CASE REPORT

Primary Synovial Sarcoma Presenting as a Huge Mass: A Report of a Rare Case and Review of Literature

Seyed Hossein Fattahi Masoom, MD[1]; Amir Hossein Jafarian, MD[2]; Alireza Sharifian Attar, MD[3]; Fatemeh Sadat Abtahi Mehrjardi, MD[4]; Mehrdad Fakhlaei, MD[4]; Leila.S Arani, MD[5]; Sharifeh Kamalimotlagh, MD[5]; Asieh Sadat Fattahi, MD[4]

Research performed at Endoscopic and Minimally Invasive Surgery Research Center, Department of Thoracic Surgery, Mashhad University of Medical Sciences, Mashhad, Iran

Received: 23 September 2019 *Accepted: 13 October 2019*

Abstract

Primary synovial sarcoma of mediastinum is very rare among soft tissue sarcomas. Only a few cases have been reported in the literatures. The best treatment is still unclear, but, surgical resection is the main therapy. In this article we report a case of a 20*20 cm (2000gr) primary giant mediastinal synovial sarcoma in a 42 year-old man. We performed radical excision of the tumor and the metastasis.

Level of evidence: V

Keywords: Chemotherapy, Mediastinal mass, Soft tissue sarcoma, Synovial sarcoma

Introduction

Soft tissue sarcomas (STS) are rare tumors that present less than 1% of malignant neoplasms among all cancers in adults. Carcinomas are the most malignant thoracic tumors. The proportion of STS is even much less; probably less than 0.01% of all malignant thoracic neoplasms (1).

Synovial sarcomas usually occur in the extremities. This kind of sarcoma is observed in young adults. Primary synovial sarcoma of mediastinum is rare. Only a few cases have been reported in the literatures (2). The best treatment specialty in unresectable tumors is unclear, but, complete resection has been the only therapy associated with long term survival.

Here in, we report a case of 20×20 cm (2000 gr) primary giant mediastinal synovial sarcoma with a 2×2 cm lung nodule (metastasis) in a 42 year-old man .We performed complete resection of the tumor or and the metastasis.

Case presentation

A 42-year-old man was referred to the Department of Thoracic Surgery, Ghaem Teaching Hospital, Mashhad, Iran, with a six-month history of exertional dyspnea and feeling of fullness in his chest and chronic cough. Physical examination did not reveal anything significant. Just he had decreased pulmonary sounds in the right side. His blood counts at presentation revealed hemoglobin level of 14.6 gm/dL, total leukocyte counts of 5600 cells/mm3, and platelet counts of 228,000/mm3. His serum electrolytes, renal function tests, and liver function tests were within normal range. The chest X-ray showed a large mass in right thoracic cavity, adjacent to the mediastinum [Figure 1A]. Computed tomography (CT) scan revealed a 20 cm mediastinal mass and a 2cm mediastinal nodule in right thoracic cavity [Figure 1B]. CT guided biopsy was done. It was reported as fibrous tissue with hyalinization. After induction of anesthesia with midazolam, sufentanil,propofol and atracurium patient intubated with 39 french left DLT. The patient underwent Right thoracotomy and excision of the tumor and metastasis was performed. The tumor measured about 20

Corresponding Author: Asieh Sadat Fattahi and Fatemeh Sadat Abtahi Mehrjardi, Endoscopic and Minimally Invasive Surgery Research Center, Mashhad University of Medical Sciences, Mashhad, Iran
Email: emis@mums.ac.ir

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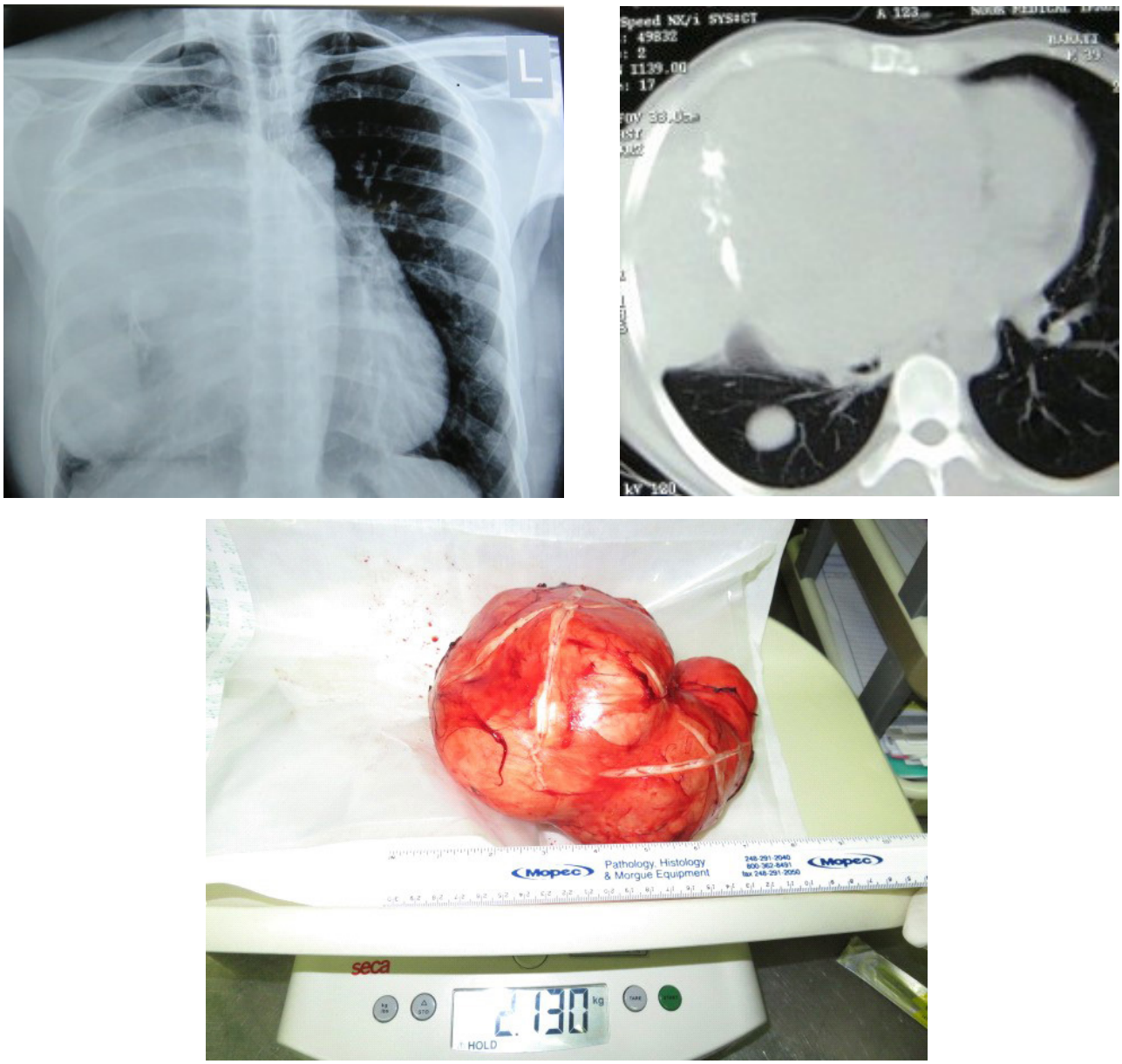

Figure 1. (A) Chest X-ray showed a large mass in Right thoracic cavity. (B) CT scan of the chest revealed a 20cm mediastinal mass and metastasis. (C) Tumor removed with thoracotomy.

by 20 by15 cm [Figure 1C]. histologically the tumor was composed of spindle cell and the Inferior lobe nodule was metastatic spindle cell sarcoma. Immunohistochemistry demonstrated strong positivity for bcl_2,CD99. Mesothelial markers and S100 were negative. Histopathological and immunohistochemical findings revealed the diagnosis of a primary biphasic synovial sarcoma of the mediastinum [Figure 2]. Radical resection of the tumor and metastasis was followed by chemotherapy.

Discussion

Synovial sarcoma is a tumor arising from the mesenchymal tissue. Most of the time, it is in the extremities of adolescents and young adults (3), while a primary synovial sarcoma in the mediastinum is very rare (4).

Due to rarity of cases and lack of data the best therapeutic strategy is not clear yet. Complete surgical resection is the treatment goal (5). Bart reviewed 3149 cases of soft tissue synovial sarcomas among which almost 1.4% were mediastinal (6).

In our case the tumor was 20×20cm and 2Kg which was completely resected along with a 2×2 cm nodule in the inferior lobe of lung without any mediastinal residue.

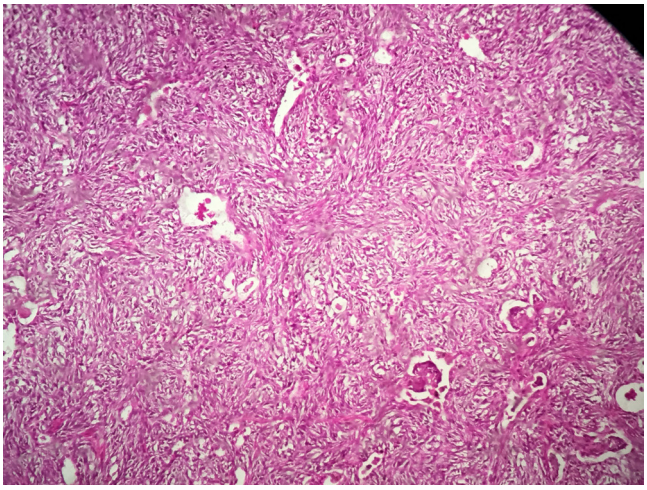

Figure 2. Histopathology Examination that shows spindle cell.

Synovial sarcomas have poor prognosis as they are highly aggressive. The tumor size is one of the important prognostic indicators. Patients with localized synovial sarcomas with the size less than 5cm have shown a 10-year survival of %88, but, with the size of 5-10 cm the 10-year survival rate drops to 38 %, and with a dimension greater than 10 cm it will be only 8% (7). Since a primary synovial sarcoma of the mediastinum is extremely rare, the best treatment is still unclear. Complete surgical resection is the mainstay of therapeutic strategy for synovial sarcomas arising in the mediastinum and the possibility of complete excision has an important role in determining the survival.

In comparison with resectable synovial sarcoma of the extremities, patients with resectable mediastinal disease seem to have worse prognoses. Unfortunately most reported cases of mediastinal synovial sarcomas with radical resection have shown local or distant recurrences. This suggests that adjuvant therapy may be recommended even with complete surgery. In our case we performed complete resection and metastasectomy followed with chemotherapy.

Conflict of Interests: The author(s) declare no potential conflicts of interests with respect to the research, authorship, and/or publication of this article.

Seyed Hossein Fattahi Masoom MD[1]
Amir Hossein Jafarian MD[2]
Alireza Sharifian Attar MD[3]
Fatemeh Sadat Abtahi Mehrjardi MD[4]
Mehrdad Fakhlaei MD[4]
Leila.S Arani MD[5]
Sharifeh Kamalimotlagh MD[5]
Asieh Sadat Fattahi MD[4]
1 Lung Diseases Research Center, Mashhad University of Medical Sciences, Mashhad, Iran
2 Mashhad University of Medical Sciences, Mashhad, Iran
3 Department of Anesthesia, Faculty of Medicine, Mashhad University of Medical Sciences,Mashhad,Iran
4 Endoscopic and Minimally Invasive Surgery Research Center, Mashhad University of Medical Sciences, Mashhad, Iran
5 University of Pennsylvania, Philadelphia, PA, USA

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