Case Report:

Pickwickian syndrome in a female farm worker

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Date of submission: 15 June 2014; Date of Publication: 22 October 2014

Abstract:
Obesity-hypoventilation syndrome (OHS), also historically described as the Pickwickian syndrome, consists of the triad of obesity, sleep disordered breathing, and chronic hypercapnia during wakefulness in the absence of other known causes of hypercapnia. Its exact prevalence is unknown, but it has been estimated that 10% to 20% of obese patients with obstructive sleep apnea have hypercapnia. OHS often remains undiagnosed until late in the course of the disease. Early recognition is important because these patients have significant morbidity and mortality. Effective treatment can lead to significant improvement in patient outcomes, underscoring the importance of early diagnosis and early treatment. We describe a case of Pickwickian Syndrome which we came across in an active farm-worker female of 35 years of age from Aurangabad district of Maharashtra.

Keywords: obesity, Pickwickian Syndrome

Introduction

Pickwickian Syndrome or the Obesity Hypoventilation Syndrome (OHS) was named after the fat, red faced boy Joe in Charles Dickens’ The Pickwick Papers and the disease is known since 1850s(1) Before this paper, other report of hypoventilation in obesity had reported by Auchincloss and Cook(2) In 1960 s various further studies and discoveries were made which led to the distinction between obstructive sleep apnoea and sleep hypoventilation(3) Obesity hypoventilation syndrome is defined as the combination of obesity (body mass index above 30 kg/m2), hypoxia (falling oxygen levels in blood) during sleep, and hypercapnia (increased blood carbon dioxide levels) during the day, resulting from hypoventilation.

Illustrative Case

A 35 year old female farm worker from Aurangabad district of Maharashtra came with complaints of swelling over face and legs, pains all over body and excessive sleepiness during daytime. On examination, morbid obesity, sleeping and snoring while being examined, was unable to tell her full name in one go without in between sleep. Her height was 153 cm and weight was 98.5 kg giving BMI of 42.07. There were no upper respiratory obstructive causes or nasal polyps. Hypoventilation was clinically obvious and hypercapnea, hypoxia were confirmed by arterial blood gases and pulse oximetry respectively. There was a history of only one year of development of all these symptons and she was not so obese before one and a half year and was hard working female farmer which eagerea us to report this case.
Classification - There are two subtypes of OHS, depending upon the nature of disordered breathing detected on further sleep and other laboratory investigations.

Type I OHS - There is the occurrence of 5 or more episodes of apnoea or hypopnea or respiratory related arousals per hour (apnea-hypopnea index) during sleep. Out of all OHS, about 90% consists of this category.

Type II OHS - This type requires a rise of CO2 levels by 10 mmHg or more after sleep as compared to awake measurements and overnight drops in Oxygen levels without simultaneous apnea or hypopnea (7, 9). These people form 10% of all OHS.

Epidemiology

There are no figures available for incidence or prevalence, mainly because the condition has been poorly defined in the past and often confused with obstructive sleep apnoea (OSA). It has been estimated that approximately 10-20% of patients with OSA have obesity hypoventilation syndrome (OHS). [9] Risk factors mirror that for obesity. Tonsillar hypertrophy is an aggravating factor in children. The peak ages of presentation are 5-7 years and adolescence, [10] although increased awareness of the condition means that more and more cases are being diagnosed in adults. [8] The exact prevalence of obesity hypoventilation syndrome is unknown, and it is thought that many people with symptoms of OHS have not been diagnosed. [7] About a third of all people with morbid obesity (a body mass index exceeding 40 kg/m2) have elevated carbon dioxide levels in the blood. [8] When examining groups of people with obstructive sleep apnea, researchers have found that 10–20% of them meet the criteria for OHS as well. The risk of OHS is much higher in those with more severe obesity, i.e. a body mass index (BMI) of 40 kg/m2 or higher. It is twice as common in men compared to women. The average age at diagnosis is 52. American Black people are more likely to be obese than American whites, and are therefore more likely to develop OHS, but obese Asians are more likely than people of other ethnicities to have OHS at a lower BMI as a result of physical characteristics. [8] Obesity and OHS are more commonly reported in the United States, where obesity is more common, than in other countries. [8]

Signs and symptoms

Most people with obesity hypoventilation syndrome have concurrent obstructive sleep apnea, a condition characterized by snoring, brief episodes of apnea (cessation of breathing) during the night, interrupted sleep and excessive daytime sleepiness. In OHS, sleepiness may be worsened by elevated blood levels of carbon dioxide, which causes drowsiness ("CO2 narcosis"). Other symptoms present in both conditions are depression, and hypertension (high blood pressure) that is difficult to control with medication. [7] The high carbon dioxide can also cause headaches, which tend to be worse in the morning. [10] The low oxygen level leads to excessive strain on the right side of the heart, known as cor pulmonale. [7] Symptoms of this disorder occur because the heart has difficulty pumping blood from the body through the lungs. Fluid may therefore accumulate in the skin of the legs in the form of edema (swelling), and in the abdominal cavity in the form of ascites; decreased exercise tolerance and exertional chest pain may occur. On physical examination, characteristic findings are the presence of a raised jugular venous pressure, a palpable parasternal heave, a heart murmur due to blood leaking through the tricuspid
valve, hepatomegaly (an enlarged liver), ascites and leg edema.[11] Cor pulmonale occurs in about a third of all people with OHS.[8]

**Diagnosis**

Obesity hypoventilation syndrome (OHS) cannot be diagnosed on history and examination alone but requires the demonstration of daytime hypercapnia.[12]

**Diagnostic criteria for OHS**

- Body Mass Index \(\geq 30 \text{ kg/m}^2\).
- Daytime PaCO\(_2\) >45 mm Hg.
- Associated sleep-related breathing disorder (sleep apnoea-hypopnoea syndrome or sleep hypoventilation, or both).
- Absence of other known causes of hypoventilation.

To distinguish various subtypes, polysomnography is required. This usually requires brief admission to a hospital with a specialized sleep medicine department where a number of different measurements are conducted while the subject is asleep; this includes electroencephalography (electronic registration of electrical activity in the brain), electrocardiography (same for electrical activity in the heart), pulse oximetry (measurement of oxygen levels) and often other modalities.[7] Blood tests are also recommended for the identification of hypothyroidism and polycythemia.[7][8]

**Management**[8][9][12]

- A return to normal bodyweight is the mainstay of treatment. Unfortunately, although they may lose weight initially, many patients are non-compliant with dietary restriction in the long term. They are furthermore restricted from increasing their physical activity due to pulmonary symptoms. Bariatric surgery may be required in severe cases.
- Continuous positive airways pressure (CPAP) is more helpful in obstructive sleep apnoea (OSA), where as patients with obesity hypoventilation syndrome (OHS) usually need assisted ventilation which may need to be supplemented by oxygen.
- The inability of these patients to increase their ventilatory capacity should be borne in mind during their management (eg when they are subjected to hospital procedures which may lead to hypercapnia).[11]
- Treat any concomitant OSA, asthma or COPD as appropriate.

In people with stable OHS, the most important treatment is weight loss—by diet, through exercise, with medication, or sometimes weight loss surgery (bariatric surgery). This has been shown to improve the symptoms of OHS and resolution of the high carbon dioxide levels. Weight loss may take a long time and is not always successful.[7] Bariatric surgery is avoided if possible, given the high rate of complications, but may be considered if other treatment modalities are ineffective in improving oxygen levels and symptoms.[8] If the symptoms are significant, nighttime positive airway pressure (PAP) treatment is tried; this involves the use of a machine to assist with breathing. PAP exists in various forms, and the ideal strategy is uncertain. Some medications have been tried to stimulate breathing or correct underlying abnormalities; their benefit is again uncertain.[8]
While many people with obesity hypoventilation syndrome are cared for on an outpatient basis, some deteriorate suddenly and when admitted to hospital may show severe abnormalities such as markedly deranged blood acidity (pH<7.25) or depressed level of consciousness due to very high carbon dioxide levels. On occasions, admission to an intensive care unit with intubation and mechanical ventilation is necessary. Otherwise, "bi-level" positive airway pressure is commonly used to stabilize the patient, followed by conventional treatment.

References


