Patients with sleep apnea often are treated by sleep disorder specialists and are studied in a sleep laboratory. The authors present two such patients who ultimately were found to harbor large benign anterior skull base lesions that caused their obstructive sleep apnea (OSA). The first patient had a massive pituitary tumor and had undergone a tracheostomy before the lesion was diagnosed. The second patient had a large frontoethmoidal encephalocele that was diagnosed at the same time as a recommendation for continuous positive airway pressure therapy was being considered. Such therapy in the presence of an encephalocele can be dangerous and even fatal. Although there are case reports of tumors causing OSA, nearly all of these lesions have been large pharyngeal lipomas (some of which were palpable in the neck during physical examination) or growth hormone–secreting pituitary adenomas. The patients reported here were completely unaware of the presence of these large lesions until imaging studies and/or nasal endoscopy were performed. These cases illustrate the need to perform nasopharyngeal endoscopy and also to obtain magnetic resonance images of the head before prescribing therapy for OSA. Neurosurgeons must be aware that large skull base lesions sometimes present only with OSA.

KEY WORDS • obstructive sleep apnea • skull base tumor • neuroendocrinology

Abbreviations used in this paper: CPAP = continuous positive airway pressure; CSF = cerebrospinal fluid; CT = computerized tomography; ENT = ear, nose, and throat; MR = magnetic resonance; OSA = obstructive sleep apnea.

Case Reports

Case 1

History. This 64-year-old man with OSA was referred for neurosurgical consultation because of a large anterior cranial fossa skull base tumor detected unexpectedly on an MR imaging study. The patient had been under the care of a sleep disorder specialist for more than 10 years. Approximately 1 year before consultation, he had undergone a tracheostomy to manage the OSA and still required it, especially for sleep at night. He had lost his ability to smell approximately 10 years earlier. He had been impotent for at least 12 years and was receiving testosterone therapy. In addition to OSA, he had insulin-dependent diabetes, hypertension, peptic ulcer disease, hypercholesterolemia and depression. He was obese and smoked heavily. A few weeks before being seen by us, he experienced inner ear infections with some buzzing in one ear and was referred for ENT consultation. An MR image of the sinuses and brain was obtained, and a very large anterior cranial fossa skull base tumor was visualized.

Examination. The patient was severely obese (268 pounds, 5'9" tall) and had anosmia. Results of his visual examination were normal except for presbyopia. Blood tests including endocrine evaluation revealed the presence of hypopituitarism; the prolactin level was only 11.5 µg/L. Admission MR images of the brain (Fig. 1) revealed the presence of a very large anterior skull base tumor that extended from the subfrontal region down to the palate.

Operation. Initial transnasal biopsy sampling of the lesion was performed, and pathological analysis showed it to be a neuroendocrine tumor. Gross-total resection was attempted via a transpalatal approach. The tumor was found to be relatively avascular, soft, and able to be suctioned; most of it was removed. The final pathology report identified the...
lesion as a nonsecreting pituitary macroadenoma after it failed to stain for any of the pituitary hormones.

Postoperative Course. Radiation therapy was administered postoperatively. The patient recovered well, and his OSA was relieved. His tracheostomy was closed off in the postoperative period.

Case 2

History. This 52-year-old woman was born with a cleft lip, palate, and nose; repair was undertaken soon after her birth. During her teenage years menarche did not occur, and she received hormone therapy that was later stopped because of adverse effects. Since childhood she had been accustomed to drinking large quantities of water and passing large amounts of urine. She was born blind in the right eye, which required enucleation and a prosthesis in later life. She had decreased vision in the left eye. Approximately 10 years before she was referred to our service, she began to experience symptoms of OSA and was evaluated by a sleep disorders specialist. Imaging studies of the head were performed further to evaluate the cleft lip and palate, and a lesion was revealed. The patient was referred to an ENT surgeon, who found a cystic mass in the nasopharynx during nasal endoscopy and referred her to our institution for neurosurgical consultation.

Examination. The patient had scars in the lip and nose in keeping with past surgery. She had a prosthesis in place of the right eye, and she was completely anosmic. Results in the remainder of the neurological examination were normal. The patient had difficulty lying flat and slept in a semire-
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cumbent position. Despite her multiple problems, she had led a fairly normal life, had a college education, and was gainfully employed.

Blood tests including endocrine evaluation revealed the presence of slight hypothyroidism and decreased follicle-stimulating and luteinizing hormone function. Laboratory tests also confirmed chronic diabetes insipidus. Admission CT and MR images of the brain (Fig. 2) revealed the presence of a large frontoethmoidal encephalocele extending from the region of the third ventricle down to the palate. There was a bone defect in the posterior part of the anterior cranial fossa. A CT scan (Fig. 2 lower) with bone windows confirmed the large bone defect in the posterior ethmoid. The presence of a large encephalocele extending from the brain to the region of the palate was clearly seen.

Treatment. The patient was given surgical and nonsurgical options for treatment. Surgical treatment was deferred because of the complexity of the operation, and it was decided that the patient’s OSA would be managed with postural adjustment and careful follow up. The patient was well adjusted to her situation and was very much reassured by having a correct diagnosis. For the first time, she had a satisfactory explanation for her chronic pituitary and hypothalamic insufficiency. If her symptoms worsen, or if CSF leakage or meningitis occurs, surgical treatment will again be considered.

Discussion

Sleep apnea is defined as the temporary cessation of breathing during sleep. It is identified by the absence of airflow past nasal or buccal monitors during sleep. There are three fundamental types, as follows.8 1) Obstructive sleep apnea is defined as the absence of airflow past nasal and buccal recorders despite abdominal and thoracic effort. This may be due to tonsillar or adenoidal hypertrophy, especially in children, or to exogenous obesity in adults, particularly in those with a short, thick neck and hypotonic pharyngeal musculature. 2) Central apnea is defined as the absence of airflow past nasal and buccal monitors because of the absence of thoracic and abdominal respiratory effort. 3) Mixed apnea is defined as the absence of airflow with no respiratory effort (central type) followed by lack of airflow despite the resumption of respiratory effort (obstructive type).

Obstructive sleep apnea is the most common type of sleep apnea. It has a higher prevalence among men (9%) than women (4%) and is typically associated with obesity.4 A patient with OSA can present with many underlying symptoms; the most prominent of them is snoring, which often alternates with periods of silence known as apneic episodes.8 Whereas apnea is due to the complete obstruction of the airway, snoring is caused by the partial obstruction of airflow. The most debilitating symptom of sleep apnea is hypersomnolence, which results from lack of the deep sleep of Stages 3 and 4.4 Individuals with sleep apnea are prone to falling asleep during the day, at work, or while driving, and they can be a danger to themselves and others. Other symptoms of sleep apnea include flailing of the extremities while sleeping, depression, memory loss, impotence, irritable mood, and hypertension.

The patients in both of our cases had received a mis-diagnosis of idiopathic OSA. The patient described in Case 1 was assumed to have OSA, and his obesity and history of smoking were thought to be contributing factors. No structural basis for his OSA was ever sought, and the large skull base tumor was discovered accidentally when imaging studies of his head were performed because he was experiencing ear infections. In retrospect, the ear infections were probably caused by obstruction of the eustachian tube by the large mass. The patient described in Case 2 was also treated for OSA and was considered to be a candidate for CPAP therapy. Her history of repair for congenital cleft lip and palate led to the performance of imaging studies that revealed the presence of a large anterior cranial fossa mass, namely, a frontoethmoidal encephalocele. Nasal endoscopy performed by an ENT surgeon confirmed the presence of a cystic nasopharyngeal mass.

Structural lesions have been reported to cause OSA. Case reports identify parapharyngeal and retropharyngeal lipomas as causative lesions.12,5,9,10 The patient described by Pellanda, et al.,10 who presented with snoring and OSA, was found to have a 5 × 10–cm mobile mass in the right side of the neck under the sternocleidomastoid muscle. The diagnosis was confirmed using MR imaging, and excision of the mass relieved the OSA. Koopmann, et al.,8 described a patient with a submandibular mobile mass in the neck. In their case, a 600-g lipoma was excised from the parapharyngeal area, leading to immediate resolution of OSA. Hockstein, et al.,7 reported the case of a 64-year-old man in whom an incidental finding of a large retropharyngeal lipoma was made on CT scans obtained while he was being evaluated for OSA. Although the diagnosis of lipoma was confirmed by needle biopsy sampling, the lipoma itself was not excised because of comorbid conditions. Aland6 reported on a 36-year-old man who presented with loud, intolerable snoring. Pharyngeal endoscopy and CT scans of the neck demonstrated the presence of an 8 × 4 × 6–cm submucosal posterior pharyngeal lipoma. The lesion was removed transorally, leading to total resolution of the snoring and OSA. Di Girolamo, et al.,7 also reported on a 36-year-old man presenting with OSA who was found to have a submucosal mass on pharyngeal examination that was confirmed to be a retropharyngeal lipoma on CT scans. Excision of the mass resulted in resolution of the snoring and OSA. Abdullah, et al.,1 reported on a 60-year-old man who presented with 30 days of daytime sleepiness. A CT scan of the head and neck demonstrated a 3 × 1 × 7–cm, well-defined lipoma in the right parapharyngeal area at the C1–2 level. No tissue diagnosis was obtained and the patient refused surgery. He was maintained on CPAP therapy.

In two reports multiple cases of growth hormone–secreting pituitary adenomas are described in patients with acromegaly and OSA.11,12 In many instances resection of the tumor and correction of the pituitary hormone levels resulted in disappearance of the sleep apnea. Nevertheless, the mechanism of OSA in acromegaly is mainly associated with hypertrophy of the tongue and the adjacent buccal pharyngeal mucosa. These changes resolve after removal of the growth hormone–secreting adenoma, and the OSA is also relieved.

Posterior fossa tumors have been reported to cause sleep apnea. These include a posterior fossa glioma7 and an acoustic neuroma.8 Nevertheless, the cause of the apnea was mainly attributable to compression of the respiratory center in the brainstem, and resection of these tumors resulted in

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decompression of the brainstem and resolution of the sleep apnea. 

We report two cases of large anterior cranial fossa lesions causing OSA. The mechanism is somewhat similar to those in the cases reported in the literature, in that when the patient lies down the lesion obstructs the upper respiratory passages. In the patient described in Case 1, this led to a tracheostomy because the lesion itself was not diagnosed. In the patient described in Case 2, a correct diagnosis was made and she is being treated with postural adjustment and careful follow up.

The lesion in the patient described in Case 2, which is an example of a frontoethmoidal encephalocele, was not treated surgically because the patient had learned to live with her symptoms. Nevertheless, these lesions can be treated surgically when patients present with symptoms such as CSF leaks, meningitis, or progressive neurological deficits. In those circumstances, surgery is best performed via a one-stage craniofacial approach with excision of the devitalized brain, repair of the skull base defect, and correction of hypertelorism or other skull base congenital anomalies. The complications associated with such procedures include CSF leakage, meningitis, and neurological deficits such as blindness.7

Conclusions

Our patients have OSA that is caused by a mass. These cases and those reported in the literature emphasize the need for patients with OSA to undergo imaging studies of the head and neck to detect possible structural lesions before therapy such as tracheostomy or CPAP is instituted. Neurosurgeons must be aware that some skull base lesions present only with OSA.

References


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