Giant Pulmonary Hamartoma-Case Report and Review of the Literature

Ebru ÇAKIR*, Funda DEMİRAĞ*, Sadi KAYA**, Hakan ERTÜRK***

* Atatürk Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim ve Araştırma Hastanesi, Patoloji Bölümü,
** Atatürk Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim ve Araştırma Hastanesi, Göğüs Cerrahisi Bölümü,
*** Atatürk Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim ve Araştırma Hastanesi, Radyoloji Bölümü, ANKARA

SUMMARY
Pulmonary hamartomas, the most common benign tumour of the lung, are usually an incidental finding and range in size between 1-5 cm in various series. There are only 11 published giant hamartoma cases in the English literature with a diameter of 9 cm or more. We report a case of a giant pulmonary hamartoma in a 43 years old man who was admitted to our hospital with chest pain lasting for 3 months. Radiologic examination showed a giant heterogenous solid intrapulmonary mass with dense punctate microcalifications in the right lung. The resected tumour was 11 x 9.5 x 8.5 cm gray, encapsulated and multilobulated solid mass. The histopathologic examination revealed chondroid hamartoma. The patient is healthy with no sign of recurrence 18 months after the operation.

KEY WORDS: Hamartoma, lung, giant size

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ÖZET
DEV AKCİĞER HAMARTOMU-OLGU SUNUMU ve LİTERATÜR İNCELEMESİ
Akciğerin en sık görülen benign tümörleri olan hamartomlar genellikle rastlantısal olarak bulunur ve boyutları farklı serilerde 1-5 cm arasında değişkenlik gösterir. İngiliz literatüründe boyutu 9 cm ve üzerinde olan sadece 11 dev hamartom bulunmaktadır. Bu çalışmada, 3 aydır devam eden göğüs ağrısı şikayeti ile hastanemize başvuran, 43 yaşındaki erkek hastada dev hamartom olgusu sunuldu. Radyolojik incelemede sağ akciğerde yoğun naktalı mikrokalsifikasyonlar içeren dev heterojen kitle mevcuttu. Rezepsiyon edilen tümör 11 x 9.5 x 8.5 cm ölçüleriinde kapsülü, multilobule, solid kitle lezyondu. Histopatolojik inceleme chondroid hamartom olarak değerlendirildi. Hasta operasyondan sonra 18. ayda sağlıklı ve nüksü yoktu.

ANAHTAR KELİMELER: Hamartom, akciğer, dev boyut

**INTRODUCTION**

Pulmonary hamartomas, also known as mesenchymomas, are the most common type of benign lung tumours, composed varying proportions of mesenchymal tissues and entrapped respiratory epithelium. They were considered as developmental abnormalities in the past but now considered as benign mesenchymal neoplasms (1-6).

The incidence of pulmonary hamartoma is 0.25% with a two to four fold male predominance. Most are found in the 6th decade. Clinical presentation is usually asymptomatic solitary nodule on the routine chest X-ray (1-3). They are usually peripheral parenchymal lesions and less than 5 cm in diameter. There are only 11 published giant hamartoma cases in the English literature with a diameter of 9 cm or more (7-17). Present case is a 43 years old male with a giant hamartoma in the right lung.

**CASE**

A 43-year-old man was admitted to our hospital with chest pain lasting for three months. Physical examination revealed decreased respiratory sounds in the right lung. Chest X-ray Im and thorax computed tomography (CT) showed a giant heterogenous solid intrapulmonary mass with dense punctate microcalcifications and minimal compression effect in the lower lobe of the right lung (Figure 1). The tumour was resected by enucleation. Macroscopic appearance of the tumour was 11 x 9.5 x 8.5 cm gray, encapsulated and multilobulated solid mass (Figure 2). Microscopically lobules of mature cartilage, loose connective tissue and mature adipose tissue interspersed by cleft like space lined by non-neoplastic bronchiolar type epithelium were seen. The tumour lobules were predominantly made of cartilaginous tissue (Figure 3). The pathologic diagnosis was chondroid hamartoma. After a period of 18 months follow-up, our patient is healthy with no sign of recurrence.

**DISCUSSION**

The term hamartoma refers to an excessive but focal over-growth of cells and tissues native to the organ in which it occurs. Although the cellular elements are mature and identical to those found in the remainder of the organ, they do not
reproduce the normal architecture of the surrounding tissue. They have been described in many organs but especially in the chest, breast, skin, brain, liver and eye (1-4).

Pulmonary hamartomas are the most common benign tumours of the lung. They are benign connective tissue neoplasms rather than tumour like malformations (3,4). Their rarity in childhood, growth in adult life and identification of chromosomal rearrangements (6p21 and 12q14-15) are similar in lipomas and leiomyomas favour them being neoplasm (1,5,6).

These tumours are constituting about 8% of all “coin” lesions in chest radiographs (18). They appear as a solitary, round nodules sometimes with punctate or popcorn calcification. While most of the usual hamartomas measure 1-5 cm in diameter, giant hamartomas reported in the English literature measure between 9 to 30 cm (1,3). Most of them are female patients in contrast with the usual hamartomas. Although the age range is wide, some of the patients are younger than the expected age (30-63 years, mean age of 48.9) (Table 1). Petheram and colleagues reported a huge hamartoma with a diameter of 30 cm (9). This patient was a 32 years old male. Most of the giant hamartomas in the literature localized in the right lung. In the left lung, lesions localized mostly in the lower lobe. None of the cases had endobronchial lesion (Table 1). Our case is a 43-year-old man. He has a giant hamartoma in the right lower lobe with no endobronchial lesion in concordance with the literature.

Unlike carcinoid tumours, more than 90% of hamartomas are peripheral. Only 10% or less are located centrally. They sometimes show a topographical relationship to small bronchi and bronchioles. Tomiyasu and colleagues documented an unusual example that penetrated the visceral pleura (19). The peripheral location of pulmonary hamartomas usually renders them asymptomatic. Occasionally, the tumours may be endobronchial and they cause hemoptysis or bronchial obstruction with signs and symptoms of coughing, wheezing, expectoration, leukocytosis and fever. Most of the giant hamartomas in the literature show symptoms like cough, pain and dyspne despite their peripheral location. Our case had also chest pain lasting for three months probably due to the giant size of the tumour.

In general, hamartomas are discovered incidentally on routine chest roentgenograms as an asymptomatic coin lesion. On chest radiographs, pulmonary hamartomas characteristically appear as well-defined solitary pulmonary nodules, which may show varying patterns of calcification, including an irregular popcorn, stippled, or curvilinear pattern or even a combination of all three.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Symptom</th>
<th>Localization</th>
<th>Size (cm)</th>
<th>EBL</th>
<th>Component</th>
</tr>
</thead>
<tbody>
<tr>
<td>Darke (7)</td>
<td>51</td>
<td>F</td>
<td>Breathlessness</td>
<td>Left lower</td>
<td>10</td>
<td>Absent</td>
<td>Cartilage</td>
</tr>
<tr>
<td>D’Altorio (8)</td>
<td>60</td>
<td>M</td>
<td>Cough</td>
<td>Right upper</td>
<td>18</td>
<td>Absent</td>
<td>Cartilage</td>
</tr>
<tr>
<td>Petheram (9)</td>
<td>32</td>
<td>M</td>
<td>NA</td>
<td>Right</td>
<td>30</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Okabayashi (10)</td>
<td>43</td>
<td>F</td>
<td>Cough, fever, pain</td>
<td>Left lower</td>
<td>9</td>
<td>Absent</td>
<td>Cartilage</td>
</tr>
<tr>
<td>Kervancioglu (11)</td>
<td>49</td>
<td>F</td>
<td>Pain, cough, dyspne</td>
<td>Right</td>
<td>26</td>
<td>NA</td>
<td>Fibrolipolefibromyxomatous</td>
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<tr>
<td>Fujino (12)</td>
<td>62</td>
<td>F</td>
<td>Asymptomatic</td>
<td>Right</td>
<td>NA</td>
<td>NA</td>
<td>Cartilage</td>
</tr>
<tr>
<td>Lee (13)</td>
<td>30</td>
<td>M</td>
<td>Asymptomatic</td>
<td>Right</td>
<td>16</td>
<td>Absent</td>
<td>Cartilage</td>
</tr>
<tr>
<td>Kim (14)</td>
<td>38</td>
<td>M</td>
<td>NA</td>
<td>Left lower</td>
<td>11.5</td>
<td>Absent</td>
<td>Cartilage, adipose</td>
</tr>
<tr>
<td>Park (15)</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Hutter (16)</td>
<td>63</td>
<td>F</td>
<td>Reduced endurance</td>
<td>Left</td>
<td>26</td>
<td>NA</td>
<td>Absent</td>
</tr>
<tr>
<td>Ganti (17)</td>
<td>61</td>
<td>F</td>
<td>Breathlessness</td>
<td>Right lung</td>
<td>25.5</td>
<td>Absent</td>
<td>Adipose, leiomyomatous</td>
</tr>
<tr>
<td>Present case</td>
<td>43</td>
<td>M</td>
<td>Pain</td>
<td>Right lower</td>
<td>11</td>
<td>Absent</td>
<td>Cartilage, adipose</td>
</tr>
</tbody>
</table>

NA: Not available, M: Male, F: Female.
Specific popcorn-type calcification is almost pathognomonic for pulmonary hamartoma. Thorax computed tomography helps to make a differential diagnosis (1-4). Our case has also typical dense punctate microcalcifications in radiologic examination and easily identified as a hamartoma.

The histology of the parenchymal lesions in both giant and usual hamartomas usually reveals a predominant chondroid differentiation (80%), with fibroblastic (12%), fatty (5%) and osseous (3%) differentiation making the rest. Endobronchial lesions tend to have more fat (9). Myxomatous connective tissue, smooth muscle, blood vessels and other mesenchymal elements may also be seen histologically. These tissues are arranged in a disorganized manner. Our case also reveals a predominant chondroid differentiation.

Most tumours grow slowly (average of 3 mm/y) during follow-up. Surgical treatment is the gold standard in intraparenchymal hamartomas including enucleation, lobectomy or sleeve resection, wedge resection, segmentectomy and pneumonectomy. Endobronchial hamartomas can be removed successfully through bronchoscopy. Although hamartomas are benign tumours, they may rarely assume malignant characteristics and may be invasive to surrounding tissues. Besides, some other lung pathologies may rarely accompany pulmonary hamartomas such as bronchiectasis, primary lung cancer, tuberculosis, and metastatic tumours of the lung. It has been recommended that patients with hamartoma should be evaluated and closely followed up with respect to the risk for associated synchronous malignancies (1-3,19,20). Our case has no associated pulmonary pathology and at 18 months follow-up he is healthy with no sign of recurrence.

Hamartomas are common benign tumours of the lung but there are only a few published giant hamartoma cases in the literature. The present case has typical histological and radiological features of chondroid hamartoma but the size of the tumour makes it an unusual case. We report a case of giant pulmonary hamartoma and review the clinical features of the usual and giant hamartomas in the English literature.

REFERENCES


Yazıma Adresi

Ebru ÇAKIR
Atatürk Göğüs Hastalıkları ve Göğüs Cerrahisi Eğitim ve Araştırma Hastanesi
Patoloji Bölümü
Keçören-ANKARA

e-mail: arabaci.ebru@gmail.com
   ebruarabaci@hotmail.com