

Pickwickian Syndrome

Zubair Ahmed^{1*}, Imran Mehdi Haidry², Muhammad Khurram³, Tanveer Hussain⁴, Abdul Slam⁵ and Arshad Rabbani⁴

¹Post Graduate Resident, Medical Unit II, Benazir Bhutto Hospital, Rawalpindi, Pakistan

²Senior House Officer, Acute Stroke Unit, Blackpool Victoria Hospital, Blackpool, UK

³Professor of Medicine, Rawalpindi Medical University, Rawalpindi, Pakistan

⁴Senior Registrar, Medical Unit II, Benazir Bhutto Hospital, Rawalpindi, Pakistan

⁵House Officer at Benazir Bhutto Hospital, Rawalpindi, Pakistan

***Corresponding Author:** Zubair Ahmed, Post Graduate Resident, Medical Unit II, Benazir Bhutto Hospital, Rawalpindi, Pakistan.

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Abstract

Pickwickian syndrome also known as obesity hypoventilation syndrome (OHS) is characterized by obesity, hypoventilation, day time somnolence and sleep disordered breathing in the absence of alternative neuromuscular, mechanical, or metabolic cause for hypoventilation. Here we describe a patient with OHS who was successfully managed at our Medical Unit.

Keywords: Obesity; Obesity Hypoventilation Syndrome

Introduction

Pickwickian syndrome also known as obesity hypoventilation syndrome (OHS) is characterized by obesity (Body Mass Index-BMI > 30 kg/m²), hypoventilation, day time somnolence and sleep disordered breathing without an alternative neuromuscular, mechanical, or metabolic cause of hypoventilation [1-3]. OHS needs to be differentiated from obstructive sleep apnea (OSA), in which patients have normal alveolar ventilation during wakefulness. OHS and OSA may coexist. 0.3% - 0.40% of general population and 10 - 20% of OSA patients have OHS [4].

OHS lead to hypoxemia and hypercarbia due to failure of adequate ventilation [2,5]. Factors other than obesity are involved in pathogenesis of the disease as not all morbidly obese patients develop OHS. It is considered that defective brain respiratory center function along with obesity cause poor respiratory effort [6]. The name Pickwickian syndrome was coined by Burwell, Robin and co-workers [7]. This was based on clinical features similarities of OHS patients with description of fat boy Joe, in Charles Dicken's book "The Posthumous Papers of the Pickwick Club" which is also called "Pickwick Papers" [8]. Here we describe a patient of Pickwickian syndrome who was managed at our Medical Unit.

Case Report

A 42-year-old female, teacher by profession presented to Outpatient Department of Benazir Bhutto Hospital. She had been known as hypertension and hypothyroidism for one year. Her main complaints were generalized body swelling, shortness of breath and day time sleepiness for 1 year. Other complaints included lethargy, low mood, loud snoring, and frequent awakening during night sleep. She did not complain of fever, cough and sputum. On examination her blood pressure was 140/90 mm Hg, pulse 80/minute and respiratory rate

20/minute. She had generalized body swelling including pedal edema. Her weight was 120 Kg and BMI 44 kg/m². Except for generalized diminished breath sound and loud pulmonary component of second heart sound (P2), systemic examination was unremarkable. Oxygen saturation was 91% on room air. She was admitted for workup and treatment. Clinical diagnosis was made as OHS, pulmonary hypertension, cor-pulmonale in back ground of obesity, hypertension, and hypothyroidism. Patient was administered diuretics, captopril, prophylactic heparin, and inhaled bronchodilators. She was taking 100 microgram of Thyroxin daily that was continued. Baseline investigations were sent along with thyroid profile.

On day two of admission, she initially became drowsy and then lost consciousness. Her blood pressure at that time was 220/120 mm Hg. Random sugar was normal. Her arterial blood gases showed pH 7.05, PO₂ 72 mm Hg, PCO₂ 100 mm Hg, HCO₃ 25 mEq/L, and O₂ saturation 82%. D-dimer testing was negative. Chest X-Ray showed infiltrates at right base. Computed tomography of brain performed at that time was unremarkable. Thyroid profile showed; T3 41.2 ng/dl (normal 58 - 159), T4 4.19 ug/dl (normal 4.87 - 11.72), and TSH 0.78 uIU/ml (normal 0.35 - 4.94). On echocardiography, left ventricular ejection fraction was 60%. Right ventricle was enlarged, right ventricular systolic pressure was 59.99 mm Hg and inferior vena cava was dilated. Details of other investigations are given in table 1.

Investigation	Result	Investigation	Result
Hemoglobin (11.5 to 16.5 g/dl)	8.6 g/dl	Total bilirubin (0.18 to 0.94 mg/dl)	0.6 mg/dl
MCV (78 to 98 fL)	61.4 fL	Alanine aminotransferase (ALT) (10 to 50 U/L)	40 U/L
Peripheral film	Unremarkable	Alkaline phosphatase (40 TO 125 U/L)	109 U/L
WBCs (4 to 11 × 10 ³ /mm ³)	8.3 × 10 ³ /mm ³	Serum Albumin (3.5 to 5.0 g/dl)	3.2 g/dl
Platelets (150 to 350×10 ³ /mm ³)	369 × 10 ³ /mm ³	Prothrombin time (PT)	WNL*
ESR	20 mm at 1 hour	Activated partial thromboplastin time (APTT)	WNL*
Urea	24 mg%	Serum Potassium	3.8 mmol/L
Creatinine	0.9 mg%	Serum Sodium	138 mmol/L
Urine R/E	Unremarkable	Serum Lactic dehydrogenase (LDH)	558 IU/L

Table 1: Hematological and biochemical investigations.

*WNL; Within Normal Limits.

Patient was shifted to Intensive Care Unit, intubated and put on mechanical ventilation for management of respiratory failure. Treatment of pneumonia, and additional illnesses was instituted. She improved subsequently and was gradually weaned of ventilator. Non-invasive ventilation (CPAP) was started. The patient was discharged as she further improved on diuretics, thyroxin, captopril, oral iron, inhaled bronchodilators, and non-invasive ventilation. She was given advice regarding weight reduction as well. Her most recent picture shows improvement.

Discussion

Alveolar hypoventilation is hallmark of OHS. It is characterized by deficient ventilatory process that causes hypercapnia (PaCO₂ > 45 mm Hg). Hypoxemia variably accompanies additionally. In addition to OHS a number of conditions are included in hypoventilation syndromes. These are; central alveolar hypoventilation, deformities of chest cage, neuromuscular disorders, and chronic obstructive airways disease. According to Pierce AM and Brown LK obesity causes restrictive lung disease, that lead to increased breathing work, utilization of oxygen, increased carbon dioxide production and its decreased removal from body [3]. Leptin resistance and sleep disordered ventilation are contributory to these [3]. Hypercapnia in OHS patients cause worsening of daytime sleepiness. It is also associated with increased blood pressure and headaches [1,9]. Hypoxemia causes pulmonary vasoconstriction. Pulmonary hypertension and cor-pulmonale complicate the scenario.

Mainstay of OHS therapy are positive airway pressure therapy, oxygen administration, and obesity management. These are in addition to treatment of accompanying illnesses and complications [10]. Daytime increase in carbon dioxide improve with continuous positive airway pressure (CPAP) and bi-level positive airway pressure (BiPAP). CPAP is generally tried earlier. Those who do not improve with it are administered BiPAP. It entails balancing expiratory and inspiratory positive pressures with clinical, and oxygen saturation improvement. We followed similar plan of management in our patient.

Many of the OHS patients are not appropriately diagnosed earlier and are managed for asthma or chronic obstructive pulmonary disease. OHS patients are generally not diagnosed alone but with diabetes, hypertension, atrial fibrillation, heart failure, chronic obstructive pulmonary disease, and bronchial asthma [3]. We noted similarly.

Marked obesity in OHS patients may cause certain other management issues [11]. These may include drawing blood for investigations, requirement of special blood pressure cuffs, difficulty in getting and interpreting imaging modalities like X-Ray, CT scan and ultrasound [11]. Fortunately we did not face any such problem. Interestingly it is to be kept in mind that Pakistan is among 10 countries who estimated have 3.5 million OHS population. Focus is thus required on management of obesity epidemic [12].

Conclusion

Chest infection and uncontrolled blood pressure were contributors to deterioration in our Pickwickian syndrome patient. Prompt diagnosis, risk factor identification and focused management lead to short and long term improvement.

Bibliography

1. Olson AL and Zwillich. "The obesity hypoventilation syndrome". *American Journal of Medicine* 118.9 (2005): 948-956.
2. Mokhlesi B and Tulaimat A. "Recent advances in obesity hypoventilation syndrome". *Chest* 132.4 (2007): 1322-1336.
3. Pierce AM and Brown LK . "Obesity hypoventilation syndrome: current theories of pathogenesis". *Current Opinion in Pulmonary Medicine* 21.6 (2015): 557-562.
4. Kaw R., *et al.* "Determinants of hypercapnia in obese patients with obstructive sleep apnea: a systematic review and metaanalysis of cohort studies". *Chest* 136.3 (2009): 787-796.
5. "Sleep-related breathing disorders in adults: recommendations for syndrome definition and measurement techniques in clinical research. The Report of an American Academy of Sleep Medicine Task Force". *Sleep* 22.5 (1999): 667-689.
6. Piper AJ and Grunstein RR. "Current perspectives on the obesity hypoventilation syndrome". *Current Opinion in Pulmonary Medicine* 13.6 (2007): 490-496.
7. Burwell CS., *et al.* "Extreme obesity associated with alveolar hypoventilation-Pickwickian syndrome". *American Journal of Medicine* 21.5 (1956): 811-818.
8. Comroe JH. "Frankenstein, Pickwick, and Ondine". *American Review of Respiratory Disease* 111.5 (1975): 689-692.
9. McNicholas WT and Phillipson EA. "Breathing disorders in sleep". Saunders Book company (2001).
10. Branstiter GW., *et al.* "Obesity hypoventilation syndrome in a 12-year-old child requiring therapeutic phlebotomy: case report and review of the literature". *Pediatrics International* 56.6 (2014): e99-e101.
11. Tatusov M., *et al.* "A case report of malignant obesity hypoventilation syndrome: A weighty problem in our ICUs". *Respiratory Medicine Case Reports* 20 (2017): 38-41.
12. Borel JC., *et al.* "Prevalence of obesity hypoventilation syndrome in ambulatory obese patients attending pathology laboratories". *Respirology* 22.6 (2017): 1190-1198.

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