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A Giant Bullae Manifesting as Left Pulmonary Arteria Hypoplasia

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Özet

Sol Pulmoner Arter Hipoplazisi Düşünüren Dev Bül

Bir hemitoraks hacminin yarısından fazlasını kaplayan büller dev bül olarak tanımlanır. Genellikle ilerleyici bir nefes darlığı olan bu hastalarda altta yatan akciğer parankiminde amfizematöz değişikliklerin olup olmadığı önemlidir. Biz de bu çalışmada, radyolojik olarak sol pulmoner arter hipoplazisini taklit eden dev tip 1 bülü olan bir olguyu sunmayı amaçladık. Hastaya sol torakotomi aracılığı ile bül rezeksiyonu uygulandı ve postoperatif dönemde sağlam parankimin uzun süreli basısına bağlı olarak reekspansiyon pulmoner ödem komplikasyonu gelişti. Hasta sorunsuz olarak taburcu edildi. Cerrahiden iki ay sonra solunum fonksiyon testlerinde mükemmel bir düzelme saptandı.

Anahtar kelimeler: *Pulmoner arter, Göğüs Cerrahisi, bül*

SUMMARY

The giant bullae is defined as the bullae formation of lung tissue which is larger than one half of a hemithorax. The presenting symptom is usually progressive dyspnea and the most important thing with this patient is that the underlying lung parenchyma has emphysematous changes or not. In this study, we aimed to presente a case with giant type I bullae whose lesion was mimicking left pulmonary arteria hypoplasia radiologically. The ginat bullae was resected via left thoracotomy and a complication of re-expansion pulmonary eudema occurred due to long time compression of viable parenchyma. The patient was discharged without any problem. The pulmonary function test showed excellent improvement after two months from surgery.

Key words: *Pulmonary artery, thoracic surgery, bullae*

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Introduction

The giant bullae is defined as the bullae formation of lung tissue when it occupies at least one third of a hemithorax. Young male smokers are usually affected and the large bullae is usually orginated from upper lobes of the lungs. Giant bullous emphysema or vanishing lung syndrome is a distinct clinical syndrome from primary bullous disease of the lung. This condition is characterized by large bullae as the predominant finding but is also associated with several forms of emphysema, which is very important in operative planning and in selection of patients. Herein we presented a case with giant type I bullae manifesting as left pulmonary artery hypoplasia who were treated with simple bullectomy successfully.

Case Presentation

A 34-year-old man admitted to a Chest Disease Department with the complaint of progressive dyspnea and ches pain last 10 months. The decrease of breath sounds on the left side was the only abnormal finding on physical examination. Routine haemogram and biochemistry examination were normal. The left sided hyperinflation was discovered on chest X-ray. Pulmonary function test revealed restrictive pattern (FEV1/FVC: 76%, FVC: 74% and FEV1: 67%). Thorax Computed Tomography (CT) revealed bullous left upper lobe, partial atelectasia due to compression of giant bullae and a hypoplastic left pulmonary artery (Figure 2 a,b). The perfusion mismatching of left upper lobe was determined in V/Q scintigrahpy. There was no endobronchial lesion in fiberoptic bronhcoscopic examination. He was refered to our clinic for surgical treatment of prediagnosis of infantile lobar emphysema or Swyer-James-Macleod syndrome. He underwent left thoracotomy and the giant type I bullae was resected (Figure 2,c). There were multiple left sided patch infiltrations on chest X-ray on the first postoperative day which was compatible with re-expansion pulmonary edema (Figure 1). The chest X-ray was normal on the seventh postoperative day and subsequently the patient was discharged in stable condition. The pulmonary function test revealed approximately 50 % improvement after two months (FEV1/FVC: 97%, FVC: 111% and FEV1: 104%).

Discussion

The association of emphysema with large bullae is a well defined clinical entitiy. The surgical treatment strategy and patient selection which is consisted of increasing bullae size, compressing to the normal lung, pneumothorax, pulmonary insufficiency, and infection within the bulla are comprehencively discussed in the literature (1,2). However, the type I giant bullae without existing of emphysema was not discussed in the literature up till now. Our case was an extremely rare because

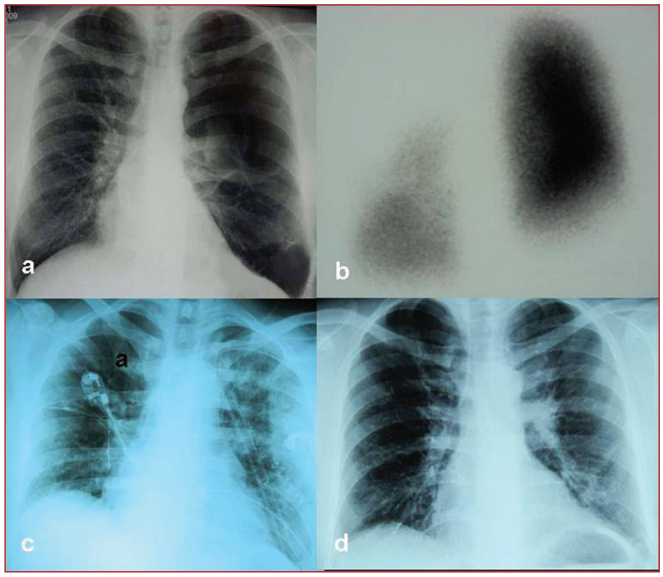

Figure1: Hyperinflation of the left lung on chest X-ray (a), there were no perfusion and ventilation for left upper lobe on V/Q scintigrahpy (b), infiltration was seen on the left side due to re-expansion of the compressed lung on the first postoperative day (c), normal chest X-ray after seven days from resection (d).

there were not any emphysematous changes in the other lung field and the prediagnosis of patient was challenging.

The differential diagnosis of this condition is including infantile lobar emphysema, Swyer-James-Macleod syndrome and pneumothorax. The distinguish of pneumothorax is challenging especially in the emergency department. Chest tube insertion is contraindicated in the treatment of giant bullae. Because the insertion of chest tube into the bulla results in sudden air drainage and asphyxia originates from bronchopleural fistula, which may be life-threatening (3). The “double-wall sign” which is defined as air outlining both sides of the bulla wall parallel to the chest wall is the distinctive finding in patients with pneumothorax (4). In our case chest X-ray was not considered pneumothorax and the only abnormal finding was hyperinflation on the left side. So we made the other diagnostic procedure not for the confirmation of pneumothorax whether present or not.

High-resolution CT is useful for the differentiation of the types of emphysema, such as paraseptal and centrilobular. The patients with giant bullae can be divide into three subgroup. Giant bullae is well demarcated and almost normal underlying pulmonary parenchyma in group 1, giant bullae is associated with diffuse emphysema in group 2, and giant bullae with a complete loss of parenchyma (vanishing lung) in group 3 (5). According to this classification our case was in group 1. However radiographic findings on CT of our case was interesting because the bullae was not consist of air density. It was presenting as emphysematous left upper lobe such as congenital lobar emphysema and hypoplastic left pulmonary artery. We made V/Q scintigrahpy for definitive diagnosis and the findings were consistent with Swyer-James-Macleod syndrome (Figure 2).

Pulmonary function test revealed moderate restrictive pattern due to compression of giant bullae but the improvement was excellent after two months form surgical removal of giant bullae.

In conclusion, when there is a giant bulla in one hemithorax, the differantial diagnosis can be challenging due to different presentation of the patients. If a giant bullae with normal

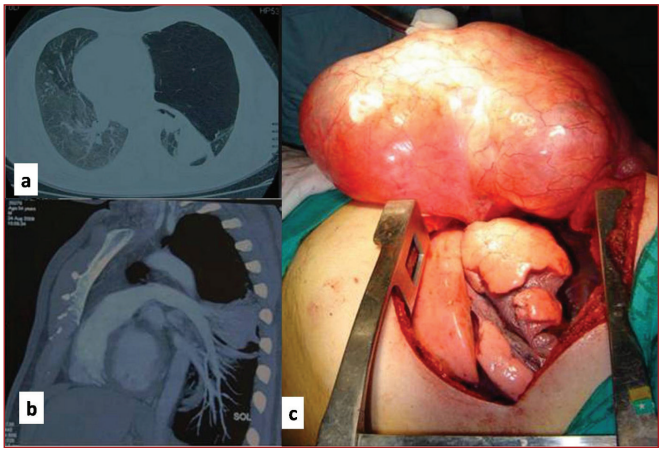

Figure2: The appearence of the bullae and compressed lung on axial and sagittal plane on thorax CT and hypoplasia of the left upper lobe artery (a,b), intraoperative view of the bullae (c).

lung parenchyma resected it will result in expansion of the compressed lung parencyhma and pulmonary function will demonstrate significant improvement and also we consider in mide that re-expansion pulmonary edema can be occurred due to long time compression of viable parenchyma.

References:

1. Laros CD, Gelissen HJ, Bergstein PG, Van den Bosch JM, Vanderschueren RG, Westermann CJ and et al. Bullectomy for giant bullae in emphysema. J Thorac Cardiovasc Surg 1986;91:63–70.
2. Mehran RJ, Deslauriers J. Indications for surgery and patient work-up for bullectomy. Chest Surg Clin N Am 1995;5 (4):717–34.
3. Gokce M, Saydam O, Altın R, Kart L. Giant bulla mimicking tension Pneumothorax. Tüberküloz ve Toraks Dergisi 2009; 57 (4): 435-8.
4. Waitches GM, Stern EJ, Dubinsky TJ. Usefulness of the double-wall sign in detecting pneumothorax in patients with giant bullous emphysema. Am J Roentgenol 2000;74:765-8.
5. Witz JP, Roeslin N. La chirurgie de l’emphyse`me bulleux chex l’adulte: ses re´sultats e´loigne´s. Rev Fr Mal Respir 1980;8 (2):121–31.