Primary Myxoma of the Lung

Akciğerin Primer Miksoması

Ulas Kumbasar [1], Serkan Uysal[1], Nazlı Eylem Alkanat [2], Kemal Kosemehmetoglu[2], Riza Dogan[1]

[1] Hacettepe University School of Medicine, Department of Thoracic Surgery, Ankara, Turkey
[2] Hacettepe University School of Medicine, Department of Pathology, Ankara, Turkey

ABSTRACT

Primary myxoma of the lung is extremely rare. In this report, we present a 36-year-old man who presented to our clinic with cough and hemoptysis. His chest CT revealed a 15 x 14 mm mass with mildly spiculated borders in the right upper lobe. A wedge resection was performed under video-assisted thoracoscopy. Histologically, the tumor was diagnosed as primary pulmonary myxoma.

Keywords: Primary myxoma, lung, surgery, VATS

Received: 08.01.2020 **Accepted:** 10.01.2020

ÖZET

Akciğerin primer miksoması oldukça nadirdir. Olgu sunumumuzda, kliniğimize öksürük ve hemoptizi şikayeti ile başvuran 36 yaşında bir erkek hastayı sunuyoruz. Toraks bilgisayarlı tomografisinde sağ üst lobda hafif spiküler sınırları olan 15 x 14 mm'lik bir lezyon tespit edildi. Video yardımlı torakoskopik yöntemle wedge rezeksiyon yapıldı. Histolojik olarak tümör, primer pulmoner miksoma olarak teşhis edildi.

Anahtar Sözcükler: Primer miksoma, akciğer, cerrahi, VATS

Geliş Tarihi: 01.08.2020 **Kabul Tarihi:** 01.10.2020

ORCID IDs. U.K. 0000-0003-0616-1326, S.U. 0000-0002-1152-2854,N.E.A. 0000-0003-3506-3856, K.K. 0000-0002-7747-0460, R.D. 0000-0003-4845-3044

Address for Correspondence / Yazışma Adresi: Serkan Uysal, MD Hacettepe University School of Medicine, Department of Thoracic Surgery Sıhhiye, Ankara, Turkey E-mail: serkan.uysal@hacettepe.edu.tr

doi:http://dx.doi.org/10.12996/gmj.2021.74

INTRODUCTION

Myxoma is a mesenchymal tumor that frequently stems from the left atrium of the heart. Primary mesenchymal tumors originating from the lung are rare. Furthermore, primary pulmonary myxoma is extremely rare and has only been documented in the literature as sporadic case reports. Most patients are asymptomatic and diagnosed incidentaly. It's diagnosis is hard to be made on needle biopsy or cytology. Because of this resection is essential. (1-3). Herein we present a primary myxoma of the lung in a middle-aged man.

CASE REPORT

A 36-year-old man presented with a history of cough and hemoptysis of 2 weeks duration. He was previously asymptomatic and has no history of malignancy. His physical examination was normal. The thorax computed tomography revealed a 15 x 14 mm mass with mildly spiculated borders in the right upper lobe and it's density is 60 HU(Hounsfield unit) (Fig. 1). Transthoracic echocardiography demonstrated no evidence of any cardiac mass such as myxoma. The management step is discussed with the patient and he refused to have a biopsy and wanted removal of the lesion. Based on the computed tomography images, this lesion is mostly considered as a benign lesion. Furthermore there were no mediastinal lymph node involvement at the thorax computed tomography. Thus, we did not consider positron emission tomography (PET) imaging. The patient was discussed in the multidisciplinary meeting and surgical removal of the lesion was recommended. A wedge resection of the mass was performed under video-assisted thoracoscopy. Intraoperative frozen section analysis revealed a mesenchymal tumor. Due to the benign result of the frozen section we decided to do a wedge resection and follow up with permanent histologic result rather than performing lobectomy. Therefore, limited resection with sufficient margins was considered to be satisfactory for local control of the tumor. The postoperative course of the patient was uneventful. Macroscopically tumor was 1,7x1,5x1,1 cm, well-demarcated and had gray-white myxoid appearance. Histological examination revealed a myxoid tumor with elongated spindle cells in a basophilic loose stroma with thin-walled vascular structure, accompanying many inflammatory cells such as plasma cells and lymphocytes. Spindle cells had minimal to moderate pleomorphism, no mitosis was seen. Myxoid matrix was positive with Alcian blue. Immunhistochemically tumor was all negative for pancytokeratin (AE1/AE3), EMA, SMA, desmin, calretinin, SALL4, DOG1, S100, CD34, ALK, DOG1, p63 and CD30. There was no EWSR1 gene rearrangement by fluorescent in situ hybridization (FISH) (Fig. 2). Given the all morphological immunophenotypical and molecular findings, tumor was diagnosed as pulmonary myxoma – mainly a diagnosis of exclusion. Based on the histopathologic diagnosis, a follow-up strategy was planned in order to identify any potential recurrence or metastasis. The patient has been followed for one year without reccurence and metastasis.

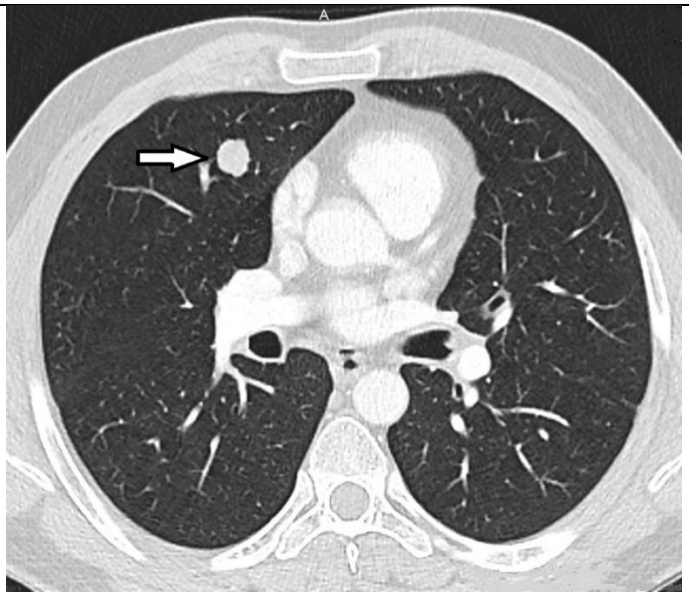

Figure 1. CT revealed a homogenous mass with mildly spiculated borders in the right upper lobe (white arrow).

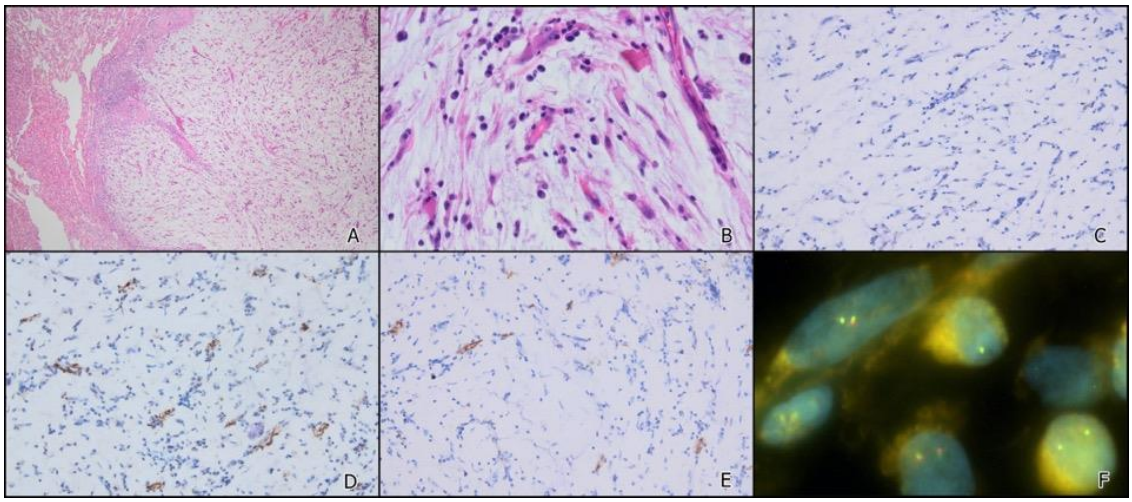

Figure 2. (A) A well-demarcated hypocellular myxoid tumor (H&E×40). (B) Scattered bland spindle cells in a basophilic loose stroma are accompanied by inflammatory cells (H&Ex400). Immunohistochemically, no expression for S100 (C) , SMA (D) and CD34 (E). (F) EWSR1 gene rearrangement was negative by FISH.

DISCUSSION

Myxomas originate from multipotent mesenchymal cells which has the potential of neural and endothelial differentiation. Histologically, they are composed of elongated and scattered cells within a mucopolysaccharide stroma. They are the most common primary cardiac neoplasm, especially originate from the left atrial wall (4). Primary mesenchymal tumors rarely occur in the lungs. Among them, primary pulmonary myxoma is unique and only five cases have been reported in the literature in English so far (1-3, 5).

Most patients were reported to be asymptomatic or had non-specific symptoms as our case. In 4 of 5 previously reported cases, the tumor was reported in the sixth or seventh decades of life. In one case it was described in a 26-year-old male which is similar to our case. The differential diagnosis of primary pulmonary myxoma includes benign mesenchymal tumors of the lung such as hamartoma, angiomyolipoma, and pulmonary lipoma. The differentiation of these clinical diagnoses would not be challenging in resected specimens (2). However; diagnosis of a myxoid mesenchymal tumor in lung is extremely challenging. Recently described primary pulmonary myxoid sarcoma was the initial differential diagnosis of this tumor. Tumor showed no EWSR1 rearrangement by FISH along with desmin and EMA negativity that ruled out this entity (6). Epithelial tumors were excluded with EMA and keratin negativity. Another possible differential diagnosis was inflammatory myofibroblastic tumor, characterized with myofibroblastic spindle cells without atypia, prominent inflammatory cells on the background, and SMA and ALK expression. Extensive myxoid change was described in inflammatory myofibroblastic tumors; however, both SMA and ALK were negative in our case disabling the diagnosis of inflammatory myofibroblastic tumor. Vascular tumors, especially epithelioid hemangioendothelioma – mainly designated by myxocolllagenous background– were excluded with CD31 and CD34 negativity. S100, SMA and p63 for myoepithelial neoplasia were all negative. Diagnosis of a possible intimal sarcoma was opposed by a long clinical history and absence of relation with a vessel, as well as lack of atypical nuclei. Finally, the tumor was diagnosed as pulmonary myxoma based on morphological (elongated spindle cells, loose myxoid stroma, no mitosis) and immunophenotypical features. The case was also investigated for the presence of cardiac myxoma, because pulmonary embolism is not an uncommon complication of cardiac myxoma (7).

Although there are very limited number of cases to recommend the appropriate surgical treatment of these tumors, based on the histopathologic resemblance of these tumors to conventional cardiac myxomas, surgical resection of the lesion with sufficient margins considered to be the best treatment option in patients with primary pulmonary myxoma. Even though metastases or recurrences have been reported in cardiac myxomas, no recurrences or metastasis have been documented for primary pulmonary myxomas yet (8). However, long-term follow-up is mandatory for early detection of potential local recurrences and/or distant metastases.

CONCLUSION

In short, we have described an unusual case of primary pulmonary myxoma in a middle-aged man. We think that complete surgical resection would be curative in this patient. Nevertheless, we planned a long-term follow-up strategy in order to identify the long-term consequences of this rare entity.

Conflict of interest

No conflict of interest was declared by the authors.

REFERENCES

1.Chen CC, Chang JM. Primary pulmonary myxoma. QJM. 2016;109(8):549-50.
2.Kalhor N, Marom EM, Moran CA. Primary myxoma of the lung. Ann Diagn Pathol. 2010;14(3):178-81.
3.Matsuoka H, Takata Y, Maeda S. Primary pulmonary myxoma. Ann Thorac Surg. 2005;79(1):329-30.
4.Pucci A, Gagliardotto P, Zanini C, Pansini S, di Summa M, Mollo F. Histopathologic and clinical characterization of cardiac myxoma: review of 53 cases from a single institution. Am Heart J. 2000;140(1):134-8.
5.Abcarian H, Lee HT, Barker WL. Primary endobronchial myxoma. Ann Thorac Surg. 1973;15(3):287-90.
6.Thway K, Nicholson AG, Lawson K, Gonzalez D, Rice A, Balzer B, et al. Primary pulmonary myxoid sarcoma with EWSR1-CREB1 fusion: a new tumor entity. Am J Surg Pathol. 2011;35(11):1722-32.
7.Thyagarajan B, Kumar MP, Patel S, Agrawal A. Extracardiac manifestations of atrial myxomas. J Saudi Heart Assoc. 2017;29(1):37-43.
8.Kaynak K, Besirli K, Arslan C, Ozguroglu M, Oz B. Metastatic cardiac myxoma. Ann Thorac Surg. 2001;72(2):623-5.