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Case Report

A case report of an obesity hypoventilation syndrome associated with obstructive sleep apnea due to a carotid body paraganglioma

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Abstract

Background and purpose: To the best of our knowledge, the association between an obstructive sleep apnea syndrome (OSAS) due to a neck mass and an obesity hypoventilation syndrome (OHS) has not been reported.

Patients and methods: We report the case of a patient with obesity hypoventilation syndrome (OHS) in whom OSAS caused by a carotid body paraganglioma contributed to recurrent bouts of severe alveolar hypoventilation.

Results and conclusions: The complete surgical excision of the paraganglioma resulted in the cure of the OSAS and contributed to a clear improvement of the clinical symptoms of OHS.

Keywords: Obstructive sleep apnea syndrome; Carotid body paraganglioma; Obesity hypoventilation syndrome

1. Introduction

OHS is characterized by the coexistence of a morbid obesity and hypercapnic respiratory failure [1]. Impaired central drive, ventilatory muscle dysfunction, abnormal load responsiveness and obstructive lung disease contribute to the pathophysiology of OHS [2]. OSAS is frequently associated with OHS, but the relationship between these two diseases remains unclear [3]. It has been suggested that ventilatory responses are reduced in obese patients with OSAS, in contrast to obese patients without OSAS [4]. OSAS is seldom caused by discrete anatomic lesions of the upper airway [5], and the association of OHS and OSAS due to a tumor in the neck has not been described. This study reports on a patient with OHS in whom anatomic obstruction of the airway due to a paraganglioma resulted in sleep-disordered breathing and contributed to recurrent bouts of severe alveolar hypoventilation.

2. Case report

A 69-year-old woman was first seen in the respiratory diseases department in October 1995, after the surgical removal of a meningioma at the base of the skull, because of dyspnea associated with morbid obesity (body mass index [BMI] = 51.2 kg m^{-2}). She had decreased breathing and no sign of right heart failure. Arterial blood gas results were: pH 7.42, Pa_{O_2} 83 mmHg and Pa_{CO_2} 41 mmHg. A ventilation/perfusion lung scan ruled out a pulmonary embolism. Spirometry showed a predominantly obstructive ventilatory deficit: forced expiratory volume in 1 s (FEV_1) = 1040 ml (54% predicted), forced vital capacity (FVC) = 1790 ml (77% predicted) and FEV1/FVC = 58%. Nocturnal oximetry tracing was pathological with minimal Sa_{O_2} (min Sa_{O_2}) 51%, mean nocturnal Sa_{O_2} (mSa_{O_2}) 89.8%, and cumulative time spent below 90% (CT_{90}) 44.1%. However, no apneic episodes were demonstrated at polysomnography (PSG) (Fig. 1). Dietary counseling was given, as well as inhaled β_2-adrenergic agonist therapy. She was then lost for follow-up until January 1997, when she presented at the emergency department with dyspnea and lethargy. Physical examination revealed decreased breath and peripheral edema. BMI was 49.6 kg m^{-2}. Initial arterial

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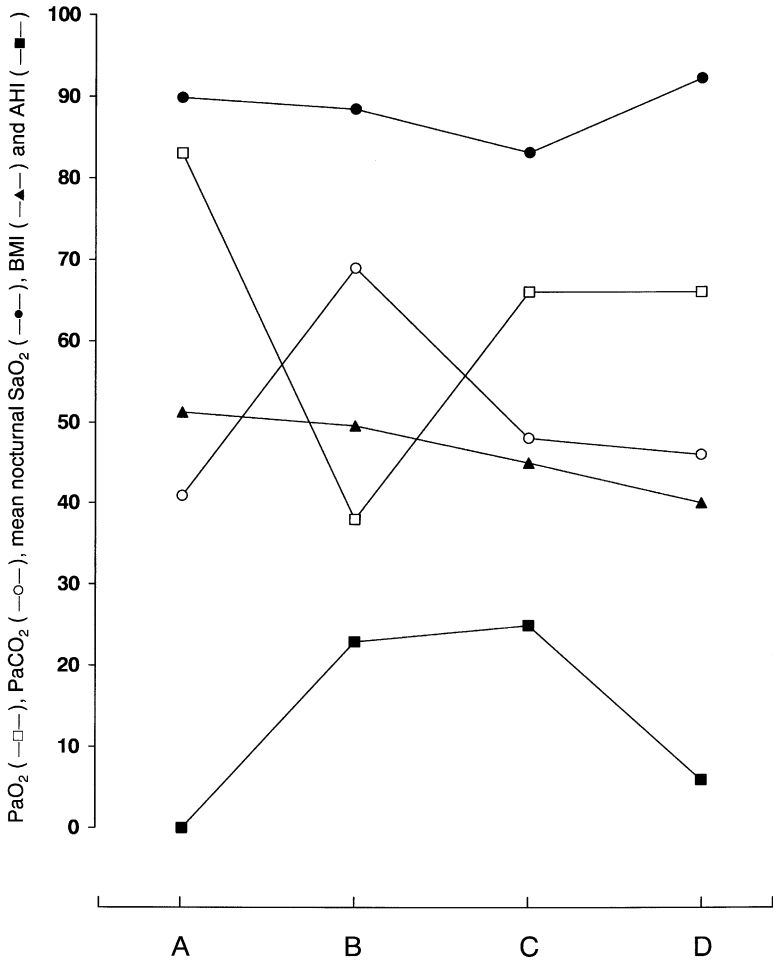

Fig. 1. Values of PaO_2 (mm Hg), $PaCO_2$ (mm Hg), mean nocturnal SaO_2 (%), apnea hypopnea index (AHI, events.h^{-1}), and body mass index (BMI, kg.m^{-2}) at initial check up of the patient (A), during the first episode of respiratory failure (January 1997, B), before (C) and after (D) surgical excision of the paraganglioma.

blood gas results were: pH 7.24, PaO_2 38 mmHg and $PaCO_2$ 69 mmHg. She was intubated and mechanically ventilated. After extubation she remained quite somnolent, with witnessed snoring and persistent hypercapnia. Following stabilization, a PSG was performed. Sleep was severely fragmented and OSAS was observed, with an apnea and hypopnea index (AHI) of 23 events h^{-1}. No central apneic episodes were noted. Nocturnal oximetry data were: min SaO_2 76%, mSaO_2 88.5%, and CT_{90} 71% (Fig. 1). A bi-level positive airway pressure ventilation (Respironics BiPAP®) was applied and the patient was urged to lose weight. She was discharged in March 1997, but because of poor compliance to the Respironics BiPAP® machine was readmitted in September 1997 and March 1998 in severe congestive failure with hypoxemia and hypercapnia. A right cervical mass was discovered during the second admission, but the patient refused any other investigation than a cervical echography, which evaluated the measurement of the mass to be $27 \times 20 \times 40$ mm. A PSG at this time confirmed the persistence of an OSAS (AHI = 23 events h^{-1} with a central apnea index <1 h^{-1}). Nocturnal oximetry data were: min SaO_2 76%, mSaO_2 83.2%, and CT_{90} 99.2% (Fig. 1). The patient was discharged and urged to use the Respironics BiPAP® machine, to agree to dietary counseling and to present for future follow-up of her neck mass. Unfortunately, her lack of observance of these therapeutic objectives led to another respiratory failure in December 1999. The neck mass was still present, but the patient again refused testing. She was lost for follow-up until December 2000, when she was readmitted for a new episode of respiratory failure and once again intubated and mechanically ventilated. BMI was 52.7 kg m^{-2}. The neck mass measured approximately $40 \times 40 \times 40$ mm. Following stabilization of the respiratory failure and weight loss to a BMI of 45.0 kg m^{-2}, the patient gave her consent for etiologic investigations of the neck mass. Arterial blood gas results were: pH 7.43, SaO_2 66 mmHg and SaO_2 48 mmHg (Fig. 1). Contrast enhanced computerized tomography (CT) scans of the neck (Fig. 2) showed an hypervascular mass embedded in the right carotid bifurcation. A carotid arteriogram confirmed the diagnosis of a carotid body tumor (Fig. 3). Hypercapnic and hypoxic ventilatory responsiveness was not studied. The tumor ($60 \times 40 \times 70$ mm) was surgically removed without complication. Pathology

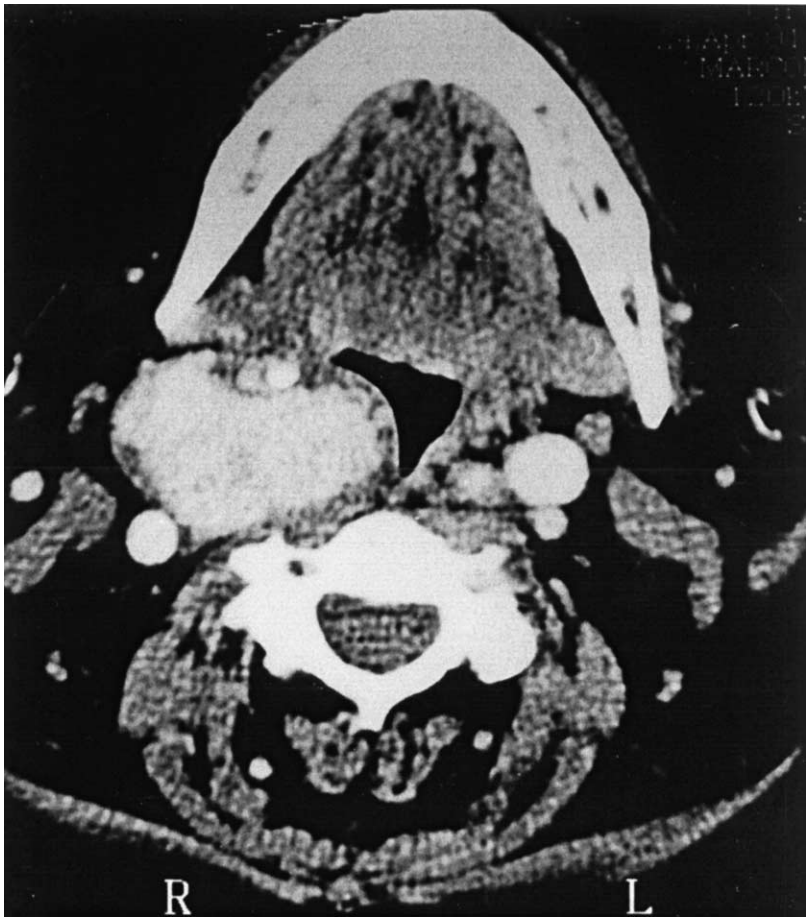

Fig. 2. Contrast enhanced computerized tomography scanning of the neck showing an hypervascular mass located in the right carotid bifurcation and bulging in the airway.

revealed typical cells of a paraganglioma with no sign of malignancy. One month after surgery, arterial blood gas levels on ambient air were: pH 7.42, PaO_2 66 mmHg and $PaCO_2$ 46 mmHg. BMI was 40 kg m^{-2}. A repeat PSG showed an AHI of six events per hour, with a central apnea index <1 h^{-1}. Nocturnal oximetry data were: min SaO_2 70%, mSaO_2 92.3%, and CT_{90} 5.5% (Fig. 1). Despite these persistent nocturnal desaturations, and probably because of the striking clinical improvement after the surgical excision of the paraganglioma, the patient remained reluctant to use of nocturnal non-invasive ventilation. She nevertheless agreed to a periodic follow-up and remains free of any respiratory deterioration 12 months after surgery.

3. Discussion

This case is remarkable for the association of an OHS complicated by recurrent bouts of respiratory failure, associated with an OSAS due to a paraganglioma. A coexistent OSAS may also be determinant for OHS in decreasing ventilatory drive, since it has been shown that ventilatory responsiveness to CO_2 is decreased in obese OSAS patients, in contrast to obese non-OSAS individuals [4]. However, little is known about the association of OSAS and OHS [6] and the link between them remains controversial [3]. It is postulated that OSAS is frequently found in OHS patients, but definitive epidemiological studies supporting the association are lacking. The proportion of patients with OHS in whom OSAS is present may range between 73.8% [7] and 88.5% [6]. In our patient, the occurrence of OSAS and the progress of a carotid body paraganglioma may be linked for the following reasons. First, the neck mass was discovered approximately one year after confirmation of OSAS, which was not disclosed at the first presentation of the patient 15 months before, despite marked nocturnal oximetry abnormalities. Becker et al. [8] have indeed reported on patients with OHS in whom sleep studies showed intermittent desaturation during sleep but no significant OSA. Second, complete surgical excision of the paraganglioma eliminated the OSAS and contributed to a clear improvement of the clinical symptoms of OHS. However, because a thermistor was used for the assessment of the airflow at the nose and mouth, rather than nasal cannulae that are a more sensitive method for the detection of obstructive respiratory events, a persistent airflow obstruction may have remained undetected.

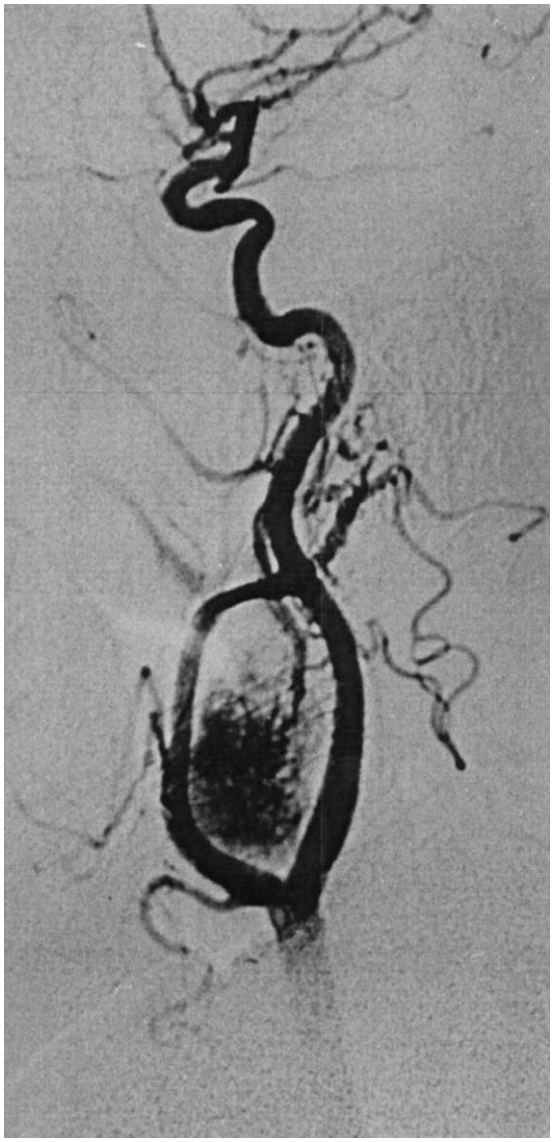

Fig. 3. Right carotid angiography showing a widening and stretching of the internal and external carotid arteries by a highly vascular carotid body tumor.

Weight loss may elicit a significant improvement in respiratory disorders during sleep in OSAS patients [9]. However, the weight loss that occurred between the first episode of respiratory failure in November 1997 and the pre-operative period was comparable to the weight loss observed before and after the surgical removal of the paraganglioma (respectively −9.3% and −11.1% Fig. 1), thus reducing the confounding effect of weight loss on the post-operative improvement of the patient.

To the best of our knowledge, the association of OHS with OSAS caused by a tumor has not been reported. As pointed out by Hoijer [10], OSAS caused by tumor is rare, but probably unrecognized. OSAS secondary to pharyngeal tumor [10], plasmacytoma [5], lipoma [11], carcinoma [12], lymphoma [13], extensive Hunter's disease infiltration of the upper airway [14], and various anatomic abnormalities located anywhere from the nasal vestibule to the larynx [15,16] have been reported. This is the second reported case of an OSAS caused by a carotid body paraganglioma [17].

The patient was not compliant to the management of her disease. She declined a complete evaluation of her cervical mass for more than two years and avoided periodic checkups. Compliance to use of the Respironics BiPAP® machine was very poor because of low tolerance. Kessler et al. have stated that psychological factors related to morbid obesity may predispose patients to underestimate or even be unaware of their disease for a long time [6]. However, as pointed out by Orliaguet et al. [14], inadequate correction of OSAS and/or intolerance to continuous positive airway pressure should prompt a complete evaluation of the upper airways. Severe respiratory failure and "near miss" deaths have been described in patients with OSA [18]. The search for precipitating factors may disclose facial trauma, lower respiratory tract infection, COPD exacerbation or use of pain medication [18]. Since anatomic narrowing of the upper airways plays a major pathophysiological role in the occurrence of OSA, our observation illustrates the importance of a careful examination of the pharynx in any patient with suspected sleep disordered breathing.

In conclusion, the compression of the upper airway by a voluminous carotid paraganglioma led to the development of OSAS and the worsening of OHS in our patient. Both manifestations improved after complete surgical excision of the tumor. This case report emphasizes the necessity of a thorough evaluation of the upper airway in unusual cases of OSAS.

Acknowledgements

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