Özgün Araştırma / Original Research

PRİMER AKCİĞER MALİGN MEZENKİMAL TÜMÖRLERİ

PRIMARY LUNG MALIGN MESENCHYMAL TUMORS

Murat Kavas[1], Cansel Atinkaya Baytemir[1], Ayşe Ersev[1], Akın Öztürk[1], Onur Derdiyok[1], Selma Aydoğan Eroğlu[1], Sümeyye Alparslan Bekir[1], İrfan Yalçınkaya[1]

[1]Süreyyapaşa Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim Araştırma Hastanesi, İstanbul, Türkiye

ÖZET

Primer mezenkimal tümörler, akciğerin oldukça nadir görülen tümörleridir. Mezenkimal tümörler tüm kanserlerin yaklaşık %1 ni oluşturur ve en sık yerleşim yerleri extremitelerdir (%59). Nadir görülmeleri nedeniyle tedavileri ve prognozları sınırlıdır. Bu çalışmada primer akciğer mezenkimal tümörlü üç olgu literatür ışığında sunulmuştur.

Anahtar Kelimeler: Mezenkimal akciğer tümörleri, Akciğer kanseri, KHDAK, Rezeksiyon sonrası, Radyoterapi

ABSTRACT

Primary mesenchymal tumors are extremely rare tumors of the lung. Mesenchymal tumors constitute about 1% of all cancers and the most common sites are the extremities (59%). Their treatment and prognosis are limited because of their rare occurrence. In this study, three cases of primary lung mesenchymal tumor are presented in the light of the literature.

Keywords: Mesenchymal lung tumors, Lung cancer, NSCLC, Post-resection, Radiotherapy

INTRODUCTION

Although several methods have been applied to classification of mesenchymal tumors, there is no definitive classification method. However mesenchymal tumors are classified according to their origin tissue in general. Tumors are extremely rare and involvement of tissues of multiple histological structures is a major challenge in the classification of mesenchymal tumors. Mesenchymal tumors can occur anywhere in the body. Locality and growth rates depend largely on the origin of the tumor [1, 2]. Although approximately 1% of mesenchymal tumors are malignant, histological diagnosis by imaging methods is not easy [3]. Intrathoracic malignant mesenchymal tumors may originate from the lung, thoracic wall, mediastinum or other tissues [2]. The clinic is usually asymptomatic. Symptoms such as chest pain, shortness of breath, and chronic cough may be associated with the site and size of the lesion in symptomatic cases. Though transthoracic needle aspiration biopsy and thoracoscopic biopsy are helpful in diagnosis, the exact diagnosis of the disease is usually obtained by surgical intervention. In this study, three patients with definite diagnosis of postoperative primary malign mesenchymal lung tumors were presented with clinical, radiological and histopathological features.

METHODS

Case 1

A 28-year-old male patient presented to our clinic for complaints of breathlessness and chest pain. There was no obvious physical examination. He had had a pneumothorax in his left hemithorax four years; also his three year-old son died of leukemia. Pneumothorax was treated with tube thoracostomy, there was no sign of any another disease in the chest tomography. On admission,

Corresponding Author / Sorumlu Yazar: Murat Kavas, e-mail: muratkavas@gmail.com

posterior-anterior (PA) radiograph showed a mass density of about eight cm size (Figure 1). Thorax ultrasonography showed lesion compatible with cystic lesion. Positron emission tomography (PET-CT) showed a 89x59 mm cystic lesion in the left upper lobe (SUV-max 2.6). Laboratory parameters were within normal limits. Pulmonary function tests were within normal limits (FVC: 3.05 lt, %98, FEV1: 2.7 lt, 95%). Endobronchial lesion was not detected in fiberoptic bronchoscope (FOB). Sputum, bronchoscopic lavage cytology showed no malignancy. Informed consent was obtained. In June 2015, a thin-walled cystic lesion with minimal adhesiveness to the chest wall was observed to be originated from lingula at thoracotomy. Frozen section was reported as benign. Therefore, the wedge resection of the lingula was performed and the thin-walled cystic lesion filled with necrotic material was excised totally. After the definitive pathology was reported as a primary malignant mesenchymal tumor of the lung (Figure 1.b), upper lobectomy and MLND was performed with rethoracotomy on September 2015. Postoperative chemotherapy was administered. No pathology was detected in the patient's 20-month follow-up.

Case 2

A 28-year-old woman was admitted to our clinic with a complaint of left shoulder pain. For 11 years there was a history of active smoking. Postero-anterior (PA) chest X-ray showed an increase in density of about three centimeters (cm) in the left lung (Figure 2.a). Thorax CT showed a soft tissue lesion of 33x42 mm in the left lower lobe of the lung (Figure 2.b). No endobronchial lesion was found in FOB. Diagnosis was not obtained with bronchoscopic lavage and transthoracic cutter needle biopsy. Thorax USG showed lesion compatible with cystic lesion. Laboratory parameters and respiratory function tests were within normal limits (FVC: 2.98 lt, 90%, FEV1: 2.65 lt, 85%). Informed consent was obtained. Left lateral thoracotomy was performed in 2015. The frozen necrotic material, thin-walled and rounded, was reported to be benign. It was observed that the lesion was densely adhesive to the pericardium and diaphragm and also to the anterior segment of the lower lobe. The wedge resection was performed to the lower lobe. A biopsy was taken from areas where the pericardial and diaphragm were also adhered. Frozen section was reported as benign. Therefore, the lesion was totally excised. Postoperative pathology was identified as an unidentifiable high-grade primary malignant mesenchymal tumor of the lung (Figure 2.c). After postoperative pathology report PET-CT and cranial MR were performed. PET-CT and Cranial MR reported no metastasis. Left lower lobectomy and MLND was performed with re-thoracotomy on March 2015. She received postoperative chemotherapy and radiotherapy. Patient follow-up was 46 months and no pathology was detected.

Case 3

A 44-year-old male patient was admitted to our clinic with a complaint of chest pain and shortness of breath with active smoking for eighteen years. His brother had died of leukemia. In the thoracic CT, an increase in smooth soft tissue density of approximately 10 mm in diameter was observed in the anterior segment of the left upper lobe of the lung (Fig. 3.a). In PET-CT, the nodule in the upper lobe of the left lung was SUVmax 2.2. No endobronchial lesions were seen in FOB. Malignancy was not detected in lavage and bronchoscopy and sputum cytology. Laboratory parameters were within normal limits. Pulmonary function tests were within normal limits (FVC: 2.94 lt, 98%, FEV1: 2.75 lt, 96%). Informed consent was obtained. Left upper lobectomy and MLND was performed because of malignant pathology on frozen section. The definitive pathology was pleomorphic primary malignant fibrous histiocytoma of the lung (Fig. 3.b). The patient did not receive chemotherapy or radiotherapy in the postoperative period. No pathology was detected in 48-month follow-up period.

RESULTS

Figure 1.a preoperative posterior anterior chest X-ray.

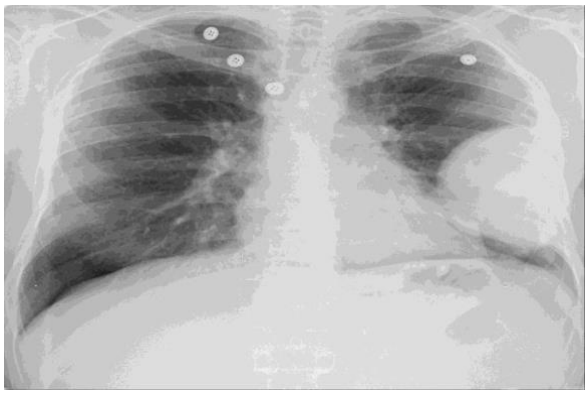


Figure 1.b histopathological evaluation
Immunohistochemical examination cd 34: negative desmin: negative panck: negative ema: negative vimentin: positive

s-100: negative sma: negative Histomorphological findings (high cellularity and mitotic index: 12-13 mitosis / 10bba) and immunohistochemical findings suggest anterior planmalign mesenchymal tumor .

Histopathological photograph of the case: closely packed spindle cells, folded pattern, atypia and mitotic fusiform nuclei (HE; x 40)

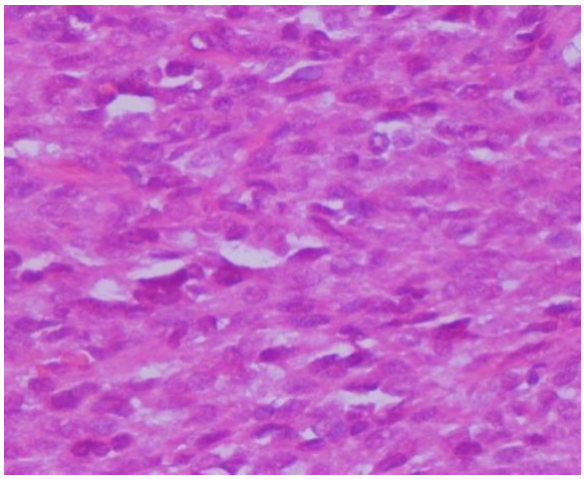


Fig.2.a PA chest X-ray showed an increase in density of about three centimeters in the left lung.

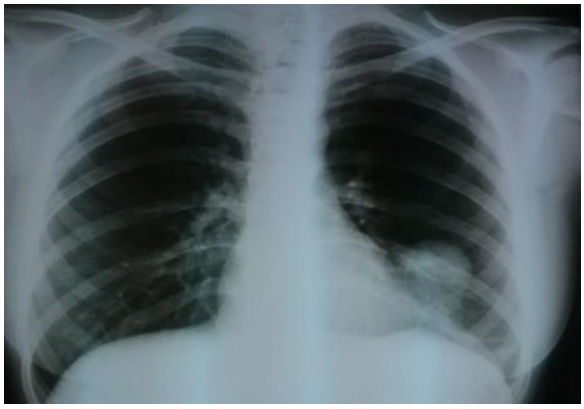


Figure 2.b CT revealed a soft tissue lesion of 33x42 mm in the left lower lobe of the lung.

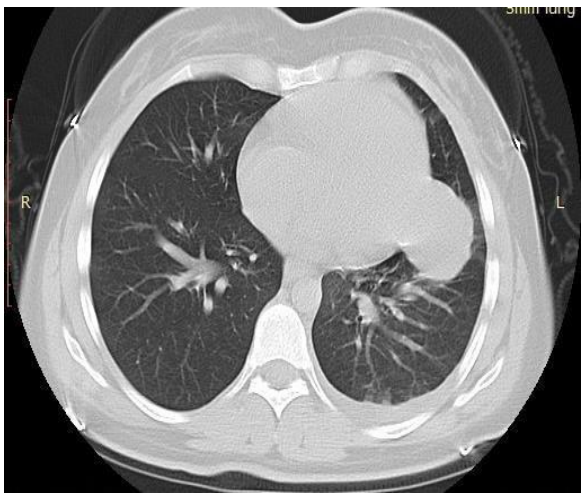


Figure 2.c histopathological photographs. : Firmly processed spindle cells, pre-patterned, atypical and mitotic fusiform nuclei HE; x 10)

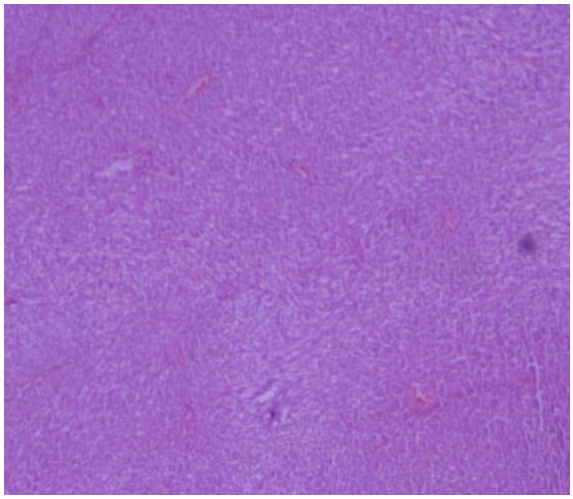


Figure 3.a CT revealed a smooth soft tissue density of approximately 10 mm in diameter at the anterior segment level in the left upper lobe of the lung

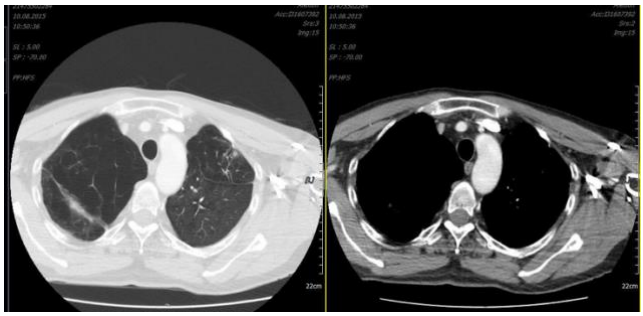
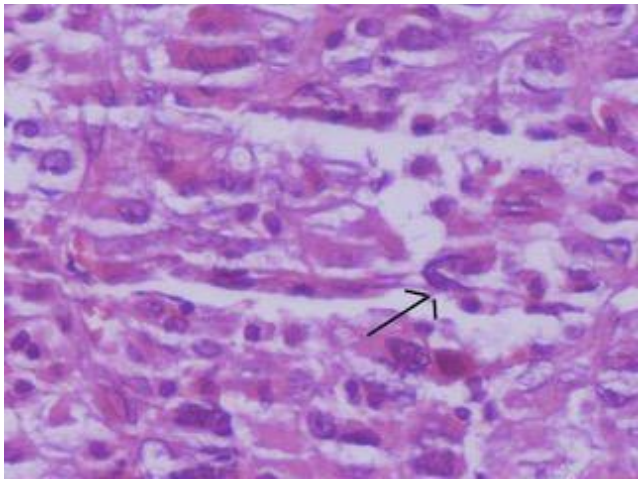


Figure 3.b Histopathological photograph of the case. : Pleomorphic cells, high atypia, rich vascularity in stroma (HE; x 40)



DISCUSSION

Mesenchymal tumors of the lung, which are rare tumors, although rarely benign, may have malignant features and recurrence [4]. Mesenchymal tumors are found in a group that differs in terms of various histological types,

survival and treatment approaches. These tumors originate from the embryonic mesoderm. Mesenchymal tumors constitute about 1% of all cancers and the most common sites are the extremities (59%). Other major localizations are thorax (20%), retroperitoneal (15%) and head and neck regions. Tumor size and depth are also important prognostic factors [5]. Although rarely benign, mesenchymal tumors have been reported to have malignant features with giant dimensions. They can be observed in a wide age range. Three cases presented in this study were observed in the third and fourth decades. Tumor is usually asymptomatic clinically. However, cough, chest pain and shortness of breath are the most common complaints due to localization of the mass. PA chest X-ray has a large preliminary in radiological imaging. Early diagnosis is difficult because they have a clear, limited appearance radiologically [6].Mesenchymal tumors tend to appear as endobronchial lesions in childhood and adolescence, while tending to be solitary in adulthood [7]. In all three cases, the lesions were round, smoothly confined and easily confused with lesions that could be confused with cyst hydatid. Lesions had a thin-walled appearance resembling necrotic material. At the same time, in two of our cases, there was an interesting history of leukemia in the family.

Mesenchymal tumors give better results with surgical chemotherapy and radiotherapy than bronchial carcinomas of lung origin [8]. Five-year survival rates for bronchial carcinomas in lung-derived patients range from about 40-70% [9]. It is not known whether mediastinal lymph node dissection is beneficial. It is generally recommended in the literature, but in most cases only anatomic resection has been performed. In a study involving all mediastinal tumors with 200 patients the metastasis rate was 42%. In the same study, the most common metastasis was found to be in the lungs with 82% [10]. Morshuis et al. reported that patients with malignant fibrous histiocytoma who received post-resection radiotherapy had no recurrence in their follow-up [11].

ETHICS COMMITTEE APPROVAL

Required Ethics Committee Approval was obtained.

CONCLUSION

When mesenchymal tumors are rare and diagnosis rate is very low with noninvasive methods, thoracotomy for diagnosis and treatment and thoracoscopy for appropriate cases can be considered. Anatomic resection must be performed.

All of our patients were young patients, their lesions were round, well-circumscribed and frequently confused with hydatid cysts. Therefore, it should be kept in mind that it may be an aggressive tumor even if the lesions are well-circumscribed lesions in young patients.

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