World Journal of Surgery and Surgical Research

Case Report
Published: 10 Jun, 2020

A Rare Case of Metastatic Low-Grade Fibromyxoid Sarcoma of the Lung

*Madonna E Lee[1], Abu Rogers[2], Hans Iwenofu[3], David Liebner[4], Daniel Mansour[1], Ronald Figura[1], Alicia Perkins[1], Robert E Merritt[1] and Desmond M D'Souza[1]**

[1]Division of Thoracic Surgery, The Ohio State University Wexner Medical Center, USA

[2]The Ohio State University, USA

[3]Department of Pathology, The Ohio State University Wexner Medical Center, USA

[4]Department of Medical Oncology, The Ohio State University Wexner Medical Center, USA

Abstract

Evans tumor, also known as Low-Grade Fibromyxoid Sarcoma (LGFMS) is rare form of sarcoma, occurring in the deep soft tissues of mostly young adult males. We describe a 36-year old man with pulmonary LGFMS that originated from possible metastasis of a primary leg lesion. The patient had initially been misdiagnosed with neurofibroma in their right leg. A right upper lobectomy and mediastinal dissection were performed to remove the tumor. The tumor was completely resected, and there has been no evidence of recurrence. LGFMS should be included as a differential for large, benign-appearing lung masses in young adults, as complete surgical resection can be effective.

Keywords: Low grade fibromyxoid sarcoma; Fibromyxomas; Evans tumor; Intrathoracic

Background

In 1987, Evans described Low-Grade Fibromyxoid Sarcoma (LGFMS) a rare soft tissue sarcoma occurring in extremities with a benign spindle cell appearance but malignant potential [1]. It is usually diagnosed in young adult males [2]. Less than 200 cases have been reported. Even rarer is the occurrence of intrathoracic LGFMS [3,4]. This case describes a metastatic pulmonary LGFMS undergoing resection in the setting of Partial Anomalous Pulmonary Venous Return (PAPVR).

Case Presentation

A 36 year-old man initially went to an emergency department with chest pain and dyspnea. On Chest X-Ray (CXR), a large right lung mass was identified. CT (Computer Tomography) Chest confirmed a Right Upper Lobe (RUL) mass measuring 11.5 cm × 10.2 cm × 10.2 cm (Figure 1). A CT-guided biopsy was performed, and it was consistent with LGFMS. A PET (Positive emission tomography)/CT demonstrated a hypermetabolic heterogeneous mass in the RUL abutting the mediastinum and hilum, consistent with neoplasm (SUV 3.8). At the age of nine, the patient was diagnosed with neurofibroma based on a small mass on his right leg. His physical exam and pulmonary function tests were unremarkable.

Operative Course

The patient was taken to the operating room for a RUL lobectomy and mediastinal dissection.

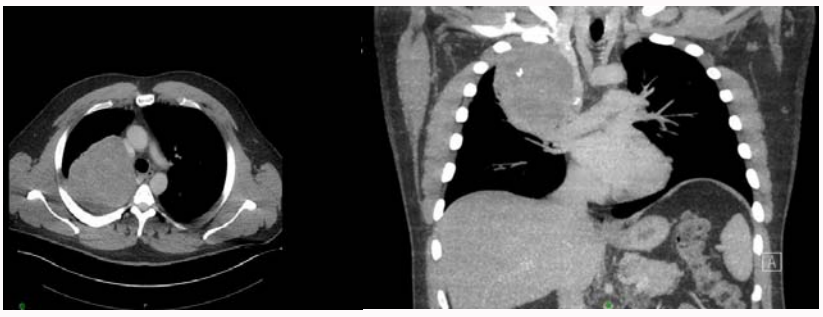

Figure 1: CT Chest–axial and coronal images.

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Correspondence:
Desmond M D'Souza, Department of Cardiothoracic Surgery, Division of Thoracic Surgery, The Ohio State University Wexner Medical Center, 410 West 10th Avenue, Columbus, OH 43210, USA,
E-mail: desmond.d'souza@osumc.edu

Received Date: *11 May 2020*
Accepted Date: *05 Jun 2020*
Published Date: *10 Jun 2020*

Citation:
Lee ME, Rogers A, Iwenofu H, Liebner D, Mansour D, Figura R, et al. A Rare Case of Metastatic Low-Grade Fibromyxoid Sarcoma of the Lung. World J Surg Surgical Res. 2020; 3: 1231.

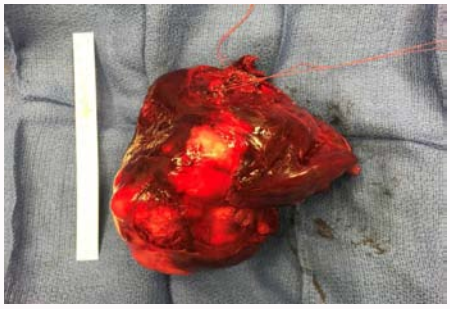

Figure 2: Intraoperative Specimen- 11.5 cm × 9.0 cm × 8.8 cm.

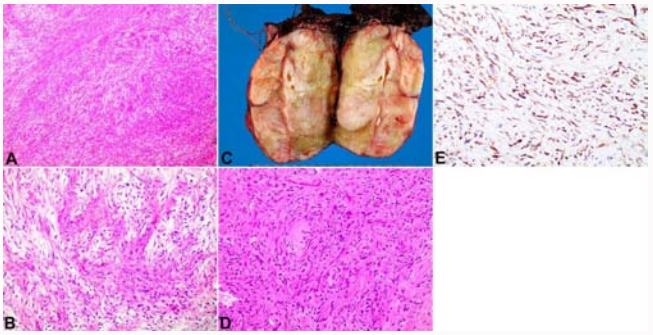

Figure 3: Histology Slides:
Figure 3A, 3B: (H&E and E, Mag x200, x400): Original biopsy material of the right leg mass showing alternating myxoid and fibrous areas with whorling growth pattern and perivascular accentuation.
Figure 3C: Cut surface of the resected mass showing a well circumscribed tumor with fibrous white-tan surface and focal myxoid areas.
Figure 3D: (Mag x200): Section showing a banal spindle cell neoplasm exhibiting a focal whorling growth pattern and marked interstitial collagen; bearing a certain semblance to the tumor from the right leg. There are no features of cellular anaplasia.
Figure 3E: (Mag x200): Immunohistochemistry for MUC4 showing strong and diffuse reactivity in the lesional cells, consistent with LGFMS.

The RUL was replaced by tumor (Figure 2). During dissection, the patient was found to have an atretic truncal right pulmonary artery branch and PAPVR draining directly into the superior vena cava. The absence of a superior pulmonary vein draining into the left atrium confirmed presence of PAPVR. The patient received an R0 resection.

Gross examination showed a circumscribed fleshy white mass with myxoid areas (11.5 cm × 9.0 cm × 8.8 cm) (Figure 3C). Histologic sections showed banal spindle cell proliferation, interstitial collagen, and swirling growth pattern. The cells were strongly positive for Mucin 4 (MUC4), focally positive for Epithelial Membrane Antigen (EMA) and negative for Smooth Muscle Actin (SMA), S-100, desmin and Signal Transducer and Activator of Transcription 6 (STAT-6) (Figure 3d, 3e). The overall features were prototypic for LGFMS.

Prompted by this histology, the previous right leg specimen diagnosed as neurofibroma was requested for review. The Hematoxylin and Eosin stained sections show a low-grade spindled neoplasm with alternating spindled and myxoid areas with whorling growth pattern (Figure 3a, 3b). The lesion on the leg was consistent with the primary lesion; whereas the lung mass was consistent with a metastatic lesion. The patient had an uncomplicated post-operative course and was discharged home on day 4. He is under surveillance with annual CT imaging.

Discussion

LGFMS tumors have potential for late recurrences and metastases, particularly to the lungs [4], as demonstrated in this case. Upon re-review of the original pathology specimen, it appears the leg lesion was initially misdiagnosed as a neurofibroma. Thus, this pulmonary LGFMS is a late, metastatic lesion, which is a classic presentation of a typical low-grade biology and metastases presenting after several decades [5]. Although pulmonary fibromyxomas are rare, LGFMS should be included as a differential for benign-appearing large lung masses in young adults, since complete surgical resection can provide a disease-free benefit [4].

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