

Case Report:**SEMILUNAR SIGN OF TRANS-MEDIASTINAL HERNIATION OF GIANT BULLAE WITH TENSION HYDROPNEUMOTHORAX****Anastasia Tjan, I Made Dwija Putra Ayusta, Dewa Gde Mahiswara**

Department of Radiology, Faculty of Medicine, Udayana University-Sanglah Public General Hospital, Denpasar, Indonesia

ABSTRACT

Herniation of bulla across mediastinum is rare, while transmediastinal giant bulla herniation accompanied with hydropneumothorax is even rarer. We report a case of an 18 years old male with dyspnea came to emergency department with trans-mediastinal giant bulla herniation, which appears as semilunar sign on chest x-ray, and right hydropneumothorax. It appeared that the giant bulla also infected by the presence of air fluid level within. Semilunar sign was seen on the contralateral left mediastinum as the hallmark finding for trans-mediastinal herniation of bulla. Chest CT further confirms the diagnosis. Subsequently chest tube insertion and symptomatic relieves were given, however the patient end up dead after 2 days of observation. Heart and lung compression by the lesions were the cause of this patient poor outcome. Bullous lung disease should be evaluated thoroughly and not underestimated since it could cause severe disease progression.

Keywords: Giant bulla; transmediastinal herniation bulla; chest x-ray; CT scan

ABSTRAK

Herniasi bula melewati mediastinum merupakan kondisi yang jarang, sementara itu herniasi giant bulla dengan hidropneumothoraks lebih jarang lagi ditemukan. Kami melaporkan sebuah kasus seorang laki-laki berusia 18 tahun dengan dispnea datang ke instalasi gawat darurat dengan diagnosis herniasi giant bulla trans-mediastinal, yang tampak sebagai tanda semilunar pada foto polos thoraks, disertai dengan hidropneumothoraks kanan. Giant bulla tersebut menunjukkan gambaran terinfeksi dengan adanya gambaran air fluid level didalamnya. Tanda semilunar yang dilihat pada sisi kiri mediastinum kontralateral merupakan tanda untuk mengetahui adanya herniasi bulla melalui mediastinum yang telah di konfirmasi melalui computed tomography scan thoraks. Insersi selang dada dan terapi simptomatis diberikan kepada pasien, namun setelah 2 hari observasi pasien meninggal. Penyakit paru bulosa harus dievaluasi secara menyeluruh dan tidak diremehkan karena dapat menyebabkan perkembangan penyakit yang parah.

Kata kunci: Giant bulla; herniasi bulla trans-mediastinal; x-ray thoraks; CT Scan

Correspondence: Anastasia Tjan, Darmo Villa 705, Surabaya/Jl. Pulau Nusa Penida 20, Denpasar, Indonesia.
Phone: 082111700491. E-mail: anz.4191@gmail.com

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INTRODUCTION

Bullous lung disease is common lung conditions which can be seen in plain radiograph as diminish density area with smooth margin and thin wall less than 1 mm, measured more than 1 cm diameter within lung parenchyma. The termed giant bulla is used if the lesion occupies more than one third hemithorax or 30% larger, giant bulla also can affect the entire lobe. Infected giant bulla is considered if there is a wall thickening with or without air fluid level (Cosgrove et al 2007).

Radiologically bulla is considered subcategories of cyst according to the third Fleischer society meeting. Bulla defined as a cyst with diameter more than 1 cm, sharply demarcated avascular lucency area with thin wall less than 1 millimeter and smooth margin (Boddu et al 2017).

In children there are three main categories of cystic lung disease, congenital, acquired and idiopathic. Acquired cystic lung disease can be caused by the sequel of prior lung infections resulting into developing cyst into giant

bullae due to its check valve mechanism, or bronchiectasis (Odev et al 2013).

The purpose of this paper is to familiarize semilunar sign from chest x-ray as the radiological appearance of trans-mediastinal bulla herniation which cause mortality and to increase clinical awareness of any bullous lung disease.

CASE REPORT

An 18 years old male was brought to emergency department for shortness of breath, chills, and fever which were noticed for 2 days prior admission. He has history of cough and no prior history of asthma, therefore he was diagnosed clinically with pneumonia. He was treated successfully with antibiotic 4 years ago due to unspecific lung infection. His physical appearance from head to toe appeared normal, neither musculoskeletal abnormalities nor craniofacial abnormalities were detected. His physical examination revealed decreased breath sounds on the right chest without any additional breath sounds from auscultation, subsequently chest X-rays were ordered.

Supine anterior-posterior projection chest x-rays showed area of avascular lucency with air fluid level on the right hemithorax, as well as a semi thinned wall bulla with wall thickening on the inferior lateral part on the right hemithorax crossing into the midline to the left hemithorax. Deviation of the heart and trachea to the left were observed, with normal left visualized lung parenchyma, on his half erect AP projection showed 3 air fluid levels, one is on the right hemithorax below the avascular lucency area, the second one was inside the bulla, and the third one was beneath the right hemidiaphragm which resembles the stomach gas. His right lateral decubitus x-rays showed the migration of the bulla to the superior part, and 3 air fluid levels were still observed. The third air fluid level was suspected with another infected bulla in the right lung base (Figure 1). Chest X-rays concludes tension hydropneumothorax of the right hemithorax, transmediastinal herniation of giant infected bulla, as well as suspicion of another bulla on the right lung base. A chest tube was performed to reduce the tension hydropneumothoraks on the right side, and then another follow up chest X-ray and CT was performed.

The patient was prepared to have bullectomy, however his condition could not meet the anesthesiologist expectation with poor forced expiratory volume below 30%, hypercapnea and severe hypoxemia, thus his surgery was postponed.

His follow-up chest x-ray showed decreased tension hydropneumothorax and normal heart and trachea position (Fig. 2). His Chest CT confirmed a giant bulla, 14 x 10.5 x 5.3 cm, on the right hemithorax which crossing midline to the anterior mediastinum with multiple bullae inferiorly which confirmed the plain film diagnosis there were another bullae on the right lung base, right hydropneumothorax and right lung collapse with slight heart deviation to the left were also seen.

Several chest x-rays were taken to evaluate disease progression while the patient was on chest tube, with slowly decrease of right hydropneumothorax. After 2 days of hospitalization, his condition was deteriorate with decreased of consciousness, respiratory distress, apnea, therefore a code blue resuscitation was alert. However, unfortunately, he was failed to resuscitate and died.

DISCUSSION

In children there are many differentials of cystic lung disease unlike that in adults. Lesions such as congenital cystic adenomatoid malformation, congenital lobar emphysema, and bronchogenic pulmonary cyst were quite common congenital causes of cystic lung disease in children should be considered (Odev et al 2015). The underlying etiology of bullous lung disease itself is diverse and this patient's bullae pathogenesis was unknown (Fatimi et al 2012). He was a healthy individual prior to the first symptoms, no previous complains of any kind congenital lung lesion manifestations.

His physical appearance was normal hence several associated syndrome can be excluded such as Ehlers-Danlos syndrome, Marfan syndrome and other connective tissue diseases (Papisiris et al 2015). Prior lung infection was acknowledged and treated successfully with antibiotics 4 years ago, therefore acquired bullae as the sequel of lung infections was still the suspected underlying mechanism of this case.

In our case the patient was an adolescent came with respiratory distress, cough and fever. Radiological finding was tension pneumothorax and giant infected bulla, which migrate crossing midline to anterior mediastinum and make a linear convexity on the contralateral side (Singh et al 2015). Tension hydropneumothorax is an emergency situation that needed direct intervention. A ruptured bulla was believed to be the nature cause of this patient's pneumothorax.

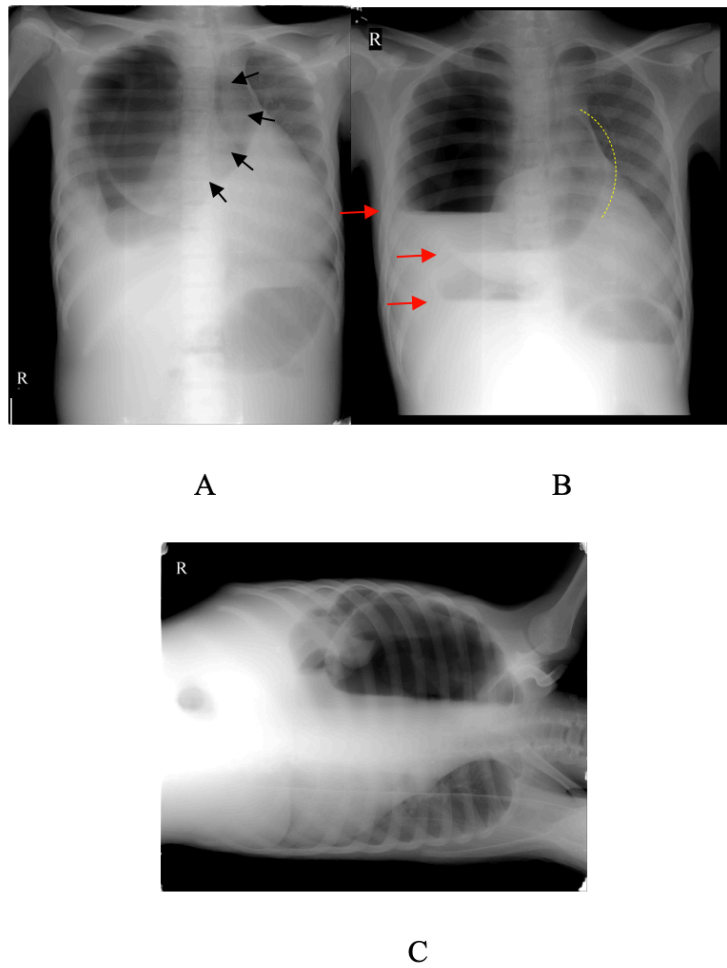


Fig. 1. Chest Radiograph with 3 positions. A) AP supine, B) Erect, C) Left lateral decubitus. There is convexity just right to the left mediastinum (arrow) which resembles a semilunar shape hence it is called semilunar sign (yellow dashes). There were 3 air fluids levels seen on erect position (red arrow).

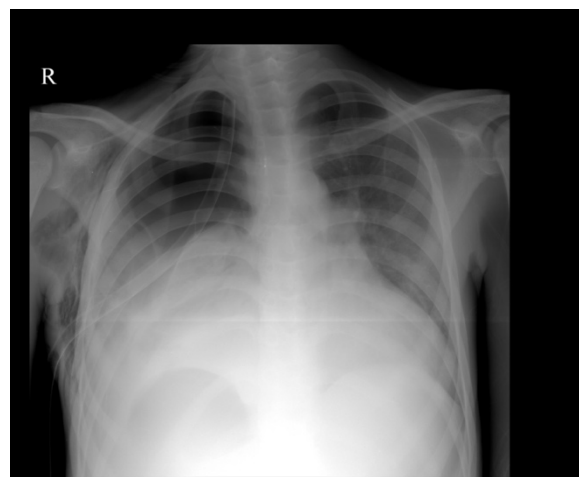


Fig. 2. Chest X-Ray after chest tube insertion.

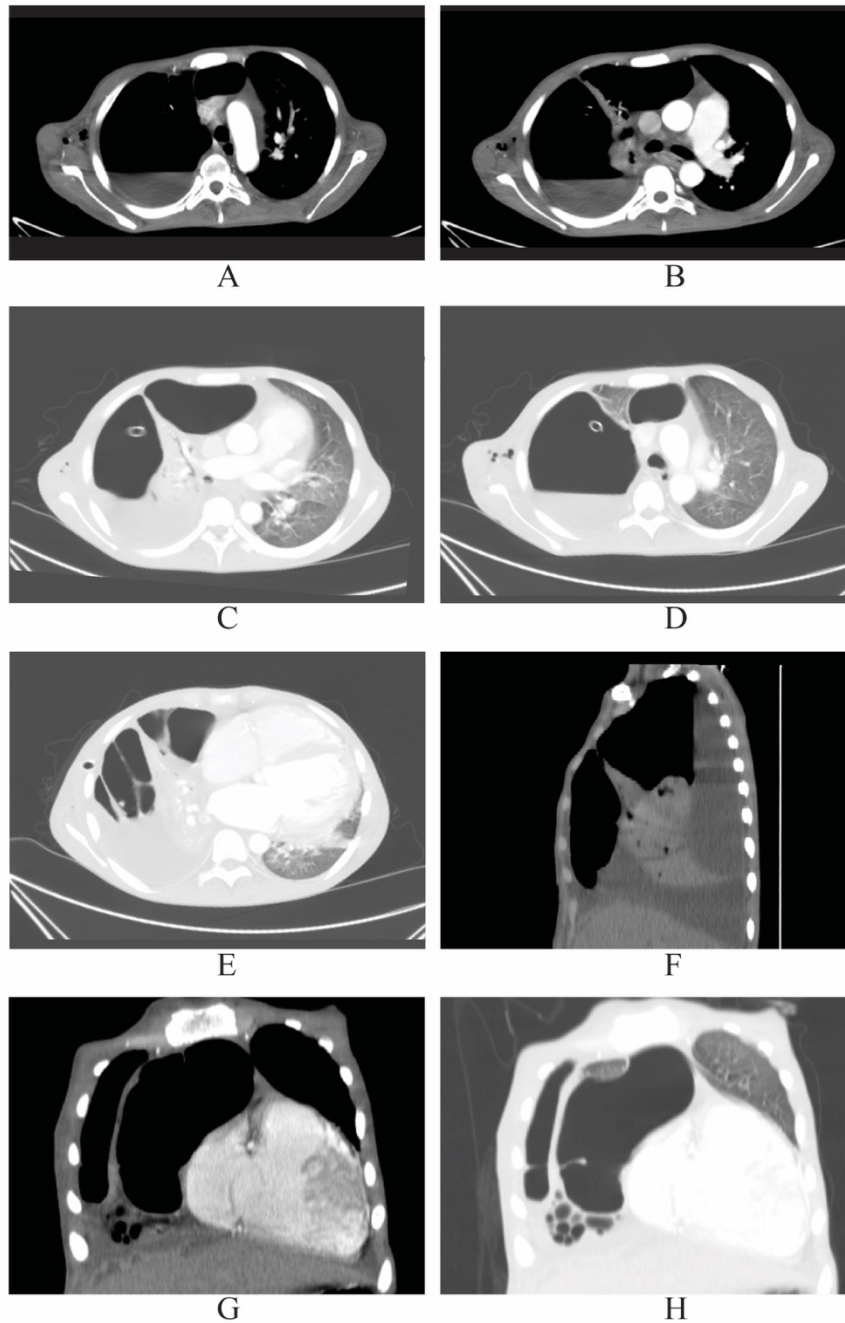


Fig. 3. Chest CT Axial, Sagittal, Coronal in mediastinum and lung window. Right giant infected bulla that herniated to the anterior mediastinum with multiple bullae in the right lung base accompanied with right hydropneumothorax and visualized chest tube distal tip.

Multiple bullae on this patient were also detected with thickening of the bullae wall and air fluid level, which indicated an infection on the bullae. Herniation of a giant bulla is a very rare condition, which can have poor prognosis such in this case. Some reported cases of

bullae herniation that were successfully treated with surgical intervention such as bullectomy and lung volume reduction surgery, while giant bullectomy itself carries significant complication following surgery (Schipper et al., 2004). Indication of surgery is severe

dyspnea with bulla larger than 30% of lung volume compressing the adjacent lung fields (Garcia-Herrerros et al 2018) that apparently well suited for this case, unfortunately his unimproved condition was unlikely to have such a big surgery. Several contraindications for bullectomy such as significant comorbid disease, forced expiratory volume below 30%, the elevation of carbon dioxide levels in blood, severe hypoxemia and cor pulmonale (Im et al 2016). Minimally invasive procedure should be considered in more advanced health care center, however is limited in our center (Batirel & Zellos 2015).

Convex line in nearby the left side mediastinum, so called semilunar sign, is the clue of trans-mediastinal shift of bulla (Singh et al 2015). The presence of pneumothorax make the margin of the bulla even more circumscribe and greatly visualized hence the herniation of bulla can be seen precisely in both sides of mediastinum, in contrast to normal lung which could make one side of the bulla wall curtailed. Plain radiograph is needed as the first evaluation of any respiratory problems. The presence of pneumothorax in supine position can be confirmed with additional projection such as cross table, contralateral decubitus, or half erect position. Tension hydropneumothorax was diagnosed from chest x-ray with the appearance of air fluid level, lung collapse and deviation of trachea and heart away from the avascular lucency area. Needle thoracostomy followed by chest tube insertion were done and another chest x-ray was performed to evaluate the patient's lung expansion and chest tube drainage.

Conservative managements were given along with chest tube monitoring for 2 days and afterwards the patient's condition became deteriorate, resuscitation was failed and subsequently death. The cause of death is believed due the impact in cardiorespiratory physiomechanics caused by the bullae and hydropneumothorax compression (Ruan et al 2011).

CONCLUSION

The underlying etiology of this patient giant bulla and multiple cystic lung appearance is considered the sequel of prior lung infection. In this case the attention goes to the herniation of a giant bulla which compromising adjacent structure, which leads to fatal complication, death. Therefore both clinicians and radiologist are

needed to emphasize diagnosis investigation for bullous lung disease after lung infections to avoid fatal complication.

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