Case Report

Enbloc resection of the largest thymic liposarcoma: A case report with literature review

Samer Alhames a, Mike Ghabally b, *

a Fellow at The French College of Vascular and Cardiothoracic Surgery; Chief of Thoracic Surgery Department at Saint Louis Hospital, Aleppo, Syria
b University of Aleppo, Faculty of Medicine, Department of Internal Medicine, Division of Cardiology, Aleppo, Syria

ARTICLE INFO
Keywords:
Case report
Liposarcoma
Mediastinal neoplasms
Thymus neoplasms

ABSTRACT

Introduction: Liposarcoma is the most common soft tissue tumor which is commonly found in the retroperitoneal region. This kind of tumor is usually well-differentiated with low to no potential to metastasize. Thymoliposarcomas are extremely rare tumors that are difficult to diagnose and differentiate from thymomas and other benign conditions.

Presentation of a case: This report presents a case of a 46-year-old male patient with dyspnea, generalized fatigue and non-specific chest pain caused by a giant anterior mediastinal mass. Computed tomography scan revealed a large mass in the anterior mediastinum. CT guided biopsy was positive for thymolipoma. The tumor was surgically resected. The histological analysis of the tumor revealed thymoliposarcoma.

Discussion: Thymoliposarcoma usually presents with non-specific symptoms. The mean age of the diagnosis is 55.8 years old with a slight predominance in males. The corner stone of the treatment remains surgical excision of the tumor while the role of adjuvant therapy is not well established.

1. Introduction

Liposarcoma is most frequently seen in the retroperitoneum; however, it can also develop in unusual sites [1,2]. Thymoliposarcoma is extremely rare type of thymus tumors that was first reported by Havlicek and Rosai in 1984, and till date, only 10 cases were reported. The diagnosis of thymoliposarcoma can be challenging, as it is hard to be distinguished from other thymus or anterior mediastinal tumors on imaging. Complete surgical resection is the standard treatment; the role of adjuvant therapy is not well established [4].

The presented case in this report is the largest thymoliposarcoma in the literature. The rarity of this tumor brings the importance to such cases to understand its presentation, clinical course and best available treatments. Furthermore, this review represents the first review in the literature of thymoliposarcoma. We believe that this review will help future doctors and researches to conduct further studies in this field.

2. Methods

In addition to the presented case, MEDLINE database for all published articles of thymoliposarcoma in the literature is also reviewed. Only liposarcoma involving the thymus gland were included. There was no restriction on language, country or publication date of the paper published. This work is reported in line with SCARE 2020 criteria [5].

3. Presentation of case

A 46-year-old male with no significant medical, surgical, psychosocial or family history presented to our institution with a yearlong complaint of fatigue, shortness of breath and non-specific chest pain (see Fig. 1). Part of the work up, the patient underwent chest x-ray, which revealed a mediastinal mass. A follow up computed tomography demonstrated 35 × 25 cm fatty mass occupying the anterior mediastinum causing collapse of the left lung and the upper lobe of the right lung (Fig. 1). CT guided biopsy was performed and was positive for thymolipoma. A thoracic surgeon with 20 years’ experience performed median sternotomy to resect the tumor, a large lobulated capsulated tumor mass was found, the mass was well circumscribed measuring 35 × 25 cm in size and occupying 95% of the left pleural cavity and 50% of the right pleural cavity causing near complete collapse of the left lung. The mass was resected in one piece and it weighs 5145 g (Fig. 2). The patient tolerated the procedure well with no complication and minimum
blood loss. On gross pathology, the mass showed white and yellow fatty areas of varying sizes and microscopically, the tumor exhibited a mixture of normal-appearing adipocytes mixed with atypical adipocytes. The atypical cells were hyperchromatic, pleomorphic and irregular. In addition, lobules of thymic tissue were embedded in the fatty component of the tumor. Atypical spindle cells were scattered in the adipose tissue and thymic cortex as well. A diagnosis of well-differentiated liposarcoma arising from the thymus was made. Patient was referred for radiation therapy. The patient has returned to his normal life and has been followed up clinically and radiologically since one year with no evidence of recurrence.

4. Discussion

Thymoliposarcoma is a very rare mesenchymal tumor that can reach a large size at diagnosis and remains asymptomatic [4]. The tumor usually expands and does not infiltrate into the neighboring structures; thus, most common symptoms are related to nearby structures compression by the tumor mass [1,3,4]. The tumor is usually encapsulated, lobulated with varying proportions of fibrosis and yellow adipose tissue [4]. The differential diagnosis of thymoliposarcoma is that of anterior mediastinal tumor including thymoma, thymolipoma and lymphoma among others [4,6].

Less than 200 cases of primary mediastinal liposarcoma have been reported so far; however, only 10 previous cases involved the thymus and were diagnosed as thymoliposarcomas (Table 1). This case represents the 11th in the literature and largest thymoliposarcoma till date. The mean age of the diagnosis is 55.8 years (range 31–77 years) with a slight predominance in males (male: female ratio 6:4). Most of the cases presented with non-specific symptoms (weight loss, fatigue, non-productive cough and dyspnea). The mainstay of treatment remains surgical resection of the tumor. The role of pre-surgical biopsy is not clear and surgical resection might be challenging given the location of the tumor. Close follow up is recommended. The role of adjuvant therapy (chemotherapy or radiotherapy) is not well documented in the literature with a lack of evidence of its potential role. However, the number of cases is very low and further studies are required in this field. Although thymoliposarcoma in general carry a favorable prognosis, recurrence was reported in three patients in the literature [3,4,8] and distal metastasis were documented in three patients with thymoliposarcoma (Vertebral [3], Nodular [7] and costolateral lung micronudules [2]).

Fig. 1. (a) Coronal and (b) sagittal Computed tomography demonstrated an anterior mediastinal tumor with irregular soft tissue density.

Fig. 2. The tumor measuring approximately 35 × 25 cm in size, 5.14 kg in weight.

Declaration of competing interest

None.

Funding

None.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.
Table 1
Thymoliposarcoma cases in the literature (UA: unavailable data; unfortunately we could not provide the full data of the third case).

| Case | Age  | Sex | Clinical Presentation | Size (cm) | Pathology | Treatment | Recurrence | Metastases | Follow up  |
|------|------|-----|-----------------------|-----------|-----------|-----------|------------|------------|------------|
| [3]  | 39   | F   | Asymptomatic (pressure sensation) | 13 × 9.5 × 5 (650 gr.) | Well differentiated to pleomorphic | Surgical Resection | 25 years after the initial surgery (surgery + radiotherapy) | 4 years after recurrence (radiotherapy) | 32 years   |
| [7]  | 1    | M   | Weight loss, malaise, night sweats, shoulder pain, chest infections | 450 gr. | Thymosarcoma with liposarcomatous differentiated | Surgical Resection | 21 months of initial treatment (treated with radiotherapy) | Nodular Metastasis | 21 months   |
| [8]  | UA   | UA  | UA                    | UA        | UA        | UA        | UA         | UA         | UA         |
| [9]  | 72   | F   | Asymptomatic           | No Data   | Well-Differentiated | Surgical Resection | –          | –          | 6 months   |
| [1]  | 77   | F   | Asymptomatic           | 9 × 8.5 × 5.4 cm | Well-differentiated + adjuvant radiotherapy | Surgical Resection | –          | –          | 29 months  |
| [5]  | 70   | F   | Non-productive cough   | 12 cm     | Lipoblastic with undifferentiated myxoid pattern | Surgical Resection | –          | –          | No Data    |
| [4]  | 36   | M   | Asymptomatic           | 12.5 × 11 × 5.3 cm | Well-differentiated | Surgical Resection | 4 years after surgery (13.2 × 7.5 × 4.4 cm, surgery and radiotherapy) | –          | 10 months after recurrence |
| [10] | 55   | M   | Non-specific chest pain| 30 × 12 × 12 (5120 gr.) | Pleomorphic | Surgical with Radiotherapy | –          | –          | 3 months   |
| [2]  | 63   | M   | Dyspnea, Weight loss   | 11 cm     | Dedifferentiated | chemotherapy (doxorubicin + ifosfamide) | –          | –          | Costolateral lung microneubules | No Data    |
| [1]  | 63   | M   | Asymptomatic           | 8.3 × 5.3 × 8.2 cm | Dedifferentiated | Surgical Resection | –          | –          | 1 year     |
| [1]  | 46   | M   | Fatigue, Dyspnea, Non-specific chest pain | 35 × 25 (5145 gr.) | Well-differentiated | Surgical Resection | –          | –          | 6 months   |

Provenance and peer review
Not commissioned, externally peer reviewed.

Guarantor
The first author is the guarantor of this article.

Acknowledgements
To Dr. Khaldoun Almhanna, thank you for your assistance in lingustic proofreading of this article; it is sincerely appreciated.

Appendix A. Supplementary data
Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2020.09.048.

References
[1] Y. Sekine, K. Hamaguchi, Y. Miyahara, Thymus-related liposarcoma: report of a case and review of the literature, Surg. Today 26 (1996) 203–207, https://doi.org/10.1007/BF00311509. PMID: 8845616, https://link.springer.com/article/10.1007/BF00311509.
[2] A. Mansuet-Lupo, F. Lococo, F. Larrousserie, M. Alifano, R. Saliceti, Dedifferentiated primary mediastinal liposarcoma mimicking a thymic tumor, Pathologica 109 (4) (2017) 401–404. PMID: 29449733. URL: https://moh-it.pure.elsevier.com/en/publications/dedifferentiated-primary-mediastinal-liposarcoma-mimicking-a-thym.
[3] F. Havlicek, J. Rosai, A sarcoma of thymic stroma with features of liposarcoma, Am. J. Clin. Pathol. 82 (1984) 217–224, https://doi.org/10.1093/ajcp/82.2.217. PMID: 6465086, https://academic.oup.com/ajcp/article-abstract/82/2/217/1860277?redirectedFrom=fulltext.
[4] M.T. Sung, S.F. Ko Sf, M.J. Hsieh, W.J. Chen, H.Y. Huang, Thymoliposarcoma, Ann. Thorac. Surg. 76 (6) (2003) 2082–2085, https://doi.org/10.1016/s0003-4975(03)03100-6, PMID: 14667651, https://www.annalsofThoracicSurgery.org/article/30003-4975/03100-6/fulltext.
[5] A.A. Agba, M.R. Borrelli, R. Farwana, K. Koshy, A. Fowler, D.P. Orgill, For the SCARE Group, The SCARE 2018 statement: updating consensus surgical Case Report (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.
[6] D.S. Klimstra, C.A. Moran, G. Perino, M.N. Koss, J. Rosai, Liposarcoma of the anterior mediastinum and thymus. A clinicopathologic study of 28 cases, Am. J. Surg. Pathol. 20 (1996) 1025–1031, https://doi.org/10.1097/00000478-199605000-00006. PMID: 8793476, https://journal.sci-hub.DO/abstract/1996/05000/AClinicopathologicStudyOf28Cases.aspx.
[7] H. Jones, M. Yaman, C.R. Penn, T. Claake, Primary stromal sarcoma of the thymus with areas of liposarcoma, Histopathology 23 (1993) 543–548. PMID: 8127637.
[8] E.G. Cristallini, S. Nati, S. Ascani, G.B. Bolis, Concurrent thymic stroma sarcoma (thymoliposarcoma) and breast carcinoma. Report of a case, Pathologica 85 (1995) 543–548. PMID: 8127657.
[9] S.J. Howling, J.D. Flint, N.L. Muller Thymoliposarcoma, CT and pathologic findings, Clin. Radiol. 54 (1999) 54, https://doi.org/10.1111/j.1365-2559.1999.tb01187.x. PMID: 8365716, https://onlinelibrary.wiley.com/doi/10.1111/j.1365-2559.1999.tb01187.x.
[10] A. Sivaraman, G.K. Jaikaran, R.V. Suresh, R.K. Sashank, Giant Thymoliposarcoma (pressure sensation) and breast carcinoma. Report of a case, Pathologica 85 (1995) 31588117.