

## CASE REPORT- MESOTHELIAL CYST OF MEDIASTINUM

Somya Chaturvedi<sup>1</sup> and Jayashree Nadkarni\*<sup>2</sup>

<sup>1</sup>Senior Resident, Department of Paediatrics, Gandhi Medical College and Associated Kamla Nehru, Hamidia and SZ Hospitals, Bhopal (M.P.) 462016.

<sup>2</sup>Professor, Department of Paediatrics, Gandhi Medical College and Associated Kamla Nehru, Hamidia and SZ Hospitals, Bhopal (M.P.) 462016.

**\*Corresponding Author: Jayashree Nadkarni**

Professor, Department of Paediatrics, Gandhi Medical College and Associated Kamla Nehru, Hamidia and SZ Hospitals, Bhopal (M.P.) 462016.

Article Received on 26/09/2017

Article Revised on 17/10/2017

Article Accepted on 07/10/2017

### ABSTRACT

A rare case of a 3 year old female child who presented with complaints of swelling all over body. Initially managed as a case of fulminant hepatic failure. Later diagnosed as a case of mesothelial cyst of mediastinum on thoracotomy missed on a contrast chest CT. Early recognition of these relatively rare lesions would lead to immediate and appropriate surgical intervention. Early surgical intervention is also important because definitive histologic diagnosis can only be established by means of surgical extirpation.

**KEYWORDS:** Mesothelial cyst, fulminant hepatic failure, mediastinal cysts.

### INTRODUCTION

Mediastinum is defined as the portion of thorax extending from the posterior aspect of the sternum to the anterior surface of vertebral bodies and includes paravertebral sulci. The mediastinum is limited bilaterally by mediastinal parietal pleura and extends from the diaphragm inferiorly to the thoracic inlet superiorly.

Mediastinum is divided into 3 spaces or compartments – anterior, middle, posterior.<sup>[1]</sup>

Anterior compartment extends from the posterior surface of sternum to the anterior surface of pericardium and great vessels. Middle compartment extends from posterior limit of anterior compartment and anterior longitudinal spinal ligament. Posterior mediastinum is the area posterior to the heart and trachea and includes the paravertebral sulci.

Most common tumors and masses of anterior mediastinum are of thymic, lymphatic, germ cell origin, vascular or mesenchymal tissue and rarely bronchogenic cyst.<sup>[2]</sup>

Neoplasms of middle mediastinum are mostly of lymphatic origin, neurogenic tumors, cystic structures associated with developmental abnormalities of primitive foregut or precursors of pericardium or pleura.<sup>[3]</sup>

Neurogenic tumors are the most common neoplasms of the posterior mediastinum. Tumors originating from

lymphatic, vascular, or mesenchymal tissues can also be found in this area.<sup>[4]</sup>

In children and infants, neurogenic tumors are the most commonly occurring tumors or cysts.<sup>[4]</sup>

Mesothelial cysts are generally made up of a capsule of fibrous tissue with an inner single layer of mesothelial cells. The most common type of mesothelial cyst is the pleuropericardial cyst which is generally located in the anterior cardiophrenic angle.

Other mesothelial cysts occurring in mediastinum are simple mesothelial cysts and lymphogenous cysts. These abnormalities are considered benign lesions and produce symptoms due to their mass effect owing to the malleable nature and small size of the pediatric airway.<sup>[3]</sup>

### CASE REPORT

A 3year old female child presented to the emergency department with complaints of fever and swelling all over the body since 15 days. On examination, she had anasarca, pallor, tachypnea, tachycardia, firm hepatomegaly of span 11 cm.

The initial investigations revealed hemoglobin of 8, SGPT of 350, INR of 2.5, Negative for HBS antigen and anti HCV antibodies, Normal urine routine microscopy and serum bilirubin within the normal range.

She was managed symptomatically as a case of fulminant hepatic failure. Liver enzymes declined to 85 and INR to 1.5. Edema decreased, but tachycardia and tachypnea persisted.

Chest radiography suggested a collection around the heart borders with an enlarged CT ratio of 0.6. Ultrasound examination of chest revealed bilateral pleural effusion and underlying left side consolidation with a moderate pericardial effusion. ECHO was done that showed a mass near the pericardium, impinging it. No significant cardiac anomaly was noted.

Contrast enhanced CT scan revealed loculated pericardial effusion with a peripheral wall enhancement suggestive of large pericardial abscess alongwith right sided gross pleural effusion with multifocal collapse and consolidation. CT also showed short segment thrombus in right brachiocephalic and SVC alongwith congestive hepatomegaly with secondary budd chiari like feature.

She underwent a subxiphoid approach to drain the pericardial effusion. A minimal effusion was found, so from the same approach, right sided pleural effusion was

drained of around 500-600 ml and the chest tube was left in situ.

The condition of the patient did not improve and tachycardia and tachypnea persisted. This time a preop ultrasound of chest was done that revealed a mass around the pericardium, impinging the pericardium.

A posterolateral thoracotomy was performed and a large cystic mass was found adhered to pericardium as well as the pleura. Partial cystectomy was performed and 2 chest tubes were placed to drain the cystic fluid. Simultaneous histopathological sample was taken.

The chest tubes remained for a total period of 16 days. Her vitals improved dramatically. A broad spectrum antibiotic coverage and adequate nutrition was given.

Histopathology revealed fibromuscular cyst wall lined by flattened mesothelial cells. cystic fluid culture was sterile and negative for gram staining.

She was discharged successfully after 1 and a half months.

**CT SCAN IMAGES OF CYSTS**



Figure-1.

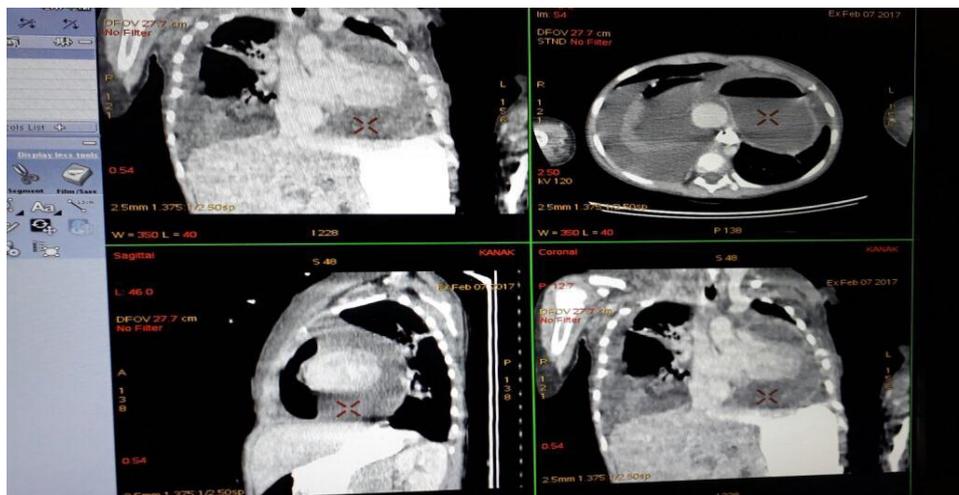
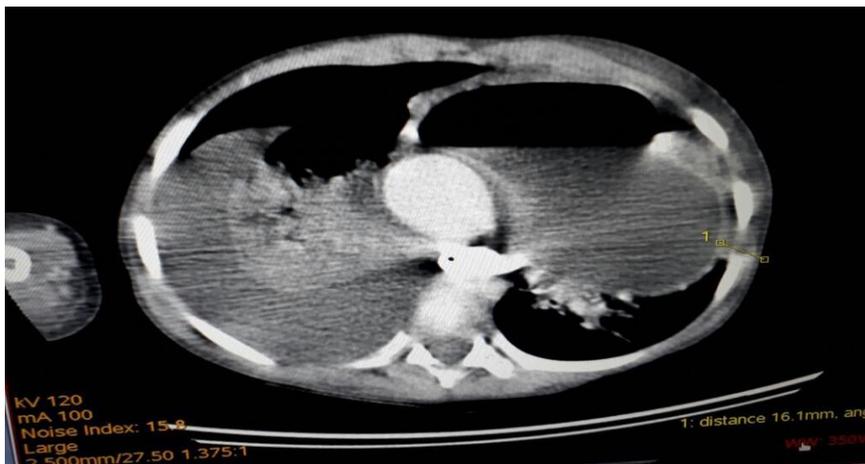


Figure-2.



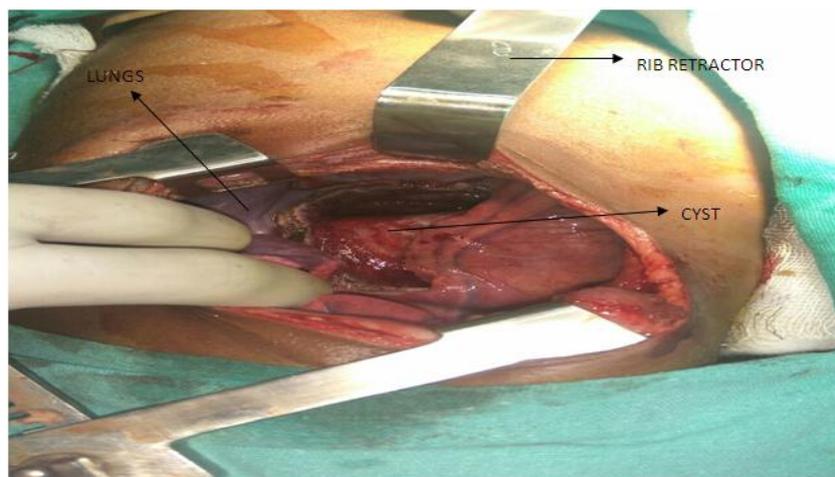
**Figure-3.**

The CECT thorax of the patient revealed loculated pericardial effusion with peripheral wall enhancement, most likely large pericardial abscess along with right

gross pleural effusion with multifocal, collapse consolidation of lung segments, and short segment thrombus in right brachiocephalic and SVC. (Figure 1-3).



**Image-1.**



**Image-2.**

The intraop images (image 1 and 2) show anterolateral thoracotomy done from the 5<sup>th</sup> intercostals space with ribs retracted by rib retractor. Image 2 reveals a large cyst densely adhered to pleura and pericardium.

**DISCUSSION**

The mediastinum is an area of the body in which a wide range of tissue variability exists. Therefore, tumors and

cysts that occur in this area can represent many different clinical entities and pathologic processes. An understanding of the embryology of this area and of the anatomic relationships of the normal structures within the mediastinum are essential in the proper determination of the exact nature of a mass or tumor located in this area.<sup>[5]</sup>

Mediastinal cysts are relatively uncommon but easily diagnosed by routine radiographic imaging procedures. These cysts comprise 12 to 30% of all primary mediastinal masses. Mediastinal cysts were further classified into foregut-derived cysts, which represented more than half of cystic lesions of the mediastinum, mesothelial cysts including pericardial pleural cysts, thymic cysts, and other rare anomalies such as thoracic duct cysts or meningoceles.

Mesothelial cysts, including pericardial and pleural cysts, are estimated to occur in approximately 1 in 100,000 persons. Ochsner and Ochsner reported 33% mesothelial cysts among 42 cysts in the mediastinum.<sup>[11]</sup> These anomalies are formed by the parietal recess persisting during development, namely aberrant recess fusion; therefore, the pericardial diverticulum, having a communication with the pericardial cavity, is regarded as an incomplete form of pericardial cyst in terms of its embryogenesis.

### SIGNS AND SYMPTOMS

A large percentage of mediastinal tumors and cysts produce no symptoms and are found incidentally during a chest radiograph or other imaging study of the thorax performed for some other reason as depicted in the image below.<sup>[4]</sup> Symptoms are present in approximately one third of adult patients with a mediastinal tumor or cyst but are more commonly observed in the pediatric population, in which nearly two thirds of individuals present with some symptoms. In adults, asymptomatic masses are more likely to be benign.<sup>[6]</sup>

Symptoms associated with the respiratory tract predominate in pediatric patients because airway compression is more likely. This occurs because of the significant malleability of the airway structures and the small size of the chest cavity in infants and children. Symptoms most commonly observed include persistent cough, dyspnea, and stridor. If the location and size of the mass produces partial or complete obstruction, obstructive pneumonia can also occur. Infectious symptomatology, and even signs of sepsis, can occur if a mediastinal cyst becomes infected.<sup>[8]</sup>

Constitutional symptoms, such as weight loss, fever, malaise, and vague chest pain, commonly occur in association with malignant tumors in pediatric patients but also may be signs associated with secondary infection of a congenital cyst, such as a bronchogenic cyst.

Symptoms associated with compression of some portion of the respiratory tract can be produced by benign lesions in adults. This is much less likely to occur in adults compared to children. Infectious symptoms or sepsis from infection of a mediastinal cyst can also occur in adults, although, again, this is very unlikely in persons in this age group.<sup>[10]</sup>

Patients with pericardial/pleural cysts have lower incidences of symptoms than other mediastinal cysts and are mostly asymptomatic. Other symptoms include-Chest pain, Dyspnea, Cough, Fever, Hoarseness, Sputum, Dysphagia, Cyanosis, Hemoptysis. Although our patient had a rare presentation of congestive hepatomegaly.<sup>7</sup>

### TREATMENT

Treatment selection for a given mediastinal tumor or cyst depends on the diagnosis of the lesion being investigated. Surgical resection is indicated in a large percentage of cases.

Surgical resection is indicated for most mediastinal cysts. All large and symptomatic bronchogenic cysts are included in the group, as are all enteric gastroenteric and neurenteric cysts. Some authors do not recommend resection of small asymptomatic bronchogenic cysts, while others advise aspiration of such cysts and resection only for those with symptoms or recurrence.

Excision of pleuropericardial cysts and simple mesothelial cysts is indicated for diagnostic purposes only. Thymic cysts require excision or enucleation. Thoracic duct cysts require excision and accompanying ligation of the thoracic duct.<sup>[8]</sup>

Surgical removal is not indicated as primary treatment for

- (1) Pleuropericardial cysts, lymphogenous cysts, and simple mediastinal cysts, provided an accurate nonsurgical diagnosis has been performed and
- (2) Some cases of bronchogenic cysts found in adult patients in which the cyst is small and the patient has no symptoms.<sup>[10]</sup>

### WORKUP

#### Chest radiography

Posteroanterior (PA) and lateral radiograph of the chest for an unrelated cause is the usual way in which an asymptomatic mediastinal mass is identified.

#### Esophagram

In the past, barium esophagram findings have been used to help delineate masses or cysts found in the mediastinum. The CT scan has generally replaced this study for the evaluation of most foregut cysts, especially bronchogenic cysts.

Barium esophagram has been used in patients who primarily present with symptoms of dysphagia. The displacement of the esophagus by neurenteric cysts and enterogenous or duplication cysts of the esophagus is usually seen clearly.

#### CT scan of the chest and mediastinum

CT scan is a routine part of the diagnostic evaluation of mediastinal tumors, cysts, and other masses.

CT scan images can greatly assist in determining the exact location of the mediastinal tumor and in determining its relationship to adjacent structures. The CT scan findings are also useful for differentiating masses that originate in the mediastinum from those that encroach on the mediastinum from the lung or other structures.

While not infallible, CT scan images are very useful in differentiating tissue densities. This greatly assists in distinguishing structures that are cystic or vascular from those that are solid.

CT scanning with contrast has, in most cases, virtually replaced the barium esophagram as an evaluation tool for these abnormalities in infants and children.

CT myelography has been performed in individuals with neurenteric cysts and neurologic symptoms to help determine the presence and level of spinal cord compression.<sup>[11]</sup>

### Magnetic resonance imaging

MRI is useful in both the initial diagnosis of a mediastinal mass and in follow-up evaluations after treatment. It provides superior vascular imaging and can help better delineate the relationship of an identified mediastinal mass to nearby intrathoracic vascular structures. It can be used to help differentiate between a possible mediastinal mass and a vascular abnormality such as an aortic aneurysm.

MRI offers direct multiplanar imaging. It can be used when iodinated contrast is contraindicated. Images provide increased detail in the subcarinal and aortopulmonary window areas and in the inferior aspects of the mediastinum at the level of the diaphragm.

MRI is superior to CT scan for the evaluation of masses located at the thoracic inlet or at the thoracoabdominal level.

In infants and children, MRI is excellent for helping delineate the relationship of a neurenteric cyst with the spinal canal and the associated vertebral abnormalities.

Because the patient is not exposed to radiation, MRI may be preferred to CT scan by some physicians for the evaluation of a mediastinal cyst in a child or infant. However, children and infants require sedation when undergoing MRI. This factor may increase the risk for those pediatric patients with signs of airway obstruction who require such an examination. The physician may have to decide which risk is greater.<sup>[12]</sup>

Radionuclide scanning: The technetium Tc 99 pertechnetate scans have been used to help identify neuroenteric cysts in the mediastinum, producing positive isotope uptake in gastric mucosa lining the cyst.

### Echocardiography and ultrasonography

Ultrasonographic methods have been used to differentiate solid from cystic mediastinal masses and to assist in determining connections between a mass and adjacent structures.<sup>[13]</sup> These studies are more useful in the evaluation of masses associated with the heart and in vascular abnormalities. In general, given the accuracy and detail provided by CT scan images, MRI, and selected radionuclide scan images, ultrasound techniques are not generally used as primary tools in the evaluation of mediastinal tumors and cysts.<sup>[14]</sup>

Prenatal ultrasonography has been helpful in the discovery of a number of abnormalities in the fetal thorax, including bronchogenic cysts. These findings can greatly enhance care of the newborn.<sup>[15]</sup>

### Arteriography

Very few indications exist for this procedure in the evaluation of a mediastinal cyst.

Arteriography may be helpful in differentiating between a bronchogenic cyst and an extralobar sequestration of the lung. It also may assist in determining whether the lesion in question originates from a mediastinal vascular structure, such as the heart or a great vessel.

### CONCLUSION

Mediastinal masses are although rare but easily diagnosed and treatable conditions. Clinical symptoms and signs are non specific, hence a strong clinical suspicion is required. Early diagnosis and prompt surgical referral may improve outcome in a majority of patients.

### BIBLIOGRAPHY

1. Priola AM, Priola SM, Cardinale L, Cataldi A, Fava C. The anterior mediastinum: diseases. *Radiol Med*, 2006 Apr; 111(3): 312-42.
2. Strollo DC, Rosado de Christenson ML, Jett JR. Primary mediastinal tumors. Part 1: tumors of the anterior mediastinum. *Chest*, 1997 Aug.; 112(2): 511-22.
3. Strollo DC, Rosado-de-Christenson ML, Jett JR. Primary mediastinal tumors: part II. Tumors of the middle and posterior mediastinum. *Chest*, 1997 Nov 5; 112(5): 1344-57.
4. Reynolds M. Foregut cysts of the mediastinum in infants and children. Shields TW, LoCicero J III, Ponn RB, eds. *General Thoracic Surgery*. 5th ed. Philadelphia, Pa: Williams & Wilkins, 2000; 2: 2393-9.
5. Whooley BP, Urschel JD, Antkowiak JG, Takita H. Primary tumors of the mediastinum. *J Surg Oncol*, 1999 Feb.; 70(2): 95-9.
6. Shields TW. Mesothelial and other less common cysts of the mediastinum. Shields TW, LoCicero J III, Ponn RB, eds. *General Thoracic Surgery*. 5th ed. Philadelphia, Pa: Williams & Wilkins, 2000; 2: 2423-35.

7. Kaiser LR. Thoracoscopic Resection of Mediastinal Tumors and the Thymus. *Chest Surgery Clinics of North America*, 1996; 6: 41-52.
8. Luketich JD, Ginsberg RJ. The current management of patients with mediastinal tumors. *Adv Surg*, 1996; 30: 311-32.
9. Giron J, Fajadet P, Sans N, et al. Diagnostic approach to mediastinal masses. *Eur J Radiol*, 1998 Mar.; 27(1): 21-42.
10. Le Pimpec-Barthes F, Cazes A, Bagan P, Badia A, Vlas C, Hernigou A, et al. [Mediastinal cysts: clinical approach and treatment]. *Rev Pneumol Clin*, 2010 Feb.; 66(1): 52-62.
11. Ochsner JL, Ochsner F. Congenital cysts of the mediastinum: 20-year experience with 42 cases. *Ann Surg*; 1966; 163: 909– 920.
12. Laurent F, Latrabe V, Lecesne R, et al. Mediastinal masses: diagnostic approach. *Eur Radiol*, 1998; 8(7): 1148-59.
13. Durand C, Baudain P, Nugues F, Bessaguet S. Mediastinal and thoracic MRI in children. *Pediatr Pulmonol Suppl*, 1999; 18: 60.
14. Ha C, Regan J, Cetindag IB, Ali A, Mellinger JD. Benign esophageal tumors. *Surg Clin North Am*, 2015 Jun.; 95(3): 491-514.
15. Ardengh JC, Bammann RH, Giovani M, Venco F, Parada AA. Endoscopic ultrasound-guided biopsies for mediastinal lesions and lymph node diagnosis and staging. *Clinics (Sao Paulo)*, 2011; 66(9): 1579-83.
16. Geibel A, Kasper W, Keck A, et al. Diagnosis, localization and evaluation of malignancy of heart and mediastinal tumors by conventional and transesophageal echocardiography. *Acta Cardiol*, 1996; 51(5): 395-408.