Diffuse Idiopathic Spinal Hyperostosis: A Rare Cause of Severe Obstructive Sleep Apnea

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Abstract
Most causes of Obstructive Sleep Apnea (OSA) arise from obstruction of soft tissue structures in the upper airway, combined with a loss in muscle tone during sleep. We describe an unusual case of OSA resulting from Diffuse Idiopathic Spinal Hyperostosis (DISH), in which ossification of the anterior longitudinal ligament presents as an extensive bulge in the posterior pharyngeal wall. This led to narrowing of the upper airway, contributing to upper airway obstruction and to symptoms of OSA.

ABBREVIATIONS
OSA: Obstructive Sleep Apnea; DISH: Diffuse Idiopathic Spinal Hyperostosis; PAP: Positive Airway Pressure

INTRODUCTION
Most causes of Obstructive Sleep Apnea (OSA) arise from obstruction of soft tissue structures in the upper airway, combined with a loss in muscle tone during sleep. We describe an unusual case of OSA resulting from Diffuse Idiopathic Spinal Hyperostosis (DISH), in which ossification of the anterior longitudinal ligament presents as an extensive bulge in the posterior pharyngeal wall. This led to narrowing of the upper airway, contributing to upper airway obstruction and to symptoms of OSA.

CASE PRESENTATION
A sixty-nine (69) year old gentleman presented with symptoms of loud snoring, excessive daytime somnolence, unrefreshed sleep, with apnoeic and choking episodes at night. He denied having any dysphagia, hoarseness, stridor or dyspnea.

He had a history of hypertension and hyperlipidemia. On physical examination, he weighed 68kg, height was 160cm, BMI was 26.6 kg/m² and neck circumference was 41cm. There was mild inferior turbinate hypertrophy with no significant septal deviation. Oropharyngeal examination revealed a smooth mucosal lined mass in the oropharynx, arising from the posterior pharyngeal wall (Figure 1). The modified Mallampati score was 1 with no notable obstruction of the airway from any elongated soft palate or uvula. Fibreoptic nasoendoscopy showed a prominent bulge along the posterior pharyngeal wall that extended from the nasopharynx to the base of tongue.

Polysomnography showed an Apnea Hypopnea Index (AHI) of 72.4/hour with a significant hypoxia nadir reaching 67%.

Computed Tomography (CT) scan (Figure 2) of the neck revealed bulky ossification of the anterior longitudinal ligament (ALL) from C1 to T1 level, consistent with DISH. Further history taking found that he had long standing neck stiffness that was not accompanied by any neurological symptoms. He was reviewed by our orthopaedic colleagues to discuss surgical options after Magnetic Resonance Imaging (MRI) of the cervical spine showed evidence of mild cord compression. Conservative management without surgery was opted for, in view of a normal neurological examination with no clinical evidence of myelopathy.

Moreover, he showed significant improvement after being started on Positive Airway Pressure (PAP) therapy for severe OSA. Subsequent follow up over the next 3 years showed an AHI range of 2.5 to 3.5 with good compliance and resolution of his initial symptoms.

Figure 1 An intraoral view of the patient with DISH. A posterior pharyngeal wall bulge (O) is seen abutting the uvula (U) and soft palate.
DISCUSSION

Diffuse Idiopathic Skeletal Hyperostosis (DISH), also known as Forestier’s syndrome is a disease characterized by calcification and ossification in the soft tissue, with predilection for the anterior longitudinal ligament. It is a rare condition that can significantly affect the aerodigestive tract. Common symptoms reported include dysphagia and hoarseness [1]. One study found a more acute presentation in which a patient presented with dyspnoea and stridor. The patient required a tracheostomy after being found to have a pharyngeal perforation, with paralysis of vocal cords and oedematous supraglottic airway obstruction [2].

There was no study identified in existing literature that describes symptoms of obstructive sleep apnea being the first presentation symptoms for patients with DISH. Instead, patients seen in the otolaryngology department typically present with symptoms such as dysphagia and are then diagnosed as having DISH. Further evaluation then shows evidence of OSA that may range from mild to severe in nature. There is limited elaboration in the existing literature regarding the extent of success with CPAP treatment [1,2].

One should consider DISH in the differential diagnosis of airway obstruction in OSA when examination of the oropharynx reveals a bulge in the posterior wall, a finding that is consistent amongst these patients. It also further reinforces the importance of upper airway evaluation by an otolaryngologist or by the sleep physician when evaluating patients with symptoms of OSA. CT is the modality of choice to confirm this diagnosis. Characteristic radiological findings include linear bone formation along the anterolateral aspect of the thoracic spine with bumpy contours, subjacent radiolucency, as well as irregular bony pointed excrescences at the cervical and lumbar vertebral margins [3].

In the process of clinical evaluation, the sleep physicians should also consider other non anatomical factors that may contribute to the patient’s sleep apnea. A detailed study of the polysomnographic data is needed to exclude other mechanisms unrelated to upper airway narrowing.

The decision on management should depend on patient and disease factors. Patient factors may include fitness for surgery, preference or compliance to conservative treatment with PAP. PAP has been shown in some cases to completely ameliorate hypoxic episodes in patients with pharyngeal tumours [4]. Disease factors may include the extent of disease, as well as other symptomatology such as myelopathy for which surgery may also be beneficial for. Recurrence after surgery has been previously reported and should also be factored into the decision making process for patients keen on surgical intervention [5]. In this patient, his OSA was controlled with PAP. He also had no dysphagia or neurological symptoms to warrant surgery. Hence, the decision was made for non-surgical management.

Patients with DISH are likely to present outside the otolaryngology department with symptoms of neck pain, stiffness and other neurological symptoms. Upon diagnosis, clinicians should consider a referral to a sleep clinician to evaluate the distinct possibility of OSA in these patients.

REFERENCES


Cite this article