CASE REPORT

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Rupture of anterior mediastinal mature teratoma in a 16-years-old girl: A case report

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ABSTRACT

Background: Mediastinal teratomas are rare neoplasms, accounting for <10% of all mediastinal tumors. The most commonly involved site is the anterior mediastinum. In prepubertal patients, teratomas account for 58% of all germ cell tumors in the mediastinum and are usually asymptomatic in 50% of children. Tumor rupture is rare and can cause pleural effusion.

Case description: A 16-year-old girl came to the hospital with the main complaint of shortness of breath three days ago. Physical examination found an increase in the respiratory rate and pulse rate, asymmetric chest wall movement, no right pulmonary breath sounds, and an increased right vocal fremitus. Blood laboratory tests show an elevated sedimentation rate. CT imaging showed a well-defined, flat edge tumor mass with heterogeneous density in solid, cystic, fat, and calcified. The tumor measured 14.8 x 10.0 x 17.4 cm, in the anterior to the medial mediastinum, urge the heart and other mediastinal organs to the left, the collapse of the right lung's inferior lobe, and right fluidopneumothorax. The tumor was suspicious of a teratoma. Right posterolateral thoracotomy and tumor resection was performed. Macroscopically, there was a multicystic tumor mass measuring 15.5 x 14.0 x 9.0 cm with an incomplete capsule. Microscopic examination of the tumor mass showed 3 components of the germ layer, namely ectodermal, mesodermal and endodermal. No immature component appears.

Conclusion: Based on clinical features, CT imaging, macroscopic and routine histopathological examinations this case was concluded as rupture of mature teratoma.

Keywords: mature teratoma, rupture, anterior mediastinum

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INTRODUCTION

Mediastinal teratomas are rare neoplasms, accounting for <10% of all mediastinal masses. This tumor can occur in men and women, before and after puberty, without sex dominance. In prepubertal patients, teratomas account for 58% of all germ cell tumors in the mediastinum and can occur even in fetuses as young as 18 weeks of gestation. In postpubertal patients, teratomas account for 93% of germ cell tumors in women and 35% in men.[1] The most common location involved by mediastinal teratoma is the anterior mediastinum.[1,2]

The mature teratomas of the mediastinum are usually asymptomatic in 50% of children and 66% of adults.[1,3,4] They are typically found incidentally on chest X-ray.[5-6] These tumors may be relatively large due to slow growth with few clinical symptoms. Symptomatic cases can be associated with chest, back, or shoulder pain, dyspnea, cough, and fever due to chronic pneumonia.[8] Respiratory distress is more common in neonates and children than in adults, usually because of the size of the lesions that take up space. Rare symptoms include superior vena cava syndrome, Horner's syndrome, and pneumothorax, which are common in adults. Tumor rupture is rare but appears to be more common in mediastinal teratomas than teratomas elsewhere, causing pleural effusion, empyema, or cardiac tamponade.[1,3] Tumor markers (α-fetoprotein and β subunit of human chorionic gonadotropin) are not elevated. Mediastinal teratomas can extend to one or both thoracic cavities and cause atelectasis.[1] Surgical resection is curative therapy and a definitive diagnosis.[3,9]

The next following reports a case of 16-years-old girl with mediastinal tumor suspicious for teratoma diagnosed 1 year before coming to the hospital. She came with worsening symptoms of shortness of breath and massive pleural effusion. This case was reported because of its rare incident.

CASE REPORT

A 16-years-old girl came to the Emergency Unit, Department of Pediatrics, complaining of shortness of breath since 3 days ago before admitted to the hospital. Shortness of breath begins with a cough with greenish phlegm and fever for three weeks previously. She also complained of decreased appetite accompanied by weight loss and weakness, making her just in bed more. The patient had

experienced the same complaint about five years ago. She had a history of control to the Thoracic and Cardiovascular Surgery Polyclinic with a diagnosis of tumors in the mediastinum about one year ago. She planned for surgery to remove the tumor, but the patient and family refused.

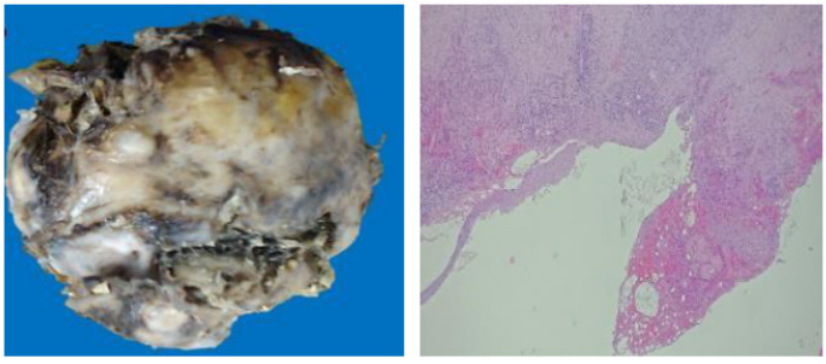
On physical examination, it was found that the nutritional status was deficient, the movement of the chest wall was asymmetric, there was no sound of right lung breath, and the right vocal premitus increased. LED results were 105.2 mm/ hour, , Hb 10,4 gr/dl, LDH 230 U/ L, β-HCG blood 0.10 mIU / ml, αFP 0.66 IU / ml. The chest X-ray could not evaluate the heart because the heart's right border was veiled, suggesting that it was pushed to the left side, and there was a homogeneous veil over the entire right hemithorax covering the pleural sinus and right diaphragm. This image suggests a massive right pleural effusion. Chest CT scan on axial slices without and with contrast showed right fluidopneumothorax and a mass with firm borders and flat edges. The tumors' densities was in the form of solid, cystic, fat and calcified. The predict size of the tumor was 14.8 x 10 x 17.4 cm in the anterior mediastinum to medius, urge the heart and other mediastinal organs to the left. It was suspected teratoma with obliteration of the right main bronchus by a mass with inferior lobe collapse of the right lung.

The patient underwent right posterolateral thoracotomy surgery with tumor resection and chest tube insertion. The operation showed a tumor mass in the anterior mediastinal region near the lung. The tumor filled 4/5 of the right hemithorax cavity, the impression that the capsule of the tumor mass was ruptured.

The tissue was then sent to the Anatomical Pathology Laboratory for histopathologic examination. Macroscopic examination showed pieces of tissue, the largest tissue measuring

15.5 x 14.0 x 9.0 cm, round, and irregularly shaped, the capsule is not intact (Figure 1A). On the cut section, the inner surface appears multilocular, gray-white, yellowish, and reddish, mostly containing sebaceous material, as well as hair and bone structure. Other tissue pieces were capsules of tumor mass in the form of irregular sheets, the largest size was 13.5 x 3.5 x 0.3 cm, and the smallest size was 3.5 x 1.7 x 0.2 cm.

Microscopic examination of the tumor capsules revealed a fibrous connective tissue stroma lined with stratified squamous epithelium with subepithelial vascular proliferation and distribution of fibroblasts and lymphoplasmacytic inflammatory cells (Figure 1B). Tumor mass showed three components of the germ layer, namely ectodermal, mesodermal and endodermal. The ectodermal component consists of the skin and adnexas, salivary glands and glial tissue. The mesodermal component consists of mature cartilage, mature fat tissue and muscle. The endodermal component consists of the gastrointestinal and respiratory epithelium (Figure 2). Based on these findings, the patient was diagnosed histopathologically as a mature teratoma with the capsule's rupture.



A B

Figure 1. A. Macroscopic appearance of the tumor shows round and irregularly shaped, and the capsule is not intact. B. Microscopic appearance of the tumor capsule shows a fibrous connective tissue stroma lined by squamous epithelium (Hematoxylin and Eosin, 40x).

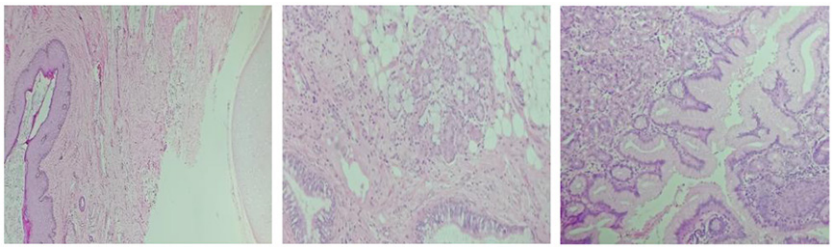


Figure 2. Histopathological evaluation of the tumor shows a mature component of the 3 germ layers.
A. Skin, adnexa, and cartilage (Hematoxylin and Eosin, 40x). B. Salivary gland, respiratory epithelium and fat tissue (Hematoxylin and Eosin, 100x). C. Gastrointestinal epithelium (Hematoxylin and Eosin, 100x).

DISCUSSION

Teratomas are germ cell tumors consisting of somatic tissue originating from two or three germ layers, namely ectodermal, endodermal, and mesodermal. Teratomas can be classified as mature teratomas consisting only of mature tissue, and immature teratomas containing immature tissue.[1] Teratomas usually arise in the gonads but can also be extra gonads. Extragonadal teratomas can occur in the anterior mediastinum, retroperitoneum, pineal, and anterior suprasellar. The most commonly involved site of the mediastinal teratoma is the anterior mediastinum.[2]

Mediastinal teratoma is a rare neoplasm and accounts for <10% of all mediastinal masses. This tumor can occur in men and women before and after puberty, without sex dominance. In prepubertal patients, teratomas account for 58% of all germ cell tumors in the mediastinum, and can occur even in fetuses over 18 of gestation. In postpubertal patients, teratomas account for 93% of germ cell tumors in women and 35% in men. Mature teratomas are more common in women than in men.[1] The etiology of teratomas is unknown, but there is a theory that gonadal and midline teratomas are derived from totipotent primordial germ cells. These cells develop among the yolk sacs' endodermal cells near the allantois's origin and migrate to the gonads during the 4th and 5th weeks of pregnancy. Some cells may lose their target to reach the destination and produce teratomas.[10]

Mature teratomas of the mediastinum are asymptomatic in 50% of children and 66% of adults.[1,3,4] This tumor is usually found incidentally on chest X-ray.[5-7] This tumor may be relatively large due to slow growth with few clinical symptoms. Symptomatic cases can manifest as the chest, back, or shoulder pain, dyspnea, cough, and fever caused by chronic pneumonia. Respiratory distress is more common in neonates and children than adults, usually due to the lesion's size. Rare symptoms include superior vena cava syndrome, Horner's syndrome, and pneumothorax, which are common in adults. Extensive granulomatous inflammation can be seen caused by ruptured cysts. Tumor rupture is rare, but are more common in mediastinal teratomas compared to teratomas elsewhere, that can causing pleural effusions, empyema, or cardiac tamponade.[1,3] Pleural effusions resulting from tumor rupture are quite rare.[11] Rupture preoperative with concurrent pleural effusion account for <25% of patients.[12] Tumor markers (α-fetoprotein and β subunit human chorionic gonadotropin) were not increased. Teratomas can also extend to one or both thoracic cavities and cause atelectasis.[1] Several hypotheses have been proposed to why mediastinal teratomas rupture, including autolysis, inflammation, ischemia, and infection. Inflammation and necrosis associated with the sebaceous or digestive enzyme originating from the tumor tissue can cause teratoma rupture. The ruptured can produce various potentially dangerous clinical manifestations; the inflammation and adhesions caused by the ruptured component significantly affected the surgical approach.[13] In these patients the symptoms complained were shortness of breath, cough, recurrent fever, and decreased appetite. These complaints have been going on for a long time, causing the patient to experience anemia and an increase in sedimentation rate and weight loss.

CT imaging is the modality of choice for preoperative tumor diagnosis.[1] The classic finding of mediastinal teratoma is well-defined tumor, and shows a multilocular cystic structure in nearly 90% of cases. Density is heterogeneous, with various combinations of soft tissue, fluid, fat, and calcium. Calcification occurs in 26% to 53% of cases. Wall calcification of tumors like shells, or bones and teeth occurs in up to 8% of cases. An adult teratoma can be mistaken for a large pleural effusion on routine chest radiographs.[1] With its characteristic imaging features, the radiologist plays a key role in diagnosing and enabling optimal surgical planning. The radiological manifestations of rupture depend on the space in which the rupture occurs. On CT scan, ruptured teratomas show inhomogeneous density in each internal compartment, possibly associated with mixing extravasation components between compartments. Additional findings also characterized ruptured teratomas, which included a fatty lump at the site of rupture, consolidation or atelectasis in the adjacent lung, and pleural effusion.[14] Immature teratomas appear more frequently as solid masses. There are no detailed reports of imaging studies for pure immature teratomas.[1] CT scan of the chest of the axial slice with contrast in this patient shows a well-defined mass, flat margins, and heterogeneous density in the form of solid, cystic, fatty and calcified, suspicious of a teratoma in the mediastinum anterior. Other features were the collapse of the right lung's inferior lobe and right pleural effusion as signs of ruptured teratoma. Pleural fluid cytology shows chronic suppurative inflammation or empyema.

Mature teratomas of the mediastinum are usually encapsulated masses, average 10 cm in diameter, ranging from 3 to 25 cm. It may also adhere to the lungs or adjacent large blood vessels. The cut surfaces variated, showing multilocular or unilocular cysts varying in size from a few millimeters to several centimeters. Cysts may contain clear fluid, mucous material, sebaceous and keratinaceous debris, hair, fat, cartilage, and rarely teeth or bone.[1] In this patient, a tumor mass measuring 15.5 x 14.0 x 9.0 cm was obtained with an incomplete capsule. The cut surface shows a multilocular cyst, mostly containing sebaceous material and a small amount of hair and bone.

A mature teratoma's histopathological features contain two or three germ layers seen throughout the tumor in varying distributions. Ectodermal tissue includes skin, skin derivatives, squamous epithelium, brain tissue, glia, retina, neuroectoderm, choroid plexus, and ganglia. Glial tissue varies in

cellularity and maybe more cellular than the normal brain without indication of immaturity. Mesodermal tissue includes muscle tissue, bone and cartilage, fat, and embryonic stroma. Endodermal tissue consists of the liver, bronchial and gastrointestinal epithelium, thyroid gland, and/or salivary gland. In the histopathological evaluation of this case, three germ layer components were found, namely ectodermal, mesodermal, and endodermal. The ectodermal part consists of the skin and adnexa, salivary glands, and glial tissue. The mesodermal component consists of mature cartilage, mature fat tissue, and muscle. The endodermal component consists of the gastrointestinal epithelium and the respiratory epithelium. Microscopic examination of tumor capsule showed a fibrous connective tissue stroma lined by stratified squamous epithelium with subepithelial vascular proliferation and distribution of fibroblasts and lymphoplasmacytic inflammatory cells.

All mature teratomas should be resected because they may become malignant, and even if they are benign, they can enlarge and impinge on nearby vital structures.[11] The approach is conservative posterolateral thoracotomy in all cases. Surgical resection without delay is a curative therapy and for definitive diagnosis of mediastinal teratoma.[3,9] The patient in this case underwent right posterolateral thoracotomy surgery and tumor resection and chest tube insertion.

An important differential diagnosis for anterior mediastinal teratoma are: another germ cell tumor in which at least two different germ cell layers are found; carcinoma, which is a very malignant epithelial tumor and does not show any other germ layer components; and sarcomas, which is containing only a mesenchymal component and showing no epithelium.[15]

CONCLUSION

Mature teratoma of the mediastinum is a rare neoplasm with a non-specific clinical manifestation ranging from asymptomatic to severe complications. CT imaging is the modality of choice for preoperative diagnosis. The histopathological features of mature teratomas contain two or three germ layers visible throughout the tumor in varying distributions, without an immature component. Early diagnosis, prompt and complete surgical excision of these tumors remain the only therapeutic approaches required.

CONFLICT OF INTEREST

There is no conflict of interest regarding the manuscript.

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None

AUTHOR CONTRIBUTION

I Gusti Ayu Sri Mahendra Dewi is responsible for the study from the conceptual framework.

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