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CASE REPORT

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Blindness caused by idiopathic intracranial hypertension in the course of the obesity-hypoventilation syndrome – case report

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ABSTRACT

A woman with morbid obesity with severe form of the obesity hypoventilation (OH) syndrome with multiple obstructive sleep apneas (OSA) and hypopneas resulting in extremely low and long-lasting nocturnal arterial oxygen desaturations presented with developing over few weeks blindness. Increases of cerebrospinal fluid pressure during sleep along with obstructive sleep apneas were observed. Idiopathic intracranial hypertension (IIH) with papilloedema was diagnosed. Noninvasive ventilation was ineffective in reversing the consequences of papilloedema despite of reversion of sleep breathing disturbances. The impaired glymphatic system functioning was hypothesized as one of the factors associated with IIH development. Sleep disordered breathing should be searched in obese patients with suddenly developing blurred vision.

KEY WORDS: blindness, obesity-hypoventilation syndrome, sleep apnea, idiopathic intracranial hypertension, respiratory failure.

INTRODUCTION

In some obese patients the obesity-hypoventilation (OH) syndrome with extreme sleep hypoxemia and several organ system derangements may develop [1, 2]. Idiopathic intracranial hypertension (IIH) is frequently associated with obesity [3]. Obstructive sleep apnea (OSA) is a syndrome associated with a number of eye manifestations including floppy eyelid syndrome, optic neuropathy, glaucoma or non-artheritic anterior ischemic optic neuropathy [4]. We present a case of young woman who presented with quickly progressive loss of vision and then was diagnosed with bilateral papilloedema in the course of severe OH syndrome.

CASE REPORT

Morbidly obese, with body mass index (BMI) of 51.3 kg/m^2, 44-years-old woman was admitted to the Neurological Department because of blindness that developed over a few weeks. She was previously diagnosed with hypertension, hypertrigliceridemia, hypercholesterolemia, and insulin resistance and has been treated for the diseases mentioned above.

Neurological examination revealed binocular blindness without light perception. The pupils reaction to light was poor. There were no signs of focal lesions in the central nervous system.

Ophthalmoscopy revealed atrophy of the optic nerves, secondary to papilloedema (Figure 1). Head magnetic resonance imaging (MRI) revealed flattening of the pituitary gland, indicating empty sella syndrome. Cerebrospinal fluid (CSF) pressure, as measured during 30-minute lumbar puncture (LP), was increased to 30 mmHg and rised up to 40 mmHg when the patient has fallen asleep during LP and experienced breathing pauses with visible respiratory efforts that ended with snoring. Neither angio-computed tomography (CT) of the head nor MRI of the cervical part of the vertebral column have shown no abnormalities.

After exclusion of other possible etiologies of intracranial hypertension, IIH was diagnosed. The treatment with acetalozamide and dexamethasone were started and appeared to be ineffective.

Sleep breathing disorder was suspected on the basis of observed extreme sleepiness, witnessed sleep apneas and loud irregular snoring. In polysomnography (PSG) study stage N1constituted 4% of total sleep time (TST), stage N2 – 66% of TST, stage N3 – 18% of TST, stage REM – 12% of TST; obstructive sleep apnea/hypopnea index (AHI) was 51/h,

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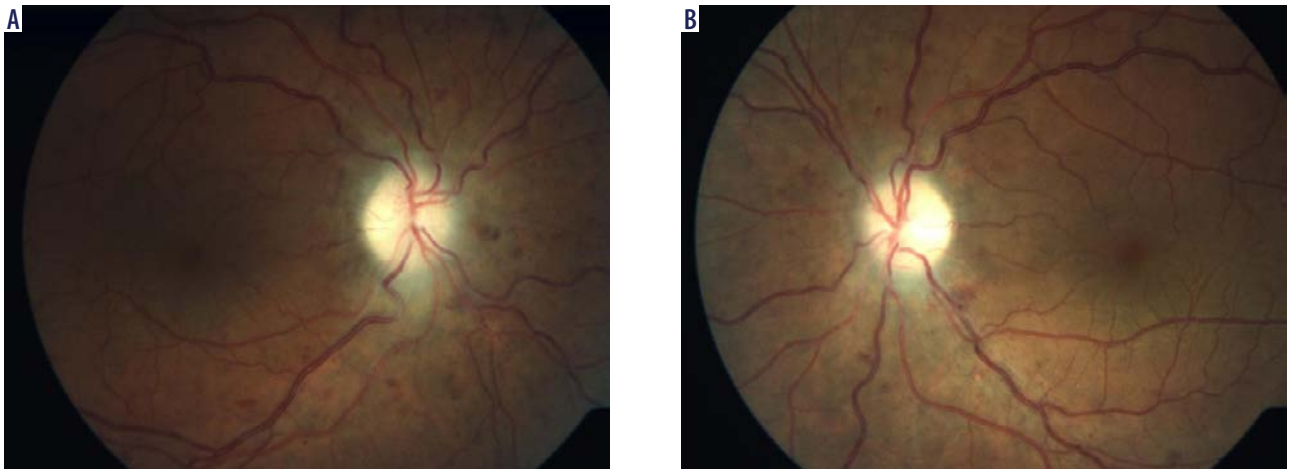

Figure 1. Papilledema

apnea duration varied from 15 s to 120 s, mean arterial oxygen saturation (SaO_2) at the end of sleep apneas/hypopneas was 75%, the minimal values of SaO_2 during sleep were 54% during non-REM sleep and 44% during REM sleep. Daytime arterial blood gases study revealed hypoxemia (PaO_2 8.02 kPa), hypercapnia ($PaCO_2$ 0.65 kPa) and increased bicarbonates (27.1 mmol/l). The results of spirometric studies were nomal.

The OH syndrome was diagnosed and during the following night non-invasive ventilation (NIV) through the nasal mask was introduced. The NIV resulted in the total abolishment of sleep apneas and hypopneas and the increase of mean SaO_2 to 92.2% during non-REM sleep and 89.5% during REM sleep. The tolerance of NIV was good. No vision improvement was observed during two-weeks long stay in-hospital. The patient was discharged home equipped with NIV apparatus. The patient admitted to an irregular use of NIV at home. After 2 months follow up the patient denied further follow-up examinations.

DISCUSSION

Along with the increasing problem of obesity worldwide cerebrovascular consequences of obesity become increasingly important.

In the presented young, morbidly obese woman with metabolic syndrome and severe OH syndrome important increases of intracranial hypertension were observed during obstructive sleep apneas and irreversible blindness developed as a consequence of papilloedema.

Idiopathic intracranial hypertension most frequently occurs in young obese women [5]. The patients may present with symptoms and signs of generalized intracranial hypertension or only with swelling of the optic disc. Papilloedema may be associated with visual disturbances, such as optic field restriction or blindness [6]. In our patient papilloedema was the first, dominating and quickly developing symptom of the disease.

Among the patients with IIH the prevalence of OSA syndrome is high [7]. The retrospective study encompassing the recordings of 156 336 patients with OSA syndrome aged 40 and more the risk of IIH was found to be two-fold greater in yet not-treated OSA patients as compared with persons without OSA [8]. In our patient OSA was present, but taking into account chronic alveolar hypoventilation without the signs of any other disease resulting in chronic daytime hypercapnia and hypoxemia – OH syndrome was diagnosed. Obstructive sleep apneas and hypopneas registered in polysomnography caused profound nocturnal hypoxemia – some episodes lasted extremely long, i.e. up to 2 minutes.

Our observation of marked increases of CSF during sleep, while the patient experienced repetitive episodes of the obstruction of the upper airways allows to link sleep breathing disorder with IIH. The pathogenetic link between sleep disordered breathing and IIH include cerebrovascular changes related to hypoxemia, hypercapnia, increased respiratory effort, and increases of systemic arterial pressure [9]. In terms of pathogenesis however, in literature, retrobulbar blood flow in patients with sleep apnea was not confirmed to be changed [10]. One of the recent hypotheses suggests the involvement of the glymphatic system impairment in the pathogenesis of IIH [11]. The presented case may confirm this hypothesis, as the function of glymphatic system is closely related to the quality of sleep and in the course of OH with numerous sleep apneas and hypopneas there is marked sleep fragmentation.

In our patient NIV was introduced as a method of choice in the treatment of OH. Although breathing disorders during sleep have been prevented by NIV and tolerance of the treatment was excellent home NIV treatment was irregular and no improvement of vision was observed. Presumably, this was caused by long-lasting optic nerve damage in the course of IIH before the initiation of the adequate treatment.

The presented case illustrates the risk of the development of IIH and permanent optic nerve damage in the course of severe OH syndrome in young morbidly obese woman. Clinicians should be aware of a risk of the development of complications of the OH syndrome leading to severe disability. In

obese patients with IIH screening for sleep disordered breathing may be indicated.

DISCLOSURE

Authors declare no conflict of interest.

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