo que mejora la supervivencia de estos pacientes.
A continuación, se describe un caso de teratoma maduro del tórax y se realiza una revisión bibliográfica.

Palabras claves: Tumor células gigantes, Teratoma, Mediastino

INTRODUCTION

We present the case of a mature teratoma in the left hemithorax of a 29-year-old male, with a classic clinical presentation of pleuritic chest pain in the left hemithorax, moderate exertional dyspnea, right trepopnea (dyspnea that occurs when adopting lateral decubitus position), and occasional dry cough. The patient was treated with complete tumor excision, with no malignancy detected in the mass, achieving full recovery six months after the surgical intervention.

Case report: medical record, diagnosis, treatment

29-year-old male patient presents with sudden-onset pain in the left shoulder, radiating to the left side of the back. The pain is intense, and worsens when adopting the right lateral decubitus position, when coughing, and with deep inspirations, and is relieved only upon taking non-steroidal anti-inflammatory drugs and antispasmodics. The patient also reports a sensation of dyspnea that has progressively worsened, now occurring even at rest, and is further aggravated by moderate exertion, supine position, and right lateral decubitus position. Symptoms partially improve when adopting the left lateral decubitus position.

Physical examination reveals a blood pressure of 106/82 mmHg, temperature of 37°C, SpO_2: 94%, heart rate of 94 bpm, and fasting blood glucose of 83 mg/dL.

Pulmonary auscultation shows good air entry in the right lung, and decreased air entry in the left lung, throughout the entire lung field, without other significant findings.

Laboratory studies, including hematology, blood chemistry, coagulation times, hormone profiles, and tumor markers, were all normal.

Spirometry results were as follows: FEV_1 (forced expiratory volume in the first second) 1.47 L (33%), FVC (forced vital capacity) 1.59 (30%), FEV_1/FVC 92% (110%), and FEF (forced expiratory force) 25-75% 2.38 L/s (45%), indicating a severe restrictive pattern. Due to the unavailability of a plethysmograph and DLCO (diffusing capacity of the lungs for carbon monoxide) in our facility, a stair-climbing test was conducted, during which the patient was able to climb 8 steps, equivalent to a DLCO of 60-80% predicted and an approximate VO_2 max of 20-25 ml/kg/min.

The chest X-ray revealed a rounded opacity with solid appearance without air bronchogram, occupying two-thirds of the left hemithorax, with a slight deviation of the mediastinum to the right (Figure 1).

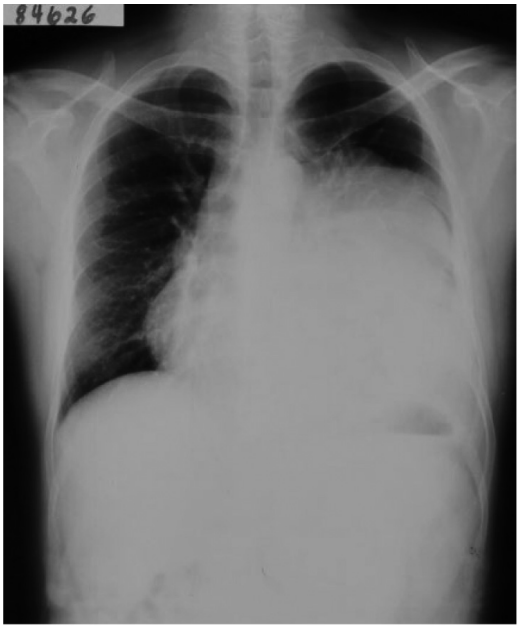

Figure 1. Chest X-ray, posterior-anterior view.

A thoracic CT scan revealed a heterogeneous, solid mass with various internal densities, including fat, calcium, soft tissue, and cartilage, suggestive of a teratoma (Figure 2).

A bronchoscopy was performed, showing only extrinsic compression of the main bronchus and segmental bronchi on the left side, without any endobronchial exophytic lesions.

Given the clinical impression of a thoracic teratoma, a surgical resection was decided. The mass was completely removed via a left posterolateral thoracotomy, measuring 17 x 10 x 6 centimeters, with macroscopic evidence of epithelial, cartilaginous, and adipose tissues, along with the presence of hair follicles and sebaceous material.

Histologically, the mass contained multiple mature tissue types, including mesenchymal tissue such as striated muscle and cartilage, as well as bone marrow, squamous epithelial tissue with skin appendages, adipose tissue, and mucin-producing tissue. (Figures 3 and 4).

The diagnosis was germ cell tumor of the mature teratoma type, without malignant components.

Following mass resection, the patient has progressed satisfactorily, achieving full expansion of the left lung, and is currently undergoing rehabilitation therapy with respiratory physiotherapy.

Case discussion

Germ cell tumors account for approximately 10-15% of mediastinal tumors. These neoplasms consist of germ cells that have migrated abnormally during early embryonic development. There are several theories related to this; some of them propose that an error occurs in the migration of primitive germ cells along the urogenital ridge[1], while others suggest that these tumors originate from totipotent cells left behind from the blastula or morula stage during embryogenesis. The mediastinum is the most frequent extragonadal location, comprising about 1-3% of all giant cell tumors. These can be subdivided into three main groups: teratomas, seminomatous tumors, and non-seminomatous tumors. The predominant histological type is the mature teratoma, followed by seminomas, non-seminomatous giant cell tumors, and finally, mixed giant cell tumors.

The teratoma is the most common mediastinal germ cell tumor, with approximately 75% being mature teratomas.[2] No risk factors for mediastinal teratomas were identified in the review; however, risk factors have been described for testicular teratomas, including low birth weight, cryptorchidism, advanced maternal age, neonatal jaundice, and retained placenta,[3] and for ovarian teratomas, including advanced age and post-menopause.[4] The occurrence of these tumors is more frequent in young adults, but they have been reported across all age groups. Men and women are affected with similar frequency. However, a descriptive study conducted between 1986 and 2012 in Costa Rica

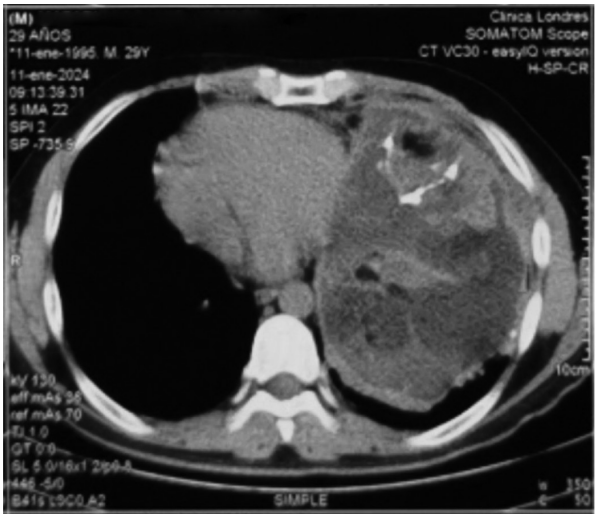

Figure 2. Thoracic CT scan.

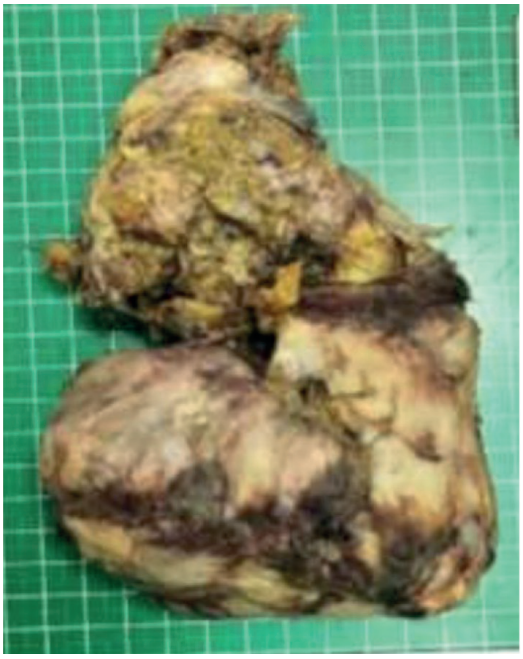

Figure 3. Macroscopic image of the resected mature teratoma, measuring 17 x 10 x 6 cm.

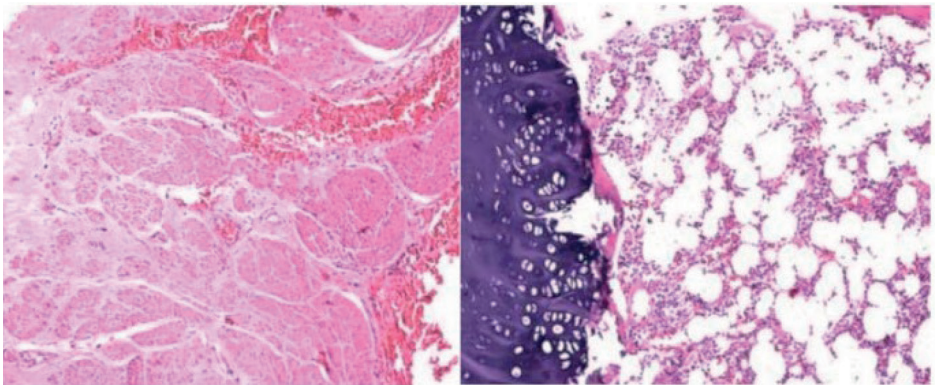

Figure 4. Histological image (40X) showing striated muscle tissue, cartilage, and bone marrow.

found a higher frequency in men, with a gender ratio of 13.5:1 and an average age of 26 years.[5]

Most patients will present with symptoms at the time of diagnosis, with only one-third of the cases being asymptomatic. The most common symptoms are retrosternal chest pain, dyspnea, and cough; other possible symptoms include dysphonia, diaphragmatic paralysis, hemoptysis, and inferior vena cava syndrome.[5] If the tumor fistulizes into the bronchial system, the patient may expectorate various types of tissues that make up the tumor.

By definition, a teratoma is composed of tissues that are different from the area in which it has grown. A teratoma can contain mature tissues from any of the three primary germ layers: the mesoderm (bone, cartilage, and muscle), the endoderm (respiratory, gastrointestinal tissues, and mucous glands), and the ectoderm (nerve fibers, epidermal appendages).[6] Ectodermal derivatives are the most common, and if only epidermal derivatives are present, it is called a dermoid cyst.

Teratomas are classified as mature, which tend to be well-differentiated and benign; immature, which contain fetal tissue and are malignant; and those with a malignant component. Giant teratomas are those that occupy half or more of the hemithorax. The most common symptoms are chest or shoulder pain, dyspnea, cough, fever, pleural effusion, and bulging of the thoracic wall.[7]

On X-rays, a teratoma usually appears as a solid, rounded, lobulated, and asymmetrical mass. It may present with cystic, calcified images and heterogeneity of densities that can usually be differentiated more precisely with chest CT scans, often providing a fairly precise diagnosis even before surgical resection.

The treatment consists of surgical resection, which yields excellent results. All teratomas must be removed due to their compressive effect on neighboring vital structures, as well as their potential for progressive growth and malignancy. Given the FEV_1 of 33% in this particular case, the surgical risk had to be carefully assessed. Given the unavailability of DLCO and plethysmograph in our center, the stair-climbing test is a good option for calculating the DLCO and approximate maximum oxygen consumption. Considering that the patient was able to climb 8 steps, equivalent to a DLCO of 60% to 80%, a VO_2 max of 20 to 25 ml/kg/min, and in the absence of coexisting parenchymal lung disease, it was determined that resecting the extrapulmonary compressive mass could improve FEV_1 and functional respiratory capacity. Therefore, it was decided to proceed with surgery, despite the moderate surgical risk that the patient still had. After the resection of the mass and one year of cardiopulmonary rehabilitation, the patient has recovered a FEV_1 that has now reached 53%, with the ability to climb 10 steps.

It is very important to perform spirometry, as well as plethysmography, DLCO, and VO_2 max studies in patients undergoing thoracic surgeries such as lobectomies, pneumonectomies, or resection of masses like these. A comprehensive assessment of all variables in each individual case is essential for decision-making. Given the lack of access to DLCO in many hospitals, an alternative is the stair-climbing test, which has a high scientific correlation with VO_2 max and a moderate scientific correlation with DLCO.[8,9,10]

Teratomas should be thoroughly examined by the pathologist throughout the entire mass to rule

out the presence of a malignant neoplastic component, as this would worsen the prognosis due to its potential to infiltrate adjacent organs or even metastasize. If malignant cells are detected, treatment should include adjuvant chemotherapy in addition to surgical resection, which will improve patient survival. Additionally, extension studies such as PET-CT should be conducted to rule out metastatic lesions at other levels.[11]

CONCLUSIONS

Mature thoracic teratoma is a rare condition. Risk factors predisposing patients to its development are not well known. A thorough histological examination of the entire mass should be conducted to rule out the presence of a malignancy within the heterogeneous tissues that make up the teratoma. All teratomas must be surgically removed, and if a malignancy is detected, adjuvant chemotherapy and tumor extension study should be considered.

Conflict of interest

The authors have no conflict of interest to declare.

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