Case Report

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ISSN: 1735-0344 *Tanaffos 2014; 13(2): 43-46*

TANAFFOS

Sleep-Disordered Breathing as Presenting Manifestation of Chiari Type I Malformation: A Case Report

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Received: 18 November 2013
Accepted: 6 February 2014

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Chiari Type I malformation (CM-I) is a rare disorder with displaced cerebellar tonsils through foramen magnum. Here we present a 30-year-old man with severe central and obstructive sleep apneas as presenting manifestations of CM-I. The patient underwent neurosurgery and the follow-up polysomnography revealed the resolution of central apnea while obstructive apnea remained unchanged. Central sleep apnea (CSA) could be associated with an underlying pathology; thus, further investigation is recommended in affected subjects.

Key words: Chiari type I malformation, Sleep disordered breathing, Central sleep apnea

INTRODUCTION

Sleep-disordered breathing consists of apnea and hypopnea, which are repeated breathing pauses and decreased breathing during sleep, respectively. Apneas may be either central or obstructive. Central sleep apnea (CSA) is repeated cessations of breathing during sleep due to loss of respiratory effort while obstructive sleep apnea (OSA) is breathing pauses due to upper airway collapse (1). CSA is less common in general population and is mostly associated with systemic conditions such as heart failure. Other less common etiologies for CSA include living in high altitude, use of opioids and central nervous system conditions (2).

CASE SUMMARIES

A 30-year-old man presented to the sleep clinic with the chief complaint of breathing cessation and gasping during sleep from three years ago. He also complained of headaches, fatigue and sleepiness during daytime, worsened in the past few months. The patient's partner reported loud snoring and several events of apneas during night. He was a civil engineer and was not taking any medications. He denied smoking, drinking or substance or drug abuse. A detailed history was taken and review of systems revealed occasional numbness and tingling in feet and hands. His past medical history and family history were unremarkable. A full physical examination was

performed with no remarkable finding except for a large neck, which was 45cm in circumference and a large tongue. He was overweight with a body mass index of 28.7 kg / m^2.

We performed a sleep study to evaluate the patient's possible sleep disorders. A 5-minute epoch of polysomnogram is depicted in Figure 1. The patient was diagnosed with SDB and as shown in Table 1, 51.6 central apneas per hour were seen. The titration study was performed the night after. During therapy with continuous positive airway pressure (CPAP), the patient's central apnea index deteriorated. Bi-level positive airway pressure (BPAP S / T) with inspiration pressure of 18 cmH_2O and expiration pressure of 14 cmH_2O together with 14 / minute rate had some beneficial effects for controlling apneas. To determine the primary pathology causing central apnea, cardiologic and neurologic workups were performed. Cardiologic workups were normal with ejection fraction of 55%. Brain MRI was done to rule out any nervous system pathology including a brain tumor with compression effect. As seen in Figure 2, cerebellar tonsils were displaced downward through the foramen magnum, consistent with Chiari type I malformation (CM-I).

The patient was referred to a neurosurgeon and underwent posterior decompression surgery. We visited the patient six months after surgery. He reported significant improvements in daytime sleepiness but still experienced apneas after surgery. A second polysomnography was performed to evaluate the patient's apneas. The detailed findings of the first and second sleep studies are shown in Table 1. Titration study was conducted the night after and the remaining apneas and hypopneas completely responded to CPAP device with a pressure of 6 cmH_2O.

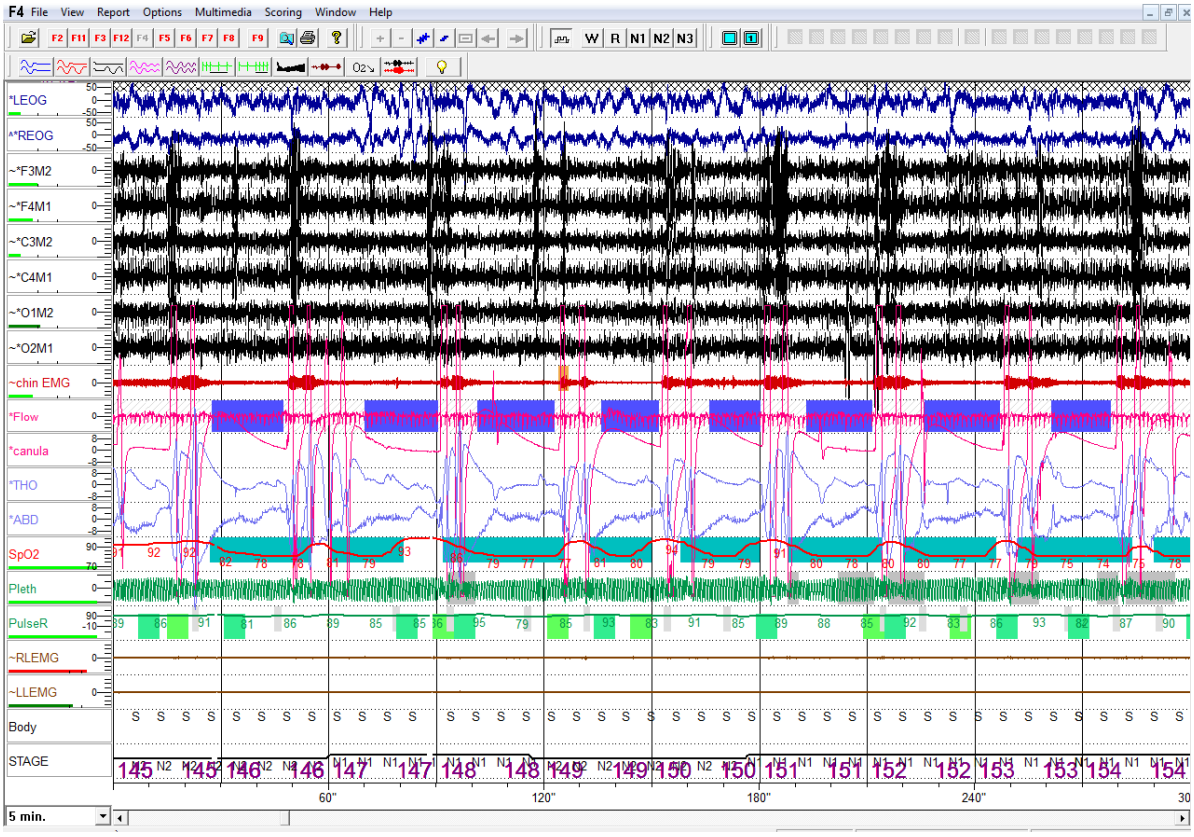

Figure 1. A 5-minute epoch of the patient's polysomnogram. Blue highlights show central apneas.

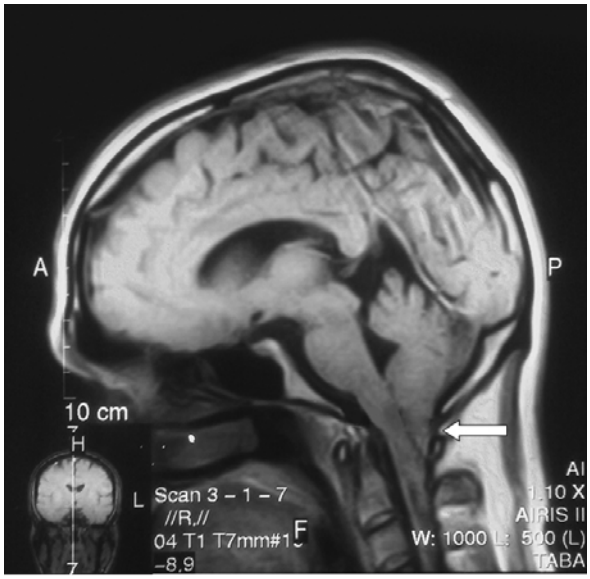

Figure 2. The brain MRI of the patient with SDB and Chiari type I malformation. The white arrow shows herniation of cerebellar tonsils through foramen magnum.

Table 1. The patient's polysomnographic report before and after decompression surgery

	Before surgery	After surgery
Sleep efficiency	83.4%	93.5%
AHI	86.9	37.2
Central apnea index	51.6	0.2
Obstructive apnea index	7.1	3.7
Mixed apneas index	6.6	0
Hypopnea index	21.6	33.3
Baseline oxygen saturation	92%	95%
Oxygen desaturation (>=4%) index	94.2	48.9

AHI: Apnea-hypopnea index

DISCUSSION

Chiari type I malformation (CM-I) is a congenital disorder characterized by abnormally shaped cerebellar tonsils displaced below the level of foramen magnum (3). The prevalence rate of this disorder is estimated to be 0.1 to 0.5%(4). Most cases are asymptomatic and diagnosed with an incidental finding in a brain MRI. The symptoms of CM-I are mainly related to brain stem compression, which may affect control of breathing (5). CSA is a known disorder associated with Chiari type I malformation. A study has shown that SDB disorder is present in 3/4 of patients diagnosed with CM-I (6). Presence of central apnea is regarded as an indication for decompression surgery in otherwise asymptomatic patients with CM-I.

In this case study, we reported a 30-year-old man with typical presentations of SDB. The sleep study revealed more than 50 central apneas per hour of sleep and also considerable number of obstructive apneas. We tried to investigate the underlying pathology of central apneas. The patient's headaches and history of occasional numbness raised a suspicion of central nervous system pathology. Brain imaging showed signs of Chiari type I malformation.

The complete resolution of central apneas after neurosurgery in patients with CM-I has been shown in several studies (5-8). In this case, the central apnea index, as anticipated, decreased from more than 50 to less than 1, while obstructive apneas only slightly decreased after surgery. Thus, there might have been two separate disorders with different pathologies: The central apnea due to a congenital brain malformation and obstructive apnea which could be due to patient's overweight and orofacial anatomy resulting in upper airway collapse during sleep.

Central sleep apnea in young adults is not common. Our patient had more than five central apneas per hour. A precise investigation including cardiologic and neurologic consultation must be carried out to find possible underlying pathologies before prescribing the device. In this case, although obstructive apneas remained after surgery, the patient's SDB was controlled with a low-pressure CPAP device instead of BPAP S/T device.

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