

Pulmonology - Case Report

Mature Cystic Teratoma of the Mediastinum

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Background --- Mediastinal masses are relatively uncommon. For those which present as supraclavicular mass are often managed as tuberculosis or thymoma.

Case --- We present a case of a 24 year old male who presented with mass at the left supraclavicular area for five years. He was initially treated as a case of PTB. A series of chest x-ray done showed an increasing soft tissue mass density extending from supraclavicular area down to the level of the aortic arch in the middle mediastinum. CT scan revealed multiloculated mass in the left infrahyoid and adjacent to the left carotid midline structure to the right; the left thyroid gland appears compressed; findings suggestive of thymic cyst. An excision of the mass was done and revealed mature cystic teratoma arising from the thymic tissue. Likewise, cell block and cytology revealed content of epithelial cyst with squamous and probable glandular elements.

Conclusion --- In patients presenting with supraclavicular mass, one has to consider teratoma as one of the possible differential diagnosis. In such patients, additional work-ups, such as chest CT scan can help in the diagnosis. Surgical excision is often curative. *Phil Heart Center J 2012;16:68-71.*

Key Words: Mediastinal mass ■ Cystic Teratoma

Literature reports that approximately 20% of primary mediastinal neoplasms are of germ-cell origin, and of these, roughly 80% are teratomatous in origin. Mediastinal teratomas are five times more common in adults than in children. The reason behind how exactly these lesions originate remains an area of conjecture and discussion.¹

We present a case of a 24 year old Filipino male, who presented with mass at the left supraclavicular area for five years. He was initially managed as PTB. However, the impression changed to thymic cyst on chest CT scan. Excision of the mass revealed Mature Cystic Teratoma arising from the thymic tissue. Likewise, cell block and cytology revealed content of epithelial cyst with squamous and probable glandular elements. This case report aims to present a case of a twenty four-year old male with mature cystic teratoma; discuss the embryologic origin of thymus, etiology, diagnosis and management of mature cystic teratoma.

CASE PRESENTATION

This is a case of a 24-year old, male who came in with a chief complaint of a mass at the left supraclavicular area. The history of present illness started five years prior to admission when the patient noticed a slowly growing mass at the suprasternal area. It was soft, non-tender in character. There was no consult done, no medications taken. Four years prior to admission, the mass progressively enlarged extending to the left supraclavicular area, thus, he consulted at nearby clinic on which a chest x-ray was requested but the result was not known to the patient. He was then started with anti-TB medications (quadruple therapy) but took this only for a month. The patient was lost to follow up after then. He consulted at Makati Medical Center for a second opinion and was advised surgery during that time, but the patient refused. Eight months prior to admission, the mass grew bigger about five times its original size, now firm, non-tender with irregular borders

almost occupying the lateral aspect of the neck at the left, thus the patient consulted at a local hospital. A chest x-ray showed a soft tissue mass density in the left paratracheal area, extending into the left supraclavicular area with narrowing of the adjacent portion of the left clavicle and clear lung fields. Likewise, chest CT scan revealed multiloculated mass measuring approximately 11cmx7cmx4cm (LxWxP) in the left infrahyoid and adjacent to the left carotid midline structure to the right; the left thyroid gland appears compressed; findings suggestive of thymic cyst and other considerations include 2nd brachial cleft cyst or 4th brachial anomaly. Six months prior to admission, FAB with C/S showed negative result. Four months prior to admission, he consulted at Pulmonary OPD and chest X-ray revealed again a soft tissue density seen in the left paratracheal area extending into the left supraclavicular area associated with narrowing of the adjacent left clavicle.

The initial impression was a thyroid mass, rule out enlarged node or vascular lesion. A repeat FNAB revealed a brachial cleft cyst, thus he was advised admission. Family History, personal and social history was unremarkable.

Physical examination revealed no cervical lymphadenopathies, and a solid mass approximately 12x7cm in diameter at the supraclavicular area extending to the anterior chest, non-tender. Rest of physical examination is unremarkable. Patient was admitted with an initial Impression of a, Left supraclavicular mass, T/C Brachial Cleft Cyst vs. Dermoid cyst. Blood examinations done showed normal results ABG done at RA showed acceptable acid-base balance. Chest x-ray done showed an increase soft tissue mass density extending from supraclavicular area down to the level of the aortic arch in the middle mediastinum displacing the tracheal air column to the right with left clavicle relatively small in size and an ovoid density seen underlying the left 5th posterior rib. The rest of the visualized lung fields are clear. The patient was then referred to Thoracovascular Surgery service for co-management. PFT done which showed moderate restriction of the volume excursion of the lung [FVC- pre- 2.97 (60%) post-3.08 (62%); FEV1- pre 2.81 (69%) post 2.96 (73%); FEV1/FVC ratio pre-95, post-96]. B-HCG and alpha feto protein requested showing normal findings.

Ultrasound of the left supraclavicular mass showed a bipolar complex mass left upper hemithorax with supraclavicular and intrathyroid component with adjacent adenopathy. MRI of the neck done showing 13.7 x 6.2 cm rim enhancing mass occupying the supero-anterior mediastinum and left supraclavicular area and it displaces the sternocleidomastoid muscle anteromedially, the ipsilateral carotid vessels posteriorly and the trachea and esophagus to the right. On the 15th hospital day, the patient underwent excision of the left supraclavicular mass. Intraoperative course was unremarkable. The mass was seen beneath the sternocleidomastoid muscle noted to be firm and smooth with distinct borders, with cheesy fatty material fluid inside. The specimens sent for examination with frozen biopsy of the mass. Frozen biopsy showed mature cystic teratoma arising from the thymic tissue. Likewise, cell block and cytology revealed content of epithelial cyst with squamous and probable glandular elements.

Final diagnosis: Mature Cystic Teratoma

DISCUSSION

Several reports revealed that approximately 20% of primary mediastinal neoplasms are of germ-cell origin, of which, majority are teratomatous in origin. Mediastinal teratomas are five times more common in adults than in children. There is a relationship with Klinefelter's disease based on cytogenetic studies. This studies showed that the most commonly associated chromosomal abnormality associated with mediastinal teratomas is an isochromosome on the short arm of chromosome 12[(12p)].¹

Mature cystic teratomas, which accounts for 95% of all primary mediastinal teratomas, are large, benign lesions composed of one or more of the embryonic germ layers (ectoderm, mesoderm, and endoderm). When they are composed primarily of mature ectoderm, they are often referred to as "dermoids." In order to be classified as benign, there should be no trace of immature mesenchymal elements in the lesion.¹

In these reports, mature cystic teratomas are identical to their gonadal counterparts as far as gross and histologic features are concerned. Classically, these lesions present as large, polypoid

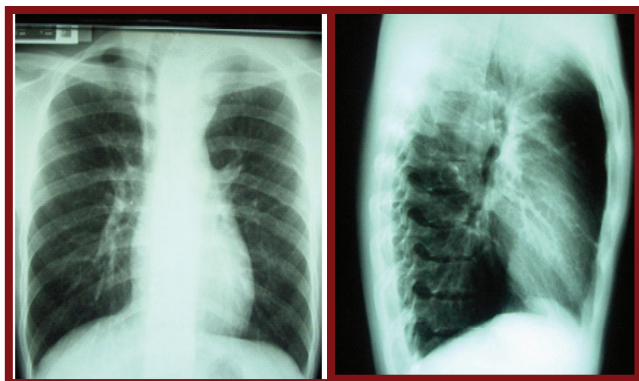


Figure 1. Chest X ray of a 24 year old male with mediastinal mass. An increased soft tissue mass density is seen extending from supraclavicular area down to the level of the aortic arch in the middle mediastinum displacing the trachea air column to the right. The left clavicle is relatively small in size. The ovoid density is seen overlying the left 5th posterior rib; bone island vs granuloma.

masses with focal intimal calcification. Residual thymus is also often adhered to the capsular surface or wall of these lesions. Sectioning usually exhibits fleshy, cystic areas with abundant sebaceous material and hair. Mature ectoderm is the most common of the three germ layers present, although bone, cartilage, respiratory epithelium and neural tissues may also be seen.¹

Reports cited that the typical presentations of these patients are symptoms related to compression and/or invasion of important structures found within the mediastinum. Of these, the most common complaints observed are cough, dyspnea, and chest pain. Occasionally, “the tumor may invade the adjacent pulmonary parenchyma and result in the expectoration of hair (trichoptysis). Additional complications, although infrequent, include pneumothorax, lipoid pneumonia, and destruction of nervous system structures”.¹ They also cited that radiographic studies such as chest x-ray and CT scans aid in the diagnosis, the only confirmation is the demonstration of the characteristic finding of teratoma such as the presence of teeth or other organized skeletal parts. Based on their report, the following should be considered as differential diagnoses: Mediastinal cystic hygromas, cysts of foregut origin, alimentary canal duplication and a variety of benign and malignant lesions.¹

Moreover, based on their reports, the prognosis of patients undergoing surgery in which the entire lesion is removed is excellent. In the absence of complications, surgery is a completely curative measure for these patients.¹

Agarwal and Kar cited in their case reports that teratomas are congenital tumors that contain derivatives of all three germ layers and arise from pluripotent embryonal cells. In their report, they state that superior mediastinal teratomas are usually asymptomatic till late, and are often discovered incidentally on chest x-ray.² In one report, the presenting symptoms such as chest pain, dyspnea or cough are a result of compression of nearby structures in these patients. Rarely, the teratoma may rupture into tracheo-bronchial tree or result in SVC syndrome or pneumonia.³ In our patient, although he presented with a rapidly enlarging, he was otherwise asymptomatic. This may be explained that the mediastinal mass found an escape route into the neck.

In the case report of Agarwal and Kar, they mentioned the utility of the chest x-ray in the diagnosis of a mediastinal teratoma. In one literature, the authors mentioned that the mediastinal CT scan demonstrates the extent of a mass better than conventional radiography. It can also detect fatty or cystic areas in mediastinal masses, but this information will not obviate the need for surgical resection to establish the final diagnosis.⁴

CT scan is useful in defining invasion of adjacent structures; thus, it assists the surgeon in the planning of surgical intervention.³ The CT scan of neck and mediastinum in our patient established the continuity of mediastinal mass into the neck and detected adherence of the mass to pericardium.

Verhaeghe et al stated that complete curative surgical removal of a mediastinal teratoma is the treatment of choice, as it establishes the diagnosis, besides preventing life threatening complications in many patients.⁵

In some reports, malignant mediastinal teratomas account for roughly 1–5% of all mediastinal tumors.⁶⁻⁷ When there is invasion or great vessels, myocardium, lung and phrenic nerves, these should be taken as indicators of malignancy, and may necessitate extensive operation in selected patients.⁶ Some studies reported that the complications of such extensive surgical procedures such as pneumonectomy, rather than the disease itself, may prove fatal.⁴ “Adherence in the mediastinal pleura and pericardium can be dealt with by removal of the involved portions.

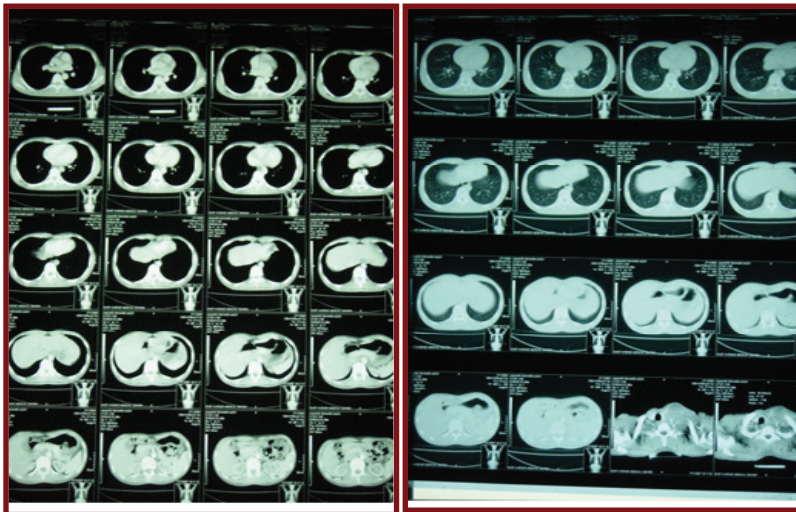


Figure 2: Chest CT Scan of a 24 year old male who presented with a mediastinal mass, which turned out to be a mature cystic teratoma.

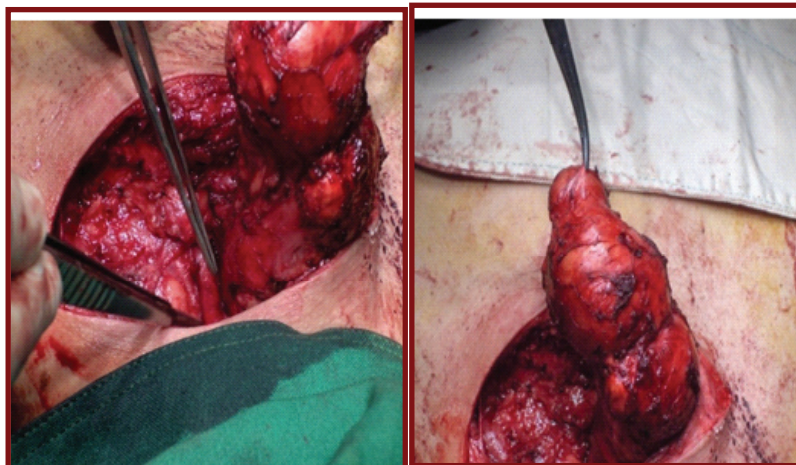


Figure 3: Intraoperative picture of a 24-year old male who presented with a mediastinal mass, which turned out to be a mature cystic teratoma.

As most mediastinal teratomas are benign, even a subtotal resection conserving adherent vital structures provides excellent results. In present era of modern surgical practices, excellent outcome has been the rule.²³

Our patient represents an unusual presentation of this not so uncommon pathological entity. Extension of a mediastinal teratoma into the neck and its cystic degeneration gave rise to this presentation.

SUMMARY

In summary, we presented a case of a 24 year old male who presented with mass at the left supraclavicular area for five years, and was initially treated as a case of PTB. A series of chest x-ray done showed an increasing soft tissue mass density extending from supraclavicular area down to the level of the aortic arch in the middle mediastinum. CT scan revealed multiloculated mass measuring approximately in the left in

frahyoid and adjacent to the left carotid midline structure to the right; the left thyroid gland appears compressed; findings suggestive of thymic cyst. The initial impression was left supraclavicular mass, T/C Brachial Cleft Cyst vs. Dermoid cyst. An excision of the mass was done and revealed Mature Cystic Teratoma arising from the thymic tissue. Complete curative surgical removal of a mediastinal teratoma was done as a treatment of choice.

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In Memory of
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