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Case Report

Posterior Mediastinal Ancient Schwannoma Mimicking a Cyst

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Abstract:

Background: Several types of neurogenic tumors can occur in mediastinum and commonly present as posterior mediastinal mass lesions. Generally they are categorized in to those arising from peripheral nerves or sympathetic ganglia. Schwannoma is the most common neurogenic tumor of this area. Generally they are presented as solid encapsulated lesions and are difficult to miss on radiology. Rarely they can be large enough to undergo cystic degeneration and cause confusion with other cystic mass lesions of the area. Large masses can also cause pressure symptoms and leads to respiratory compromise.

Conclusion: Posterior mediastinal schwannomas though usually are benign and asymptomatic but can prove fatal if attain a very large size. It should always be included in the differential diagnosis of posterior mediastinal large cystic masses. Degenerative nuclear atypia associated with ancient schwannoma should not be confused with malignancy. Patient can lead a symptom free life after surgical resection of these tumors.

Keywords: Posterior mediastinum, Schwannoma, Peripheral nerves

Introduction

Mediastinal masses are frequently encountered in clinical practices of pulmonologists and cardiothoracic surgeons and include various types of cysts and benign or malignant tumors. They differ by location and age of the patient. For anterior, middle and posterior mediastinal compartments each has different sets of these entities[1]; Thymic tumors, Hodgkins and non Hodgkins lymphoma and parathyroid adenomas are commonly located in anterior mediastinum.[2] Thyroid tumors, lymphoma, tracheal tumors etc. can be seen in middle mediastinum, while several types of neurogenic tumors, esophageal tumors, and duplication cysts are usually found in the posterior mediastinum.[3]

Neurogenic tumors constitute about 20% of all adult and 35% of all pediatric mediastinal neoplasms. They are grouped as those arising from peripheral nerves, sympathetic ganglia, or rarely parasympathetic ganglia. Schwannoma, neurofibroma, and malignant tumor of nerve sheath origin arise from the peripheral nerves while ganglioneuroma, ganglioneuroblastoma, and neuroblastoma arise from the sympathetic ganglia.[3, 4]

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Schwannomas (or neurilemomas) are the most common benign neurogenic neoplasms of posterior mediastinum. They believed to be originated from costovertebral sulcus as a solitary benign lesion.[4] Mostly they are asymptomatic in adults but can attain a large size to cause various cardiopulmonary problems.[5, 6] They are diagnosed radiographically as encapsulated, smooth, lobulated and rounded mass in paravertebral region and can rarely exhibit cystic degenerative changes. If these changes are extensive they can easily be confused with variety of other cystic masses of the region especially those of congenital and infectious etiology.[7] Same is true for the current case which was clinically and radiologically puzzled with Hydatid and Bronchogenic cysts due to large size and cystic nature of the lesion but proved to be posterior mediastinal anciant schwannoma on histopathology.

Case Presentations

A 43 years old lady with no other co-morbid presented with mild to moderate left sided dull chest pain and productive cough for last 6 months. It was associated with malaise and body aches. She used to work with household cattle and was occasional huqqa smoker. Workup for pulmonary tuberculosis was already done and no evidence of tuberculosis was seen. She also received 2-3 courses of antibiotics but there was no

improvement in her condition. Her chest examination revealed mild local tenderness along with decreased air entry in left upper chest. Right sided chest was unremarkable. CT chest revealed a cystic mass occupying upper and middle left hemi thorax. It was thought to be originated from posterior mediastinum because the lung parenchyma was unremarkable. Collapse of upper left lung with compression of lower part was attributed to its mass effect. Figure. 1

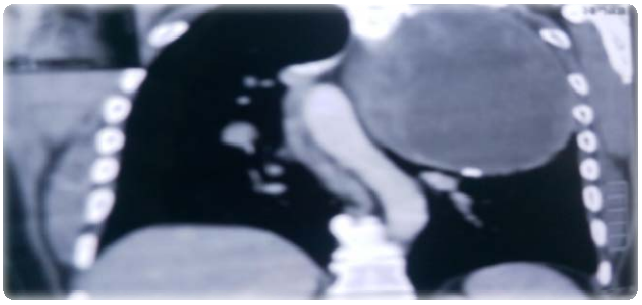

Figure-1: CT scan showing a cystic mass in left upper hemi thorax with surrounding compressed lung parenchyma.

Bronchogenic and Hydatid cysts or any mass lesion with cystic degeneration were among the top differentials in this case. Hydatid serology proved to be negative and abdominal ultrasound revealed unremarkable liver. For confirmation of diagnosis left thoracotomy was performed. A well circumscribed mass lying free of lung in upper mediastinum was identified which was attached to the inner side of 2nd rib. Mass was completely excised and sent for histopathology. Figure. 2

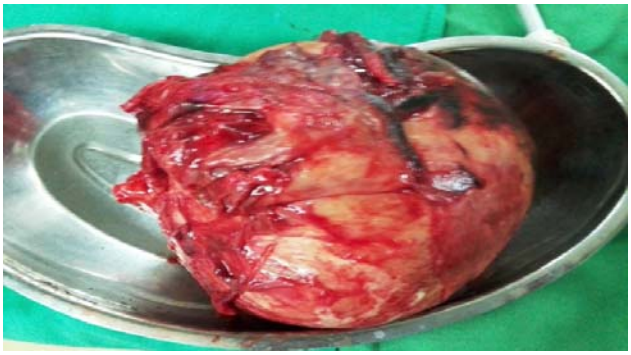

Figure-2: Completely excised left posterior mediastinal mass.

Grossly the mass measured 20x13x11 cm and weighed around 698 grams, Cut section revealed multiloculated cystic areas filled with straw coloured fluid admixed with solid areas. No necrosis or hemorrhage seen. Microscopy revealed a partly encapsulated, paucicelluar benign spindle cell tumor containing thick walled hyalinised vessels. Areas of haemorrhage and dystrophic calcification were also seen. Tumour cells show mild nuclear atypia with focal Verocay body formation. Figure. 3. Tumor cells were S-100 positive and show negativity for SMA and CD34. Ki-67 index was <1%. Figure. 4. Based on above histological and immunohistochemical findings diagnosis of Ancient schwannoma was made.

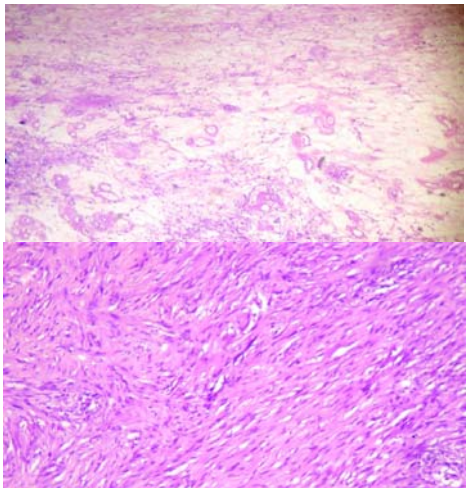

Figure 3: H&E: Photomicrographs revealing Antoni A, B areas, thick walled blood vessels and mild nuclear atypia. (H&E X 40 and H&E X 100)

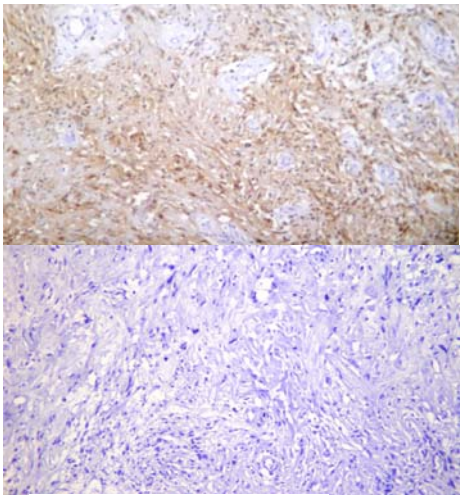

Figure. 4: Immunohistochemistry: Photomicrographs showing diffuse S-100 positivity and <1% Ki-67 positivity. (IHC X 40).

Patient was discharged after a week of surgery and her course of recovery was unremarkable. At a follow-up period of six months she was recurrence free.

Discussion

Neurogenic tumors are the fourth most common mediastinal tumors after thymoma, germ cell tumors and lymphoma. They share 12-20% bulk of all these tumors in adults.[4, 8] Mostly they involve posterior mediastinum and schwannoma is commonest of them all. This benign neurognic tumor is originated from Schwann cells which are main neuroglial cells of peripheral nerves.[1, 9] In mediastinum they commonly arise from intercostals nerves and generally located in paravertebral regions. Mostly they arise as a single lesion in costovertebral sulcus and rarely as multiple masses. From intervertebral opening they can also extend to thoracic cavity and may form dumb-bell tumors.[4] Prevalence is equal in both sexes and ages of 40 or more are commonly affected.

Most patients remain asymptomatic but if it attains a large size it can cause pressure symptoms by compressing lung and air way, esophagus, right heart or even great veins.[6] Large tumors especially those which underwent cystic degeneration can be confused with other cystic lesions of mediastinum including those of congenital, infectious, or neoplastic etiology. Main differentials are parasitic (Hydatid) cyst, foregut cysts including bronchogenic, esophageal duplication and and neurenteric cysts, meningocele, cystic teratoma. esophageal tumors, lung primary with cystic degeneration.[7]

Initial work-up for their evaluation requires radiographic assessment. Asymptomatic cases are mostly discovered on plain radiographs. To locate the exact position of mass and its relationship with adjacent structures CT scan and MRI can be very helpful.[6] Grossly schwannomas are encapsulated, firm grey masses and at times seen in close relationship with the nerve of origin. On microscopic examination the usually exhibit a dimorphic histology with Antoni A and B areas. Antoni A areas are more cellular and composed of interlacing bundles of smooth showing focal nuclear palisading and total lack of or low mitotic activity. In contrast Antoni B areas are hypocellular with loose myxoid stroma and lack any specific histological features.[10] Ancient schwannomas also exhibit certain types of degenerative changes including mild to moderate nuclear atypia, hyalinization of vessel walls and xanthomatous change. All these changes have no prognostic effect and their malignant transformation is quite a rare event.[11] Diffuse and strong staining of S-100 is required to make a final diagnosis while low or negative Ki-67 expression highlights its low proliferation capacity.[10]

Surgical resection is the treatment of choice and Posterolateral thoracotomy is a preferred surgical approach for large posterior mediastinal tumors.[12]

Based on above mentioned facts the peculiar histological features of spindle cells with fibrous background, degenerative changes, diffuse and strong S-100 staining, negligible Ki-67 index and negativity for Pan-CK favors the diagnosis of Ancient schwannoma for the case under discussion.

In conclusion, though Ancient schwannoma is a benign tumor but it can attain large size to cause compressive symptoms and can develop cystic degenerative changes. This degenerative nuclear atypia and its larger size should not be confused with malignancy and it should also taken as one of differentials while evaluating the cystic masses of mediastinum.

References

1. Pakseresht K, Reddymasu SC, Oropeza-Vail MM, Fan F and Olyaee M. Mediastinal Schwannoma Diagnosed by Endoscopic Ultrasonography-Guided Fine Needle Aspiration Cytology. Case Reports in Gastroenterology. 2011; 5(2):411-415.
2. Strollo DC, Rosado de Christenson ML and Jett JR. Primary mediastinal tumors. Part I: Tumors of the anterior mediastinum. Chest. 1997; 112:511–522.
3. Strollo DC, Rosado de Christenson ML and Jett JR. Primary mediastinal tumors. Part II. Tumors of the middle and posterior mediastinum. Chest. 1997; 112: 1344–1357.
4. Negri G, Bandiera A, Carretta A, Puglisi A, Mandelli C, and Ciriaco P, et al. Unusual presentation of mediastinal neurogenic tumors. Case Rep Surg. 2013; 2013:414260.
5. Kato M, Shiota S, Shiga K, Takagi H, Mori H and Sekiya M, et al. Benign giant mediastinal schwannoma presenting as cardiac tamponade in a woman: a case report. J Med Case Rep. 2011; 5:61.
6. Amin R and Waibel BH. An Unusual Presentation of a Posterior Mediastinal Schwannoma Associated with Traumatic Hemothorax. Case Rep Surg. 2015; 2015:175645.
7. Odev K, Arıbaş BK, Nayman A, Arıbaş OK, Altınok T, and Küçükapan A. Imaging of Cystic and Cyst-like Lesions of the Mediastinum with Pathologic Correlation. J Clin Imaging Sci. 2012; 2:33.
8. Whooley BP, Urschel JD, Antkowiak JG and Takita H. Primary tumors of the mediastinum. J Surg Oncol. 1999; 70(2):95-9.
9. Reeder LB. Neurogenic tumors of the mediastinum. Semin Thorac Cardiovasc Surg. 2000; 12(4):261-7.

10. Quartey B, Lenert J, Deb SJ, Henry LR. Giant posterior mediastinal ancient schwannoma requiring thoracoabdominal resection: a case report and literature review. World J Oncol. 2011; 2:191–4.
11. Ortigara L, Rosemberg N, Siqueira R, Neto F. Resection of a mediastinal schwannoma using video-assisted thoracoscopy. J Bras Pneumol. 2006; 32(2):172-5.
12. Fierro N, D'ermo G, Di Cola G, Gallinaro LS, Galassi G, Galassi G. Posterior mediastinal schwannoma. Asian Cardiovasc Thorac Ann. 2003; 11(1):72-3

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