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HUGE POSTERIOR MEDIASTINAL LIPOSARCOMA: A CASE REPORT

Surgery

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ABSTRACT

Liposarcomas of the Mediastinum are extremely rare tumours, which become symptomatic only when they are unusually large and therefore producing compressive symtpoms. We present a case of a 60-years-old male who presents to surgical thoracic clinic with signs and symptoms of congestive heart failure secondary to a huge mass in the mediastinum compressing his heart. He underwent curative surgical excision and was followed with radiation therapy, thereby reversing the patient's initial illness.

KEYWORDS

Mediastinal, Liposarcoma, huge mediastinal tumor, lateral thoracotomy, myxoid

INTRODUCTION:

Liposarcomas are common soft tissue sarcoma tumors, usually occurring in extremities or the retroperitoneum. However they are quite rare to arise from mediastinum, representing <10% of primary sarcomas (1) and <1% of all mediastinal tumors (2). They are usually large, rapidly progressive and silent in early illness or associated with compressive symptoms if late in the disease process. Liposarcomas frequently recur, therefore location of the tumor plays a major prognostic role, as it determines the feasibility of complete curative surgical excision, and later recurrence. If not achieved patient is required to follow up with other therapy modalities, such as radiotherapy and/or chemotherapy, with surgical re-excision being one of them.

This report discusses a patient with posterior mediastinal liposarcoma diagnosed on fine needle aspiration biopsy. Written consent was obtained for the academic purposes of sharing this case.

Case Presentation:

A 60-year-old man with known history of Asthma presented to our hospital with rapidly progressive dyspnoea, orthopnea & dysphagia over the past 3 months.

Physical examination of the patient was unremarkable, and laboratory investigations were normal.

However, chest Xray showed wide mediastinum and a well-defined hyperdensity occupying the posterior mediastinum. Past surgical history was only significant for a mass excision from the right popliteal fossa performed in 2001 and later followed by chemotherapy.

Subsequently he got a contrast enhanced Computed Tomography of the chest which revealed; a non-enhancing fluid density in the posterior mediastinum extending from T2-T11 measuring 16x13x9cm, displacing nearby pulmonary vessels, bronchi, descending thoracic aorta, heart and the oesophagus with proximal dilation. CT guided biopsy revealed sticky hemoserous fluid, with negative culture and AFB.

Cytology revealed low grade myxoid spindle cell lesion. There were oval & spindle cells with moderate amount of esinophilic tailing cytoplasm, vesicular nucleus with mild anisonucleosis, coarse chromatin, small inconspicuous nucleoli arranged in sheets with no particular pattern. Interstitium shows myxoid change. Tumor is interspersed with very few lymphocytes, plasma cells and proliferating blood vessels lined by prominent endothelial cells. No evidence of increased mitosis and anplasia. One cluster of reactive mesothelial cells noted. Immunohistochemistry reported negative for S100 and SOX100, as well as negative Ki67 factor therefore ruling out neurogenic or high-grade tumours respectively.

Video Assisted thoracoscopic Surgery (VATS), was attempted initially, as there were some discussion that large part of this mass is cystic and could be aspirated. However significant adhesions were encountered, reduction of the tumor size couldn't be done. Therefore, adhesion lysis done and surgery converted to open right postero-lateral thoracotomy. Incision was commenced at the 5th intercostal space. Serratus anterior muscle was preserved, while other muscles were divided. Adequate field of vision was achieved with self-retaining retractors. The mass was carefully dissected from surrounding structures including the right upper and lower lung lobes, carina, spine and oesophagus, with preservation of the right phrenic and vagus nerves. Feeder vessels were respectively ligated, and haemostasis secured. A remnant of the tumour was inevitably left behind as it enclosed the descending thoracic aorta, thereby risking further manipulation. The chest was closed after inserting two chest drains.

The mass weighed 1130 grams and measured 175 x 150 x 80 mm. Histological examination revealed myxoid soft tissue sarcoma, with the differential of extraskeletal myxoid chondrosarcoma versus myxoid / round cell liposarcoma.

Serial post-operative chest X-rays confirmed adequate expansion of the lungs and recoil of mediastinal structures, both chest tubes were removed and patient had uneventful recovery and was later discharged on post-operative day 9 with complete clinical recovery.

Patient was last seen at a 4-week follow-up visit, he reported complete resolution of primary complains and total return to his baseline line activity. A follow-up CT scan was done at the follow-up visit which revealed mediastinal structures filling up normally and a fully expanded lung. Following that, patient is currently undergoing radiotherapy targeting the residual part of the mass located at the posterolateral border of the aortic arch and descending thoracic aorta towards left side. He is simultaneously followed at our clinic for surveillance in case of recurrence.

DISCUSSION:

Liposarcoma is the most common malignant tumour in adults, accounting for 15-20% of all soft tissue malignant tumours (3,4). They are commonly located to lower limbs and retroperitonium. However, liposarcomas located to the mediastinum are extremely rare and account for less than 0.75% of all mediastinal tumours (5). These tumours are often diseases of the adult with peak incidence at the age of 51 years (2), with no preference to either genders.

Considering the slow nature of this tumour and especially when located in the mediastinum, patients tend to remain asymptomatic and the underlying disease may be identified incidentally. Consecutively, when tumours become significantly enlarged; patients become symptomatic due to the pressure effect produced by the mass on intra-thoracic structures. Compression symptoms may include dyspnoea, tachypnea, chest pain, dysphagia (all of these were noted in our patient), wheezing, hoarseness, superior vena cava compression, arrhythmias and eventually heart failure. (1,5).

The predominant finding of mediastinal liposarcoma on imaging modalities is usually widened mediastinum, and may vary on computed tomography between fat-containing mass to solid mass, with attenuation levels similar to or higher than normal fat (3,6).

Pathologic evaluation of surgical specimen is the gold standard of diagnosis. Liposarcomas are circumscribed, encapsulated lobulated masses that are histopathologically classified into; well differentiated, dedifferentiated, myxoid, round cell and pleomorphic liposarcomas. Patients with well differentiated type, which is the most common (2), had better overall survival and less recurrence rate unlike the other types (2,5) (7). Hence, subtypes are closely related to prognosis.

Surgical resection is the gold standard treatment, and when en bloc resection is unattainable due to surrounding structures invasion, debulking surgery is the main stay of treatment as it will alleviate the symptoms. Various surgical approaches can be considered, depending on the mass location, size and structures invaded. Ranging between midline sternotomy, lateral thoracotomy or clamshell incision (1).

40% of mediastinal liposarcoma recur within 11.5 months from initial diagnosis (8), therefore a follow up period of at least 1 year is essential. Adjuvant chemotherapy and radiotherapy of Liposarcomas is of controversial effect (9), yet it is recommended especially in cases of positive margin resection. Myxoid type tumours are considered especially sensitive to chemotherapy and radiotherapy (6,10). All in all, due to recurrence and poor response to adjuvant therapy, liposarcomas are considered diseases with poor prognosis requiring close follow up with short life expectancy from time of diagnosis (8).

Post operatively, patient provided documents proving that he had undergone mass resection from the lower limb 19 years prior to the onset of his current disease. Histopathology reports proved that the tumour excised was myxoid liposarcoma grade I and was followed by adjuvant radiotherapy.

CONCLUSION

In conclusion, huge mediastinal liposarcomas are rare tumours treated surgically requiring close follow up and occasionally adjuvant therapy. The diagnosis and management of these tumours could present a real challenge.

The authors declare no conflict of interests.

REFERENCES:

1. Toda M, Izumi N, Tsukioka T, Komatsu H, Okada S, Hara K, et al. Huge mediastinal liposarcoma resected by clamshell thoracotomy: a case report. Surg Case Reports [Internet]. 2017;3(1). Available from: http://dx.doi.org/10.1186/s40792-017-0291-5
2. Hahn HP, Fletcher CDM. Primary mediastinal liposarcoma: Clinicopathologic analysis of 24 cases. Am J Surg Pathol. 2007;31(12):1868–74.
3. Greif JOEL, Or SMARM, Y OMESK, Er FKOVN, Inbar MOSHE. Prim ary liposarcom a of the m ediastinum. 1998;205–7.
4. Fan Z, Tian XF, Tang SX, Zhang YY, Pan JY, Wang S. Myxoid liposarcoma in the abdominal wall: A case report. Med (United States). 2014;93(28):e239.
5. Nguyen DC, Olatubosun O, Yu W, Loor G, Burt BM. Giant Mediastinal Liposarcoma: A Rare Yet Distinct Clinical Entity. Ann Thorac Surg [Internet]. 2018;106(3):e117–9. Available from: https://doi.org/10.1016/j.athoracsur.2018.03.018
6. Endara SA, Davalos GA, Vinueza AL, Montalvo N, Duran PG, Barzallo DE. Mediastinal Myxoid Liposarcoma with Intrapericardial Involvement and Large Pericardial Effusion. Heart Surg Forum. 2015;18(5):192.
7. Ortega P, Suster D, Falconieri G, Zambrano E, Moran CA, Morrison C, et al. Liposarcomas of the posterior mediastinum: Clinicopathologic study of 18 cases. Mod Pathol. 2015;28(5):721–31.
8. Mei X, Li M, Xia Y. A huge mediastinal, well-differentiated liposarcoma with heterogenous smooth muscle differentiation: a case report. Int J Clin Exp Pathol. 2019;12(7):2763–6.
9. Pui WC, Ling WHY, Najah M, Soon SY. Successful resection of a giant thoracic myxoid liposarcoma. Asian Cardiovasc Thorac Ann. 2018;26(5):410–2.
10. Punpale A, Pramesh CS, Jambhekar N, Mistry RC. Giant mediastinal liposarcoma: a case report. Ann Thorac Cardiovasc Surg. 2006;12(6):425–7.