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A CASE OF GIANT LYMPHANGIOMA OF NECK AND MEDIASTINUM PRESENTING WITH BREATHLESSNESS

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ABSTRACT
Lymphangiomas are rare benign lesions of lymphatic origin mostly present at birth and consist of focal proliferations of well-differentiated lymphatic tissue. They represent 0.7%–4.5% of all mediastinal tumors, majority are discovered during the first 2 years of life. They are most common in the neck and axilla, and about 10% extend into the mediastinum. Approximately 1% of all Lymphangiomas are confined to the chest. In this study we present a case presented with breathlessness with an incidental right sided neck swelling noticed during examination. Patient was evaluated with chest X-ray, USG and CECT thorax which revealed a cystic mediastinal lesion with internal septa and superior extension to posterior triangle of neck on right side. The lesion was excised and the histopathological examination revealed features of Lymphangioma.

HISTORY AND CLINICAL FINDINGS

- 40 year old female.
- Presented with breathlessness for 4months
- No history of pain, fever or trauma.
- On examination- A swelling was incidentally noted on right side of the neck in the posterior triangle deep and posterior to right sternocleidomastoid muscle.
- Patient was advised a chest X ray for the chest discomfort and USG for the neck swelling followed by CECT thorax.

CHEST X RAY

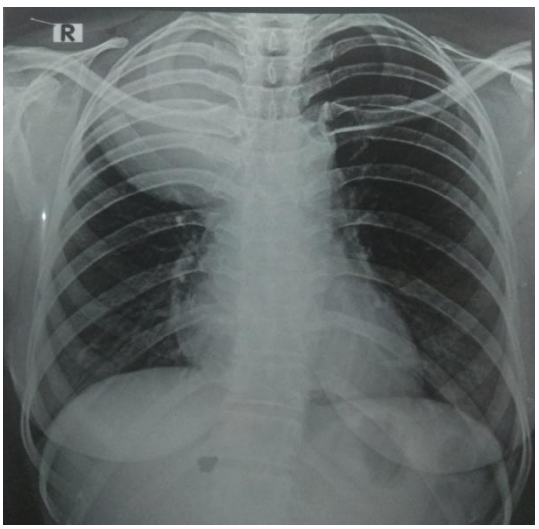

Fig 1: Chest X ray PA view of the patient showing a well defined homogeneous opacity in the right side of superior mediastinum.

- The chest X-ray showed a well defined homogeneous opacity in the right side of superior mediastinum with mediastinal widening and extending to right apical and upper zones.

- There was no significant tracheal or mediastinal shift.

USG OF THE NECK SWELLING

- The neck swelling on USG showed a well defined cystic lesion with few thin hyper echoic septa within. No vascularity was noted on colour doppler evaluation.
- There was no infiltration to adjacent tissues. There was no associated lymphadenopathy.

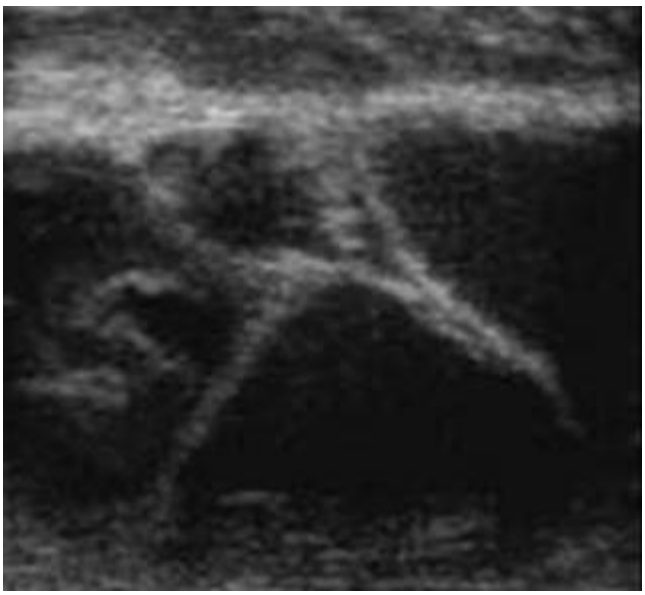

Fig 2: USG of the neck swelling showing multiple anechoic areas with thin septa within.

HRCT AND CECT OF THORAX

HRCT and CECT study of chest revealed a well defined lesion of fluid attenuation in the right side of superior mediastinum, extending to right hemi thorax compressing right upper lobe antero-laterally. The lesion was encasing trachea and primary bronchi and insinuating between mediastinal structures. The lesion measured 31x 14x 12 cm in greatest craniocaudal and cross sectional diameters. The lesion is noted extending superiorly to neck conforming to a dumb-bell shape. The cervical component of the lesion was noted extending to the right posterior triangle of neck. On post contrast study there is no significant enhancement of the lesion suggesting cystic characteristics.

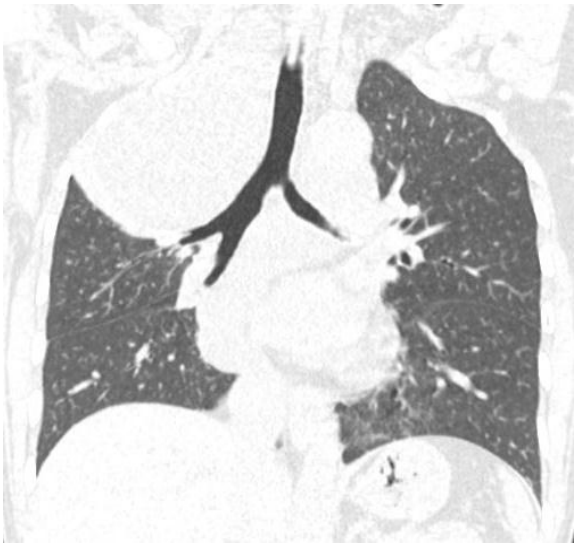

Fig 3: HRCT chest coronal image shows a well defined opacity in the right side of superior mediastinum, extending to right hemi thorax.

Fig 4: HRCT chest axial image shows compression of right upper lobe anterolaterally by the lesion

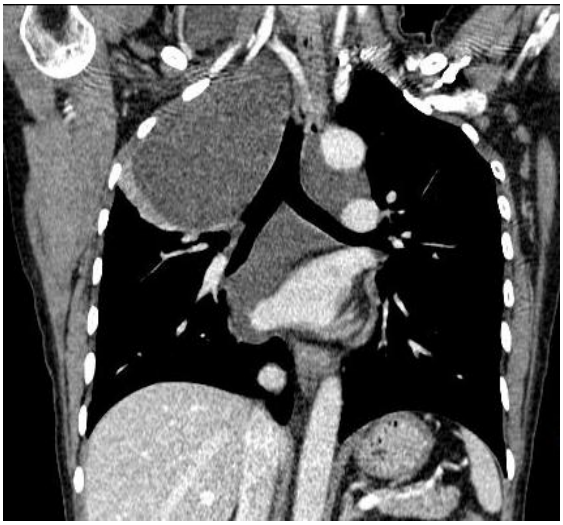

Fig 5: CECT chest coronal image shows a well defined non enhancing cystic lesion involving superior mediastinum encasing trachea and primary bronchi and insinuating between mediastinal structures.

Fig 6: CECT chest and neck coronal image shows the superior extension of the mediastinal lesion to the neck causing a dumbbell shape.

Fig 7: The CECT thorax axial image shows insinuation of the lesion between mediastinal structures, with mild compression of SVC anteriorly.

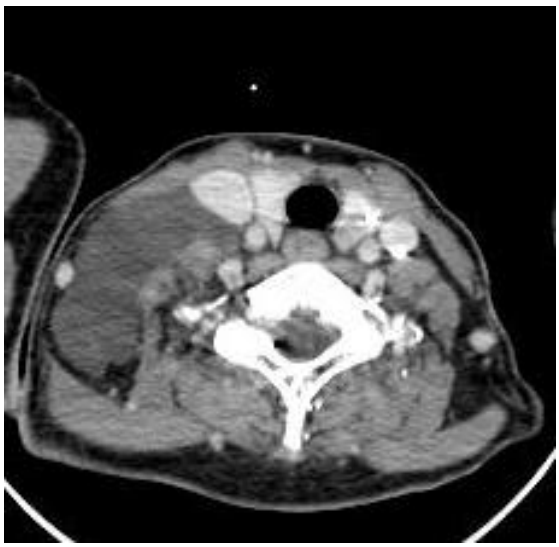

Fig 8: CECT axial image of neck shows the cervical component of the lesion extending superiorly to the right *posterior triangle* of neck.

The patient underwent thoracotomy with excision of the lesion under general anesthesia. The lesion was sent for histopathological examination

HISTOPATHOLOGY

Multiple lymphatic channels within stroma of loose connective tissue. There were peripheral lymphoid aggregates- suggestive of Lymphangioma.

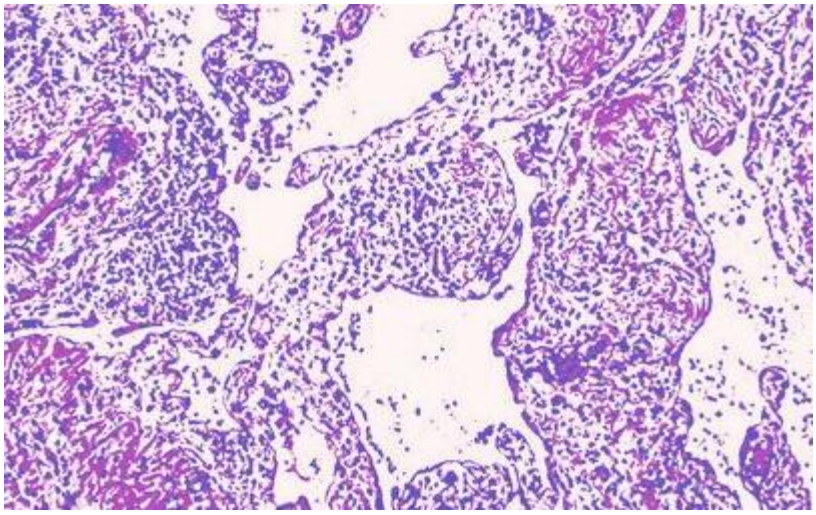

Fig 9: lymphatic channels with peripheral lymphoid aggregates

DISCUSSION

- Lymphangiomas are rare benign lesions of lymphatic origin and are mostly present at birth.(1)
- They consist of focal proliferations of well-differentiated lymphatic tissue and represent 0.7%–4.5% of all mediastinal tumors. Majority are discovered during the first 2 years of life. They are most commonly seen in the regions of neck and axilla, with about 10% extend into the mediastinum. One percent of all Lymphangiomas are confined to the thorax (2,3).
- **PATHOPHYSIOLOGY**- Lymphangiomas are caused by congenital or acquired abnormalities of the lymphatic system. The congenital form typically presents before the 5 years of age and is caused due to abnormal connection of lymphatic channels to the main drainage duct. Acquired Lymphangiomas occur as a result of surgery, malignancy, trauma, and radiation therapy due to interruption of previously normal lymphatic drainage such as.(4)
- **CLASSIFICATION**-They are histologically classified as simple capillary, cystic or cavernous, depending on the size of the channel within them. Cystic Lymphangiomas are the most common type.
- **COMPLICATIONS** include secondary infection, chylothorax, airway compromise and chylopericardium
- **IMAGING**
- On **chest radiographs**, Lymphangiomas appear as well-defined, focal mass in upper mediastinum with mediastinal widening(1).
- In **CT** imaging the lesion appears as a lobulated, smooth mass, which may mold to the adjacent mediastinal structures (2). Lymphangiomas usually have homogeneous low attenuation, but may show higher attenuation .Calcification is rare. Lymphangiomas may be either unilocular or multilocular. Thin septations within the mass may be seen sometimes (5).
- At **MR imaging**, the lesions have heterogeneous signal intensity on T1-weighted images. They usually have high signal intensity on T2-weighted images, reflecting their fluid content (2).

TREATMENT

- For any type of Lymphangioma, whenever feasible the treatment of choice is surgical excision. As recurrence is common. Wide local excision of the affected lymphatic channels is necessary

CONCLUSION

- Integrated and correlative diagnostic imaging and HPE is useful for the diagnosis of Lymphangiomas.
- Imaging plays an important role in clearly demonstrating lesion and its extension, thus aiding management.
- Mediastinal Lymphangioma is a rare condition, especially in adults and because of their insinuating nature, complete surgical resection of Lymphangiomas may be difficult, and follow-up may be needed to exclude recurrence

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