A Rare Complication of Myasthenia Gravis : Pulmonary Hypertension

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Although myasthenia gravis is frequently mentioned in standard textbooks and journal articles as a rare cause for pulmonary hypertension, only one case report actually has been found in the literature. The case described in this report is the first documented case of compansated myasthenia gravis manifesting itself as irreversible pulmonary hypertension. [Journal of Turgut Özal Medical Center 1998;5(1):71-72]

Key Words: *Myasthenia gravis, pulmonary hypertension*

Myastenia gravisin nadir bir komplikasyonu : pulmoner hipertansiyon

Myastenia gravis, pulmoner hipertansiyonun nadir bir sebebi olarak textbook ve makalelerde sıklıkla belirtilmesine rağmen, literatürde sadece bir vaka sunumu bulunmaktadır. Bu yazıda bahsedilen vaka, irreverzibl pulmoner hipertansiyon gösteren kompanse myastenia gravisli ilk dökümante edilmiş olgudur. [Turgut Özal Tıp Merkezi Dergisi 1998;5(1):71-72]

Anahtar Kelimeler: *Myastenia gravis, pulmoner hipertansiyon*

Chronic neuromuscular disorders such as poliomyelitis, Guillian-Barre syndrome and myasthenia gravis (MG) have been documented and reported to be rare causes of acute and chronic neuromuscular paralysis that may be associated with respiratory failure (1-3).

MG is a neuromuscular disorder characterized by weakness and fatigubility of skeletal muscles. The underlying defect is decrease in the number of available acetylcholine receptors at neuromuscular junctions due to an antibody-mediated autoimmune attack. Respiratory muscle involvement may cause respiratory insufficiency. Hypoventilation resulting from respiratory muscle weakness leads invariably alveolar hypoxia. Hypoxia, especially when associated with acidosis may cause pulmonary hypertension (4,5). Chronic hypoxia causes irreversible changes in pulmonary vascular bed.

CASE REPORT

A 40-year-old women, known to have MG for 7 years, was admitted to the hospital for easy fatigubility, weakness, and dypnea both at rest and exertion. Physical examination revealed cyanosis and a chronically ill appearance. Her blood pressure was 140/90 mmHg, heart rate was 90 beats per minute, regular and respiratory rate was 16 per minute. Chest examination showed diminished breath sounds bilaterally. S2 was accentuated on the second left intercostal space and there was 2°/6 holosystolic murmur best heard on the left side of the sternum. Liver was 3 cm palpable. There was minimal pretibial edema bilaterally. EKG revealed right axis deviation and p pulmonale. Telecardiography showed increased cardiothorasic ratio and prominent pulmonary vascular marking. Pulmonary function testing showed minimal restriction. Echocardiographic examination

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disclosed normal left ventricular systolic function with enlarged right ventricular chamber. There was first degree tricuspid regurgitation. Pulmonary pressure of 45 mm Hg was calculated from tricuspid regurgitation by doppler echocardiography. No abnormality was found by contrast echocardiographic examination.

Arterial blood gases on room air were as follows: PO_2=78 mm Hg, PCO_2=50 mm Hg, pH=7.33. Hemoglobin level was 16.1 mg/dl; WBC was 5700 /ml. Ventilation-perfusion scanning of the lungs showed no abnormality. Electromyography showed compensated myasthenia gravis.

By the right and left heart catheterization, coronary and pulmonary angiograms were performed. Pulmonary angiogram showed enlarged main pulmonary artery (Figure 1). Coronary angiogram and left ventriculography were normal. The oxygen saturations were as follows; superior vena cava = 39 %, right atrium = 39 %, right ventricle = 81 % and aorta = 80 %. The systemic and pulmonary vascular resistances were 1050 and 650 dynes-sec-cm^{-5}, respectively. The pressures were as follows; right atrium = 8 mm Hg, right ventricle = 55/0/9 mm Hg, pulmonary artery = 55/34/40 mm Hg, left ventricle = 120/0/23 mm Hg, and aorta = 120/60/80 mm Hg. Inhalation of 100% O_2 showed no change in pulmonary artery pressure (50/30/36 mm Hg) and pulmonary vascular resistance (622 dynes-sec-cm^5)

DISCUSSION

Hypoventilation resulting from respiratory muscle weakness leads to alveolar hypoventilation and it is accompanied by alveolar hypoxia. Alveolar oxygen reflexly controls pulmonary vascular resistance. Pulmonary vascular resistance rises when alveolar oxygen pressure decreases (2,6). This type of induced pulmonary hypertension is reversible with treatment and control of underlying cause. But, chronic hypoxia induces irreversible changes in pulmonary vascular tree.

Although myasthenia gravis is frequently mentioned in standard textbooks and journal articles as a rare cause for pulmonary hypertension and right heart failure, to our knowledge there is only one case report in the literature. We thought that pulmonary hypertension in this myasthenia gravis case was due to chronic hypoxia caused by hypoventilation related to respiratory muscle involvement by MG.

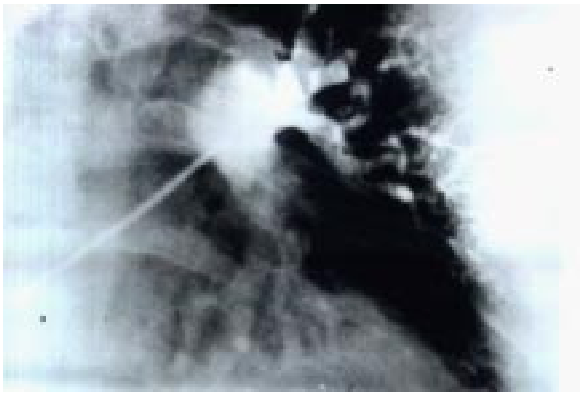

Figure 1. Pulmonary angiogram of the patient

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