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Duchenne musküler distrofili bir hastada görülen spontan pnömotoraks: Beklenmedik bir mortalite

Spontaneous pneumothorax in a patient with Duchenne muscular dystrophy: an unforeseen mortality

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Duchenne musküler distrofisi bulunan 26 yaşındaki erkek hasta ani başlayan dispne ve sırt ağrısıyla başvurdu. Hastaya başka bir merkezde sol pnömotoraks tanısı konmuştu. Merkezimizde çekilen göğüs grafisinde solda total pnömotoraks saptandı ve mediastenin sağa kaydığı görüldü. Uygulanan tüp torakostomiye karşın sol akciğerde ekspansiyon sağlanamadı. Hastanın genel durumunun uygun olmaması nedeniyle rijit bronkoskopi yapılamadı. On iki saat sonra hastada solunum yetmezliği başladı, arteryel kan gazı değerleri bozulunca hasta yoğun bakım ünitesine alındı. Mekanik ventilasyona karşın kan gazları düzelmeyen hasta kaybedildi.

Anahtar sözcükler: Musküler distrofi/komplikasyon; pnömotoraks/etyoloji.

A 26-year-old male patient with Duchenne muscular dystrophy presented with acute onset of severe dyspnea and back pain. The patient was diagnosed as left pneumothorax in another medical center. Chest X-ray demonstrated total pneumothorax on the left side with a mediastinal shift towards the right side. Tube thoracostomy did not provide pulmonary expansion in the left lung. Rigid bronchoscopy was not performed because of the general medical condition of the patient. Twelve hours later respiratory insufficiency developed and the patient was transferred to the intensive care unit due to the deteriorated arterial blood gas measurements. Despite mechanical ventilation, no improvement on the arterial blood gas levels was achieved and the patient died.

Key words: Muscular dystrophies/complications; pneumothorax/etiology.

Spontaneous pneumothorax (SP) has been reported to ensue in Duchenne muscular dystrophy (DMD) patients with great majority experiencing an uneventful disease course. Opposingly, in this report we render a case whereby the clinical scenario did end with mortality.

CASE REPORT

A 26-year-old young man was admitted to our clinic with acute onset of severe dyspnea and back pain. He had been diagnosed with left pneumothorax in a previous center in the last 24 hours. The previous medical history comprised DMD for the last 15 years -like his 2 year old brother. He weighed only 25 kg and was physically disabled due to severe generalized muscle atrophy. The physical examination of the chest revealed absence of respiratory sounds on the left hemithorax. Laboratory evaluations were unremarkable. Chest X-ray of the patient yielded total pneumothorax on the left side with a mediastinal shift towards the right side (Fig. 1a). Accordingly, an immediate tube thorachostomy was carried out. As the control chest X-ray after the procedure did not reveal any expansion of the left lung, the patient was closely monitorized. Fiberoptic bronchoscopy was performed to demonstrate any other underlying pathology which might have led to the deficiency of pulmonary expansion but no pathology was found. Negative suction with 15 cm H_2O was performed but no change occurred. The computed tomography after tube thoracostomy revealed total pneumothorax on the left hemithorax with compression of the structures on the right side (Fig. 1b). Rigid bronchoscopy under general anesthesia was planned but it

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was thought that medical condition of the patient was not suitable for it. Twelve hours later he started to have respiratory insufficiency and due to the deteriorated arterial blood gas measurements (PO_2: 70 mmHg, PCO_2: 39 mmHg) he was transferred to the intensive care unit (ICU).

Thereafter, as the arterial blood gas levels worsened (PO_2: 41 mmHg, PCO_2: 62 mmHg, pH:7.3) and the respiratory rate increased (38/min), mechanical ventilation was commenced. However, despite mechanical ventilation, no improvement on the arterial blood gas levels could be achieved and the heart rate decreased (30/min), at that time there was wide QRS rhythm on ECG and this was considered as pulseless electrical activity that had developed secondary to hypoxia and acidosis. Then the patient died despite a 45 minute cardio-pulmonary resuscitation.

DISCUSSION

The prevalence of spontaneous pneumothorax (SP) in the normal population has been reported to be 5.8 and 16.7/100.000/year among females and males, respectively.[1] Among DMD patients this ratio was found to be elevated up to 18%.[2] Furthermore, it is also known that DMD patients older than 20 years of age are 7.4 times more susceptible than those younger than 20.[2]

The most common presenting complaints of SP in otherwise healthy individuals are dyspnea and chest pain that are usually (87%) experienced at rest.[3] Interestingly, Yamamoto et al[2] have reported that all of their DMD patients were asymptomatic and recovered completely without any sequelae.[2] Opposingly, due to the severe symptomatology and the untoward clinical outcome of our patient, it would be pertinent to remark that the clinical scenario may also turn out to be an impasse.

The treatment of SP consists of four basic principles: These are; evacuation of the intrapleural air, allowing re-expansion of the lung, to facilitate pleural healing and diminishing the probability of recurrence.[4] Observation and needle aspiration are the two ways of treatment in small degrees, whereas tube thoracostomy must be the first choice in mild-high degrees of pneumothorax. If re-expansion does not occur or massive air leaks persist, negative suction may be performed. On the other hand, the tracheobronchial tree must be examined with rigid or fiberoptic bronchoscopy. The control of the air leaks via thoracotomy or video assisted thoracoscopic surgery is the most effective way if there is no response to other procedures.

The two major risk factors for the development of SP in DMD patients have been reported to be; age over 20 years and body weight less than 30 kg.[2] Both of these factors were present in our patient and we think that they did contribute since the diaphragm and the accessory respiratory muscles of our patient have conceivably atrophied and were less functioning. It is also known that familial predisposition exists for SP patients.[5] Therefore keeping in mind the increased susceptibility of the other sibling -more than the other DMD patients- we have informed the family about the relevant clinical symptoms and signs along with an algorithm of simple management.

Overall, we imply that the concomitance of DMD and pneumothorax –where both disorders are known to be inherited- can end up with mortality because of an excessive pulmonary insufficiency due to this association. When pneumothorax occurs in patients with mus-

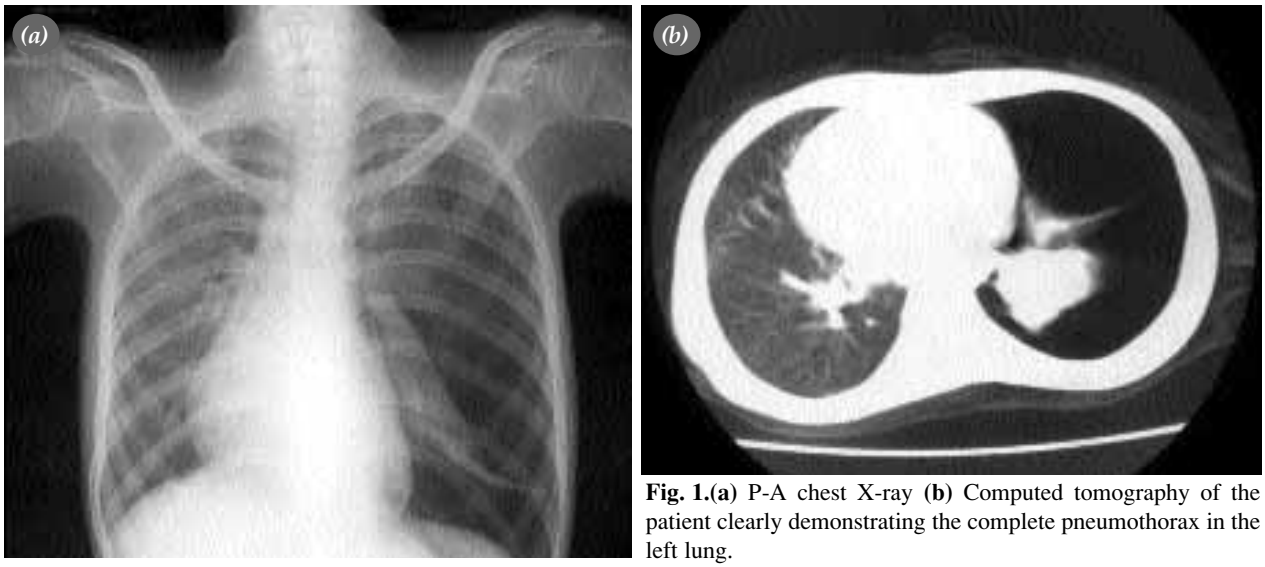

Fig. 1.(a) P-A chest X-ray **(b)** Computed tomography of the patient clearly demonstrating the complete pneumothorax in the left lung.

cular disorders, a great care should be taken promptly, avoiding an unforeseen mortality. The family members of such patients should be warned against the relevant symptomatology and the clinicians should be vigilant against such an adversity.

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