Development of Tracheobronchomegaly during Mechanical Ventilation: An Unusual Case Report

Mekanik Ventilasyon Sırasında Gelişen Trakeobronkomegali: Ender Bir Olgu Sunumu

Abstract

Tracheobronchomegaly, first described in 1932 by Mounier–Kuhn, is a well-known clinical and radiological entity characterized by marked weakness of the trachea. It can be congenital or acquired. Some well-known inflammatory and infectious conditions have been associated with the acquired form. Endotracheal intubation can traumatize the trachea and usually results in focal stenosis, but may infrequently cause focal tracheobronchomegaly. It is not clearly understood why some cases result in stenosis and others in dilatation. This study reports the case of a patient whose clinical course was complicated by the development of intrathoracic focal tracheobronchomegaly during mechanical ventilation.

Key Words: mechanical ventilation; Mounier–Kuhn syndrome; tracheobronchomegaly

Özet

Trakeobronkomegali, ilk defa 1932'de Mounier–Kuhn tarafından tarif edilmiş belirgin trakea zayıflığı ile karakterize, konjenital veya kazanılmış olabilen klinik ve radyolojik olarak iyi bilinen bir tablodur. Bazı iyi bilinen enflamasyonlu ve enfeksiyöz durumlar kazanılmış formu ile birlikte olabilmektedir. Endotrakeal entübasyon, trakeayı travmatize edebilmekte ve genellikle fokal darlık ile sonuçlanabilmektedir, ancak çok seyrek olarak fokal trakeobronkomegaliye sebep olabilmektedir. Bazı olgularda darlık veya dilatasyonun niye oluştuğu açıkça anlaşılamamıştır. Biz, mekanik ventilasyon sırasında klinik seyri komplike hale gelmiş olan bir intratorasik fokal trakeobronkomegali olgusu sunuyoruz.

Anahtar Kelimeler: mekanik ventilasyon; Mounier–Kuhn sendromu; trakeobronkomegali

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INTRODUCTION

Tracheobronchomegaly was first described in 1932 by Mounier–Kuhn, who associated it with radiographic appearances (1). It is characterized by marked dilatation of the trachea. The development of focal tracheobronchomegaly during mechanical ventilation is an extremely rare entity. It is characterized by marked dilatation of the trachea associated with Ehlers–Danlos syndrome, chronic smoking, chronic bronchitis, pulmonary emphysema, cystic fibrosis, multiple chondritis, and pulmonary fibrosis (2). It is always autosomal recessive, but the development of focal tracheobronchomegaly during mechanical ventilation is very rare. Early recognition of the condition may prevent further deterioration.

CASE

A 34-year-old woman was admitted with fever and dyspnea. Radiological and clinical evaluation revealed massive pleural effusion and thickened pleura in the left hemithorax. After obtaining informed consent from the patient's relatives, the patient underwent video-assisted thoracoscopy surgery (VATS) for drainage and obtaining samples. VATS showed that the lung parenchyma was covered with thick fibrin plaques that impeded the inflation of the lung. Thoracotomy was performed to decorticate and re-expand the lung. The first postoperative day was complicated with respiratory insufficiency. A chest radiograph displayed diffuse opacity over the entire right lung, while blood gas analysis revealed hypercarbia and severe hypoxia (pH: 7.26, pCO_2: 56.7 mmHg, pO_2: 38.6 mmHg, HCO_3: 29.2 mEq/L).

The patient's vital signs were noted a follows: blood pressure: 99/44 mmHg, heart rate: 115 beats/min, and O_2 saturation 95%. She was diagnosed with contralateral re-expansion pulmonary edema. She was taken to the intensive care unit, reintubated (endotracheal tube no. 7.5), and mechanically ventilated. Blood gas values were in the normal range throughout the course of ventilation, but the patient was intolerant of extubation. Serial chest radiographs displayed persistent infiltrates in the right lung. On the fifth postoperative day, bronchoscopy was performed by propelling the bronchoscope through the intubation tube, which revealed normal bronchoscopic findings. Daily chest radiographs were taken and these displayed a minimal but gradually enlarging tracheal lumen, but with the lack of awareness of tracheobronchomegaly, the diagnosis was missed. On the 12th postoperative day, tracheal enlargement was observed because of a huge tracheal air column that was evident on the chest radiogram (Fig.1). The endotracheal tubal pressure was 28 mmHg.

Urgent bronchoscopy was performed by propelling the bronchoscope through the endotracheal tube, but normal bronchoscopic findings were found. Computerized tomography (CT) at that time displayed focal tracheal enlargement at vertebrae C7–T3 (Fig.2). The transverse and sagittal diameters were 7.2 cm and 6.5 cm, respectively (Fig.3). Retrograde examination of the chest radiograms displayed enlargement of the lumen that had become minimally apparent on the third postoperative day, 4 cm above the carina, where there was contact between the cuff of the tube and the tracheal wall. No specific treatment was instituted for the condition. On the 17th day, the patient deteriorated, becoming tachycardic and tachypneic. Hypotension ensued. Cardiotonic infusion was started. In the following days, the deteriorated vital signs did not normalize. On the 20th day, cardiopulmonary arrest developed and the patient died.

DISCUSSION

Tracheobronchomegaly is a distinct clinical and radiological entity characterized by marked dilation of the trachea. The clinical features were described by Mounier–Kuhn. Destruction or atrophy of the elastic tissue and smooth muscle of the trachea is a dominant pathological finding (2), with an incidence of about 1/500 (3).The disease predominantly occurs in men in their third and fourth decades of life (4).

The pathogenesis of tracheobronchomegaly is multifactorial and complicated. It can be congenital or acquired. The congenital form is frequently diffuse in character and may be associated with connective tissue disorders such as Mounier–Kuhn or Ehlers–Danlos syndrome (5,6).

The acquired form of tracheobronchomegaly is rare, and is usually seen after a number of inflam-

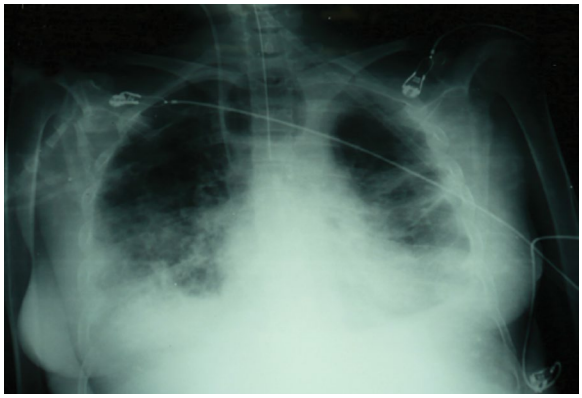

Figure 1: Chest radiogram on the 12th day showing enormous dilatation of the trachea.

matory and infectious conditions. Chronic cigarette smoking, chronic bronchitis, emphysema, cystic fibrosis, severe upper lobe fibrosis, and relapsing polychondritis may result in focal tracheobronchomegaly (3,7). In this case, no other associated comorbid conditions were present (diabetes, hypertension, hyperlipidemia, or immunosuppression) (8).

Endotracheal intubation by either the nasal or the oral route may result in trauma to the tracheal wall. Impaired blood supply, pressure necrosis, infection, friction of the tube, and dried tracheal mucosa are some known disadvantages of endotracheal intubation (7). The incidence of tracheal trauma following tracheal intubation ranges from 6% to 21%.Tracheal trauma mostly occurs around the cuff of the tube. When the tube cuff pressure is higher than the venous pressure, circulation ceases and ischemia ensues. Prolonged ischemia progresses to necrosis of the mucosa and cartilaginous rings. Healing usually occurs with granulation. Stenosis of the tracheal lumen, tracheomalacia, and fistula formation are some commonly encountered late complications of endotracheal intubation (9).

Tracheobronchomegaly is diagnosed when the transverse diameter of the trachea is greater than normal, exceeding 25 mm and 21 mm in men and women, respectively. The internal transverse diameter of the trachea is measured 2 cm above the top of the aortic arch on posterior anterior chest radiographs (10). Normally, the transverse diameter is13–25 mm in men and 10–21 mm in women (11). In this report, chest CT revealed the transverse diameter to be 7.2 cm and the sagittal diameter 6.5 cm. (Fig.3).

An enlarged tracheal shadow on a standard chest radiogram raises the suspicion of tracheobronchomegaly. CT is the gold standard in establishing the diagnosis (12).

The clinical manifestations of tracheobronchomegaly are nonspecific (7). The clinical course ranges from asymptomatic status to severe disability. Symptoms include severe cough and chronic respiratory infections (2). The weakness of the trachea is believed to result in inefficient cough. Diminished clearing of secretions leads to recurrent bronchopulmonary infections (2,10). A pulmonary function test reveals increased residual volume and obstruction (2,7).

Preventive measures are important to inhibit the deterioration of the patient's condition. Physiotherapy to assist in clearing secretions and appropriate antibiotics during infectious exacerbations are the cornerstones of treatment. Surgery has no role in the diffuse forms of the condition, but tracheal stenting may be useful in advanced cases (2). Resection of diseased segment and end-to-end anastomosis in focal forms can be an alternative to the conservative approach (9).

The case presented here is noteworthy in several respects. First, it is thought to be the first reported case of tracheobronchomegaly that developed and was diagnosed during mechanical ventilation. A few cases of tracheobronchomegaly associated with endotracheal intubation have been reported in the literature, (8,11,13,14), but these patients were sometimes diagnosed after discharge and presented with respiratory complaints.

Normal chest radiographs preoperatively and in the first 3 days of the postoperative period suggested that tracheal enlargement developed after endotracheal intubation. Retrograde examination of chest radiograms showed that the emerging point of the enlargement site was 4 cm above the carina, where there was contact between the cuff of the tube and the tracheal mucosa. Due to a lack of awareness of the condition, the diagnosis was missed even though several chest radiograph scans were taken, all of which revealed the development and progression of tracheobronchomegaly. Evaluation of chest radiograms by a radiologist might be helpful for early recognition.

Early observation of the condition might prevent further deterioration. Mechanical ventilation and antibiotic prophylaxis might mask the signs and symp-

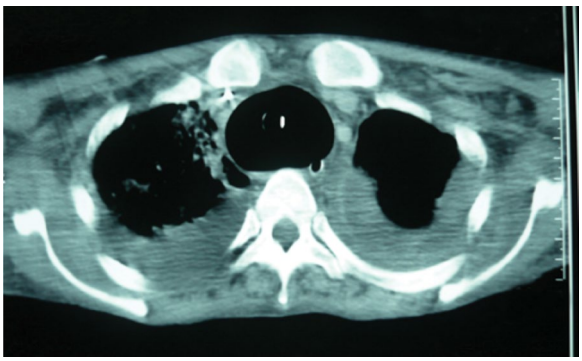

Figure 2: CT scan revealing a posteroanterior tracheal diameter of 6 cm at the level of the second thoracic vertebra; the normal trachea at the same level is about 2.5 cm.

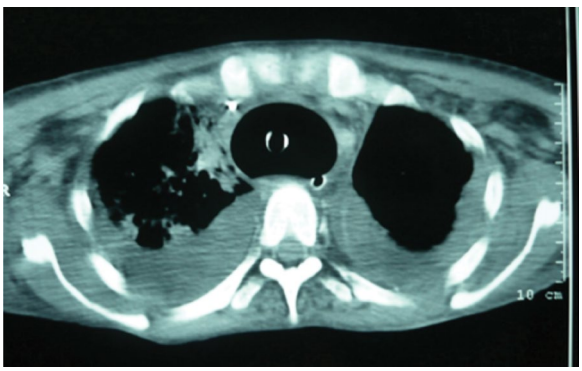

Figure 3: CT scan confirms marked enlargement of the trachea, displacement of the oesophagus, and marked parenchymal infiltrates.

toms and impede early recognition. Bronchoscopy was performed via propagation in the endobronchial tube, but the appearance of the tracheal wall beneath the endobronchial tube lumen might be imperceptible to clinicians.

Endotracheal intubation can traumatize the trachea; this usually results in focal stenosis, but very infrequently may cause focal tracheobronchomegaly. It is not known why some cases result in stenosis and others in tracheobronchomegaly. It is supposed that when trauma or inflammation is restricted to the tracheal mucosa, healing usually occurs with granulation tissue formation and is associated with narrowing of the lumen. However, if trauma is prolonged and exaggerated, as in this case, destruction of the entire tracheal wall and peritracheal tissues might be inevitable. Here, the insult resulted in the loss of structural integrity of the tracheal wall and the formation of focal tracheobronchomegaly.

It is believed that recognition of this condition provides the opportunity of early diagnosis for many diseases including radiological signs of tracheobronchomegaly in hospitalized patients in the intensive care unit.

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