

Massive Cystic Teratoma Of The Anterior Mediastinum In A Young Child

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Abstract:

Background: Mediastinal teratomas are rare congenital germ cell tumors derived from pluripotent embryonic cells and composed of elements from all three germ layers. Although most are mature and benign, large anterior mediastinal teratomas may lead to severe cardiorespiratory compromise due to compression of adjacent vital structures.

Case Presentation: We report the case of a 3-year-old girl presenting with a one-week history of intermittent dry cough, fatigue, and dyspnea. Imaging studies revealed a large, well-encapsulated multicomponent mass (12.0 × 8.7 × 10.1 cm) occupying the right hemithorax and exerting significant mass effect on the mediastinum, with near-total compression of the right lung. Serum alpha-fetoprotein levels were within normal limits. Given the extent of mediastinal compression, urgent surgical resection was undertaken via wide posterolateral thoracotomy. Complete excision of the tumor, together with a portion of the thymus, was achieved while preserving the phrenic nerve and surrounding vital structures.

Results: Histopathological examination confirmed a mature benign cystic teratoma containing well-differentiated derivatives of ectodermal, mesodermal, and endodermal origin. The postoperative course was uneventful.

Conclusion: Giant anterior mediastinal teratomas in pediatric patients may present with significant mass effect and potential life-threatening complications. Prompt diagnosis and carefully planned complete surgical excision allow safe management and are associated with an excellent prognosis.

Keywords: mediastinal teratoma; pediatric mediastinal mass; cystic teratoma; anterior mediastinum; germ cell tumor; surgical resection; mediastinal mass syndrome.

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I. Introduction

Teratomas are rare congenital tumors that arise from pluripotent embryonic cells and contain derivatives from all three germ layers, including ectoderm, mesoderm, and endoderm. According to some reports, teratomas occur in approximately 1 in 4,000 live births worldwide [1]. The anterior mediastinum is the most common location for extragonadal germ cell tumors (over 60% of all germ cell tumors), with only 5% located in the posterior mediastinum [2, 3]. Germ cell tumors are divided into three groups: epidermoid cysts, dermoid cysts, and teratomas. These tumors may be cystic or solid, with some presenting a combination of both. They contain multiple germ cell layers and are composed of tissue foreign to the organ or anatomical site in which they arise [4].

Most mediastinal teratomas are mature and are characterized by slow growth. The clinical presentation ranges from an asymptomatic course to symptoms such as dysphagia, cough, stridor, chest pain, and severe respiratory distress, with symptom severity primarily determined by compression of adjacent thoracic structures [5,6]. Giant tumors of the anterior mediastinum may result in significant hemodynamic and respiratory compromise secondary to mass effect, a condition commonly referred to as mediastinal mass syndrome. Owing to the confined mediastinal space, large anterior mediastinal tumors are particularly prone to mechanical compression or invasion of surrounding structures. Circulatory collapse and respiratory failure may occur at any time in patients with sizeable mediastinal masses, including during diagnostic procedures such as biopsy or during surgical intervention [7]. Therefore, mediastinal mass syndrome in the pediatric population frequently poses a significant challenge with respect to perioperative management strategies and surgical approach [8]. In this context, we present a case of successful surgical management of a large cystic teratoma located in the anterior mediastinum of a young child.

II. Case Presentation.

In this context, we report a case of successful surgical treatment of a large cystic teratoma located in the anterior mediastinum in a young child.

A 3-year-old female patient presented with a periodic dry cough, fatigue, and dyspnea that had begun one week earlier, prompting medical consultation. The mother reported that the pregnancy had been uneventful and that the child was born at term. Thoracic ultrasound suggested a space-occupying lesion localized in the right hemithorax, and the patient was referred to our institution.

Physical examination revealed slight bulging of the right hemithorax with localized retraction in the lower intercostal spaces. Laboratory results were within normal limits except for decreased hemoglobin (105 g/L) and elevated ALT (53.50 U/L), AST (99.60 U/L), and ESR (35.0 mm/h). Serum alpha-fetoprotein levels were within normal limits.

Echocardiography revealed dilation of the right heart chambers, mild pulmonary hypertension (PSAP 34 mmHg), grade I mitral regurgitation, and grade I tricuspid regurgitation, with preserved left ventricular systolic function. Contrast-enhanced chest computed tomography demonstrated a massive space-occupying lesion in the right hemithorax measuring 12.0 × 8.7 × 10.1 cm. The lesion was well-defined and encapsulated, with a capsule thickness of 0.5 cm, peripheral fine calcifications, and focal adipose inclusions, as well as pathological post-contrast enhancement of the capsule. The mass exerted a pronounced mass effect on the mediastinum and the right main bronchus, with subtotal compression of the right lung. An incomplete vascular ring was also observed—an aberrant right subclavian artery with a retrosophageal course. No additional pathologies were identified (Fig. 1).

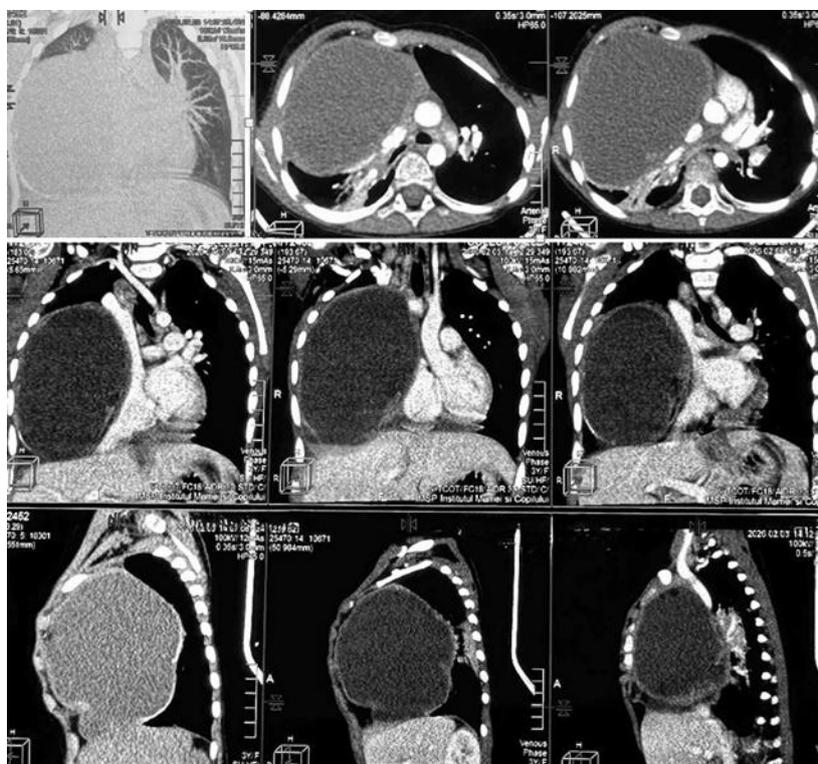


Fig. 1. Patient E.C., 3 years old. CT imaging findings suggestive of a massive, multicomponent space-occupying lesion in the right hemithorax – possible teratoma? Incomplete vascular ring – aberrant right subclavian artery.

Given the significant compression of mediastinal structures and the right lung, urgent surgical intervention was indicated. The choice of surgical approach for mediastinal tumor resection depends on tumor size, anatomical location, and the relationship with adjacent vital structures, with options including median sternotomy, anterolateral thoracotomy, posterolateral thoracotomy, clamshell incision, or thoracoscopic surgery [9].

In the present case, a wide posterolateral thoracotomy via the fifth intercostal space was performed. The patient was positioned in the left lateral decubitus position, providing optimal exposure for safe resection.

Intraoperatively, the tumor was localized to the right middle and superior mediastinum, occupying approximately 80% of the pleural cavity. Anteriorly, the mass was adherent to the thymus, and posteriorly, it compressed the right lung. The tumor demonstrated close mass adherence to the pulmonary hilum and the right pericardial surface, exerting pressure on the right atrium, the azygos vein arch, and the superior vena cava. The right phrenic nerve was also adherent to the tumor surface. Moderate pericardial effusion was observed.

Surgical management commenced with cyst fluid aspiration, followed by careful blunt dissection with preservation of the phrenic nerve. Blunt dissection combined with electrocautery enabled safe separation of the tumor from surrounding structures, achieving complete excision of the mass along with a portion of the thymus (Fig. 2).

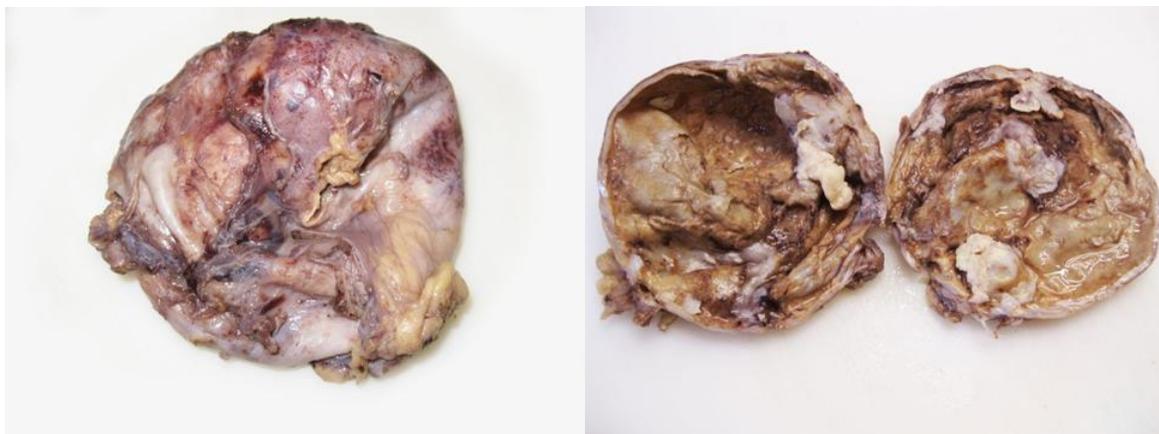


Fig. 2. Macroscopic appearance of the resection specimen after aspiration of the liquid content (A) and on section (B).

A small pericardial fenestration was performed, allowing drainage of clear pericardial fluid. The pleural cavity was drained with a 12 French chest tube inserted in the sixth intercostal space. The procedure was completed with closure of the pleural cavity and restoration of the normal anatomical layers of the chest wall.

The postoperative course was uneventful. The patient was extubated on the first postoperative day. A chest radiograph obtained on postoperative day two demonstrated full re-expansion of the right lung with no evidence of pleural effusion (fig. 3). The chest tube was removed after 48 hours, and the patient was discharged on postoperative day nine.



Fig. 3. Patient E.C., 3 years old. Chest radiograph performed on postoperative day 2: The lungs are uniformly aerated. The pulmonary markings are accentuated, with reduced hilar structure. The contours of the mediastinum and diaphragm are clearly visualized. No infiltrative or nodular opacities are detected. The pleural sinuses are free. A tubular drain is visible in the pleural cavity.

Histological examination of the resection specimen revealed features characteristic of a mature benign cystic teratoma composed of germ cell derivatives from all three germ layers. The ectodermal components included skin with a predominance of glandular structures, particularly sebaceous glands, epidermoid cysts of various sizes, and isolated hair shafts. Endodermal derivatives were represented by cartilage, muscle tissue, adipose tissue, connective and fibrotic patches, and bands of lymphoid tissue. The mesodermal component consisted of bronchial tissue, gastric and intestinal mucosal glandular tissue, and renal-type tissue. For the most part, the main cyst was lined by non-dysplastic stratified squamous epithelium. Papilliform structures composed of differentiated thymic tissue were observed, as well as papillary folds formed by lymphoid follicles and mixed gastro-intestinal or bronchial-type epithelium (fig. 4).

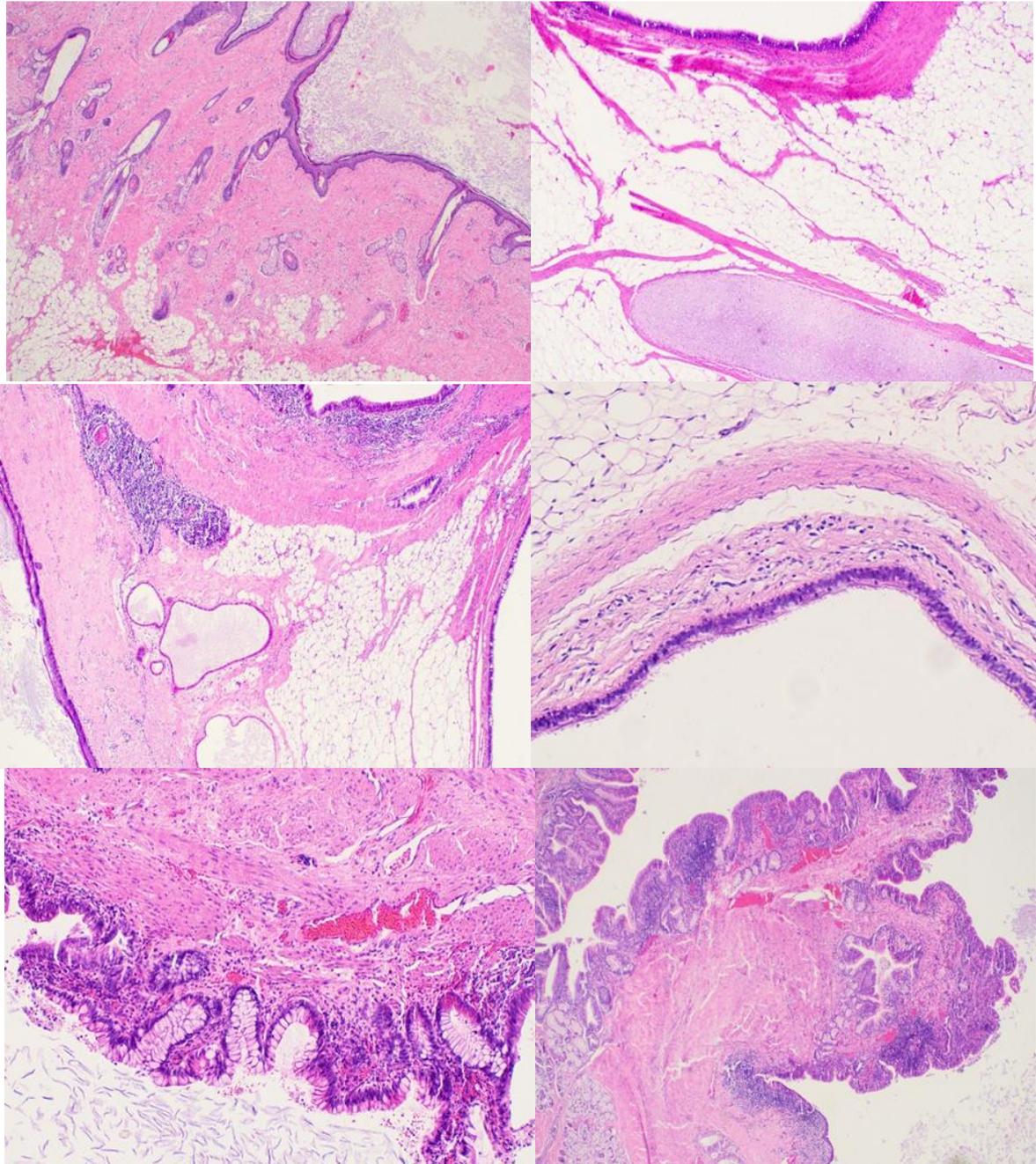


Fig. 4. Histological aspects of the three germ layer derivatives: ectodermal (A), endodermal (B), and mesodermal (C). H&E staining, x40. Histological features of mesodermal germ layer derivatives: ciliated epithelium (D), intestinal mucosa (E) and fibro-polypoid structures of bronchial origin (F). Color. H&E staining, x150.

III. Discussion

Teratomas are the most common mediastinal germ cell tumors, accounting for 60–70% of cases, and represent the most frequent extragonadal germ cell tumors in both prepubertal and postpubertal patients, regardless of sex. They comprise approximately 15% of all mediastinal masses in adults and about 25% in children [10]. Malignant transformation of mature teratomas is rare, occurring in 1–3% of cases, and has been reported as sarcomas, adenocarcinomas, squamous cell carcinomas, and carcinoid tumors [11–13].

The term “teratoma” is derived from the Greek words *terato*, meaning “monster,” and *onkoma*, meaning “swelling” [8]. Two main theories regarding the development of germ cell tumors have been proposed. The embryonic theory suggests that totipotent cells capable of differentiating into multiple tissue types escape normal regulatory control and develop locally in the mediastinum without migrating to the gonads. The germ cell theory posits that each germ cell tumor originates from an embryonic germ cell [14].

The clinical presentation of mediastinal teratomas is highly variable. Over 60% of patients are asymptomatic, with tumors often discovered incidentally. Symptomatic patients may present with chest pain, cough, dyspnea, or, occasionally, hemoptysis, with symptom severity primarily determined by compression of adjacent structures [15]. Respiratory distress is more common in children under 2 years of age, with more than three-quarters exhibiting signs of tracheal compression [16].

The clinical course of mediastinal teratomas may be complicated by life-threatening events. Rarely, these tumors rupture into adjacent structures such as the pleural space, pericardium, lung parenchyma, or tracheobronchial tree [17]. Pleural effusions secondary to tumor rupture are relatively uncommon; preoperative rupture with concomitant pleural effusion occurs in fewer than 25% of patients. Proposed mechanisms for rupture include autolysis, inflammation, ischemia, and infection, with inflammation and necrosis associated with sebaceous or digestive enzymes produced by tumor tissue potentially contributing [18]. Clinical manifestations of rupture include sudden, severe chest pain, dyspnea, and hemoptysis. Chest radiography typically demonstrates features of a mediastinal mass accompanied by unilateral pleural effusion [19]. Large mediastinal teratomas may also be complicated by superior vena cava syndrome, dysphagia, or pericardial effusion, which in severe cases can progress to cardiac tamponade [20].

Anterior mediastinal teratomas diagnosed prenatally and treated in the neonatal period are extremely rare. These tumors are usually detected incidentally during postnatal examination, with perinatal ultrasound and computed tomography playing crucial roles in early diagnosis [21].

Chest radiography remains the initial imaging modality for mediastinal masses. The radiologic appearance of a mature teratoma varies with its composition: cystic teratomas typically appear as well-circumscribed, rounded opacities, whereas solid teratomas demonstrate lobulated and irregular contours. Intralesional calcifications are nonspecific [22]. Non-contrast and contrast-enhanced computed tomography is essential for diagnosis, allowing evaluation of mass effect and assessment of invasion into adjacent structures. CT commonly demonstrates a capsulated lesion with septations of variable density, with mature mediastinal teratomas appearing as round or lobulated masses containing soft tissue, fluid, fat, and calcifications. The presence of spontaneous air–fluid levels is an important diagnostic clue. Magnetic resonance imaging reveals heterogeneous signal intensities, with fluid detected in 88% of tumors and fat in 63% [23–25].

The differential diagnosis of cystic teratomas includes cystic thymoma, lymphoma, and complex pericardial cysts. Anterior mediastinal masses with contrast-enhancing solid components may also represent lymphoma, bronchogenic cysts, cystic thymoma, or other germ cell tumors. Cross-sectional imaging is typically required for accurate differentiation [6,26,27]. Rare cases of mature mediastinal teratomas associated with multilocular thymic cysts have also been reported [28]. Serum tumor markers such as alpha-fetoprotein and beta-human chorionic gonadotropin are essential in differentiating mature teratomas from malignant germ cell tumors; normal levels support a diagnosis of benign mature teratoma [29].

Complete surgical resection is the treatment of choice for mediastinal tumors, both to distinguish benign from malignant lesions and to relieve or prevent associated symptoms. Median sternotomy is often considered optimal for exposure, whereas anterolateral thoracotomy may be preferred for tumors confined to one hemithorax [30–32]. Since 1990, minimally invasive techniques have demonstrated efficacy and advantages over open procedures [25,33,34]. Video-assisted thoracoscopic surgery (VATS) is effective even for large mediastinal tumors, providing a safe and efficient alternative to conventional thoracotomy or sternotomy with improved postoperative outcomes [35,36]. A practical approach is to begin with VATS and assess intraoperative findings before conversion to open surgery, allowing selection of the most appropriate technique based on the patient's condition [37]. In pediatric patients with tumors adherent to major vascular structures, some authors recommend a combined approach using mini-thoracotomy with video assistance [38]. In cases of extensive adhesion to critical vascular or neural structures, complete separation may be challenging; partial resection may be considered to protect surrounding tissues, with residual tissue left for natural resorption [39].

Nonetheless, complete surgical excision remains the definitive treatment for mediastinal cystic teratomas, offering excellent prognosis and minimal risk of recurrence. Histopathological confirmation is essential for accurate diagnosis [40].

The asymptomatic developmental phase and nonspecific clinical manifestations contribute to delayed diagnosis in children. Radical surgical resection of large anterior mediastinal teratomas is challenging due to tumor extension and strong adhesion to vital structures. Careful surgical planning, adequate exposure, and meticulous dissection - accounting for the anatomical particularities of major vascular structures and nerves - enable safe radical excision while minimizing the risk of severe complications.

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