

# Cervical Sympathetic Neurofibroma Masquerading as Obstructive Sleep Apnea Syndrome

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## ABSTRACT

Obstructive sleep apnea is a chronic condition characterized by frequent episodes of upper airway collapse during sleep. Collapsibility can be increased by underlying anatomic alterations and/or disturbances in upper airway, neuromuscular control, or both, which play key roles in the pathogenesis of obstructive sleep apnea. Neurofibromas of the parapharyngeal space are the second most commonly encountered primary tumor of the nerve sheath origin. A parapharyngeal neurofibroma of the cervical sympathetic chain, presenting as obstructive sleep apnea with all the features mimicking that condition is reported here for its rarity in modern clinical practice. A transcervical approach was adopted to excise the tumor in toto, following which patient was completely relieved of the symptoms, especially those of respiratory distress and features of OSAS.

**Keywords:** Parapharyngeal tumor, Cervical neurofibroma, Obstructive sleep apnea, Transcervical approach, Cervical sympathetic chain, Horner's syndrome.

## INTRODUCTION

Obstructive sleep apnea syndrome (OSAS) is a chronic condition characterized by frequent episodes of upper airway collapse during sleep.<sup>1-3</sup> Obstructive sleep apnea is insidious and patients are often unaware of the associated symptoms. There are several risk factors like obesity, sex, age and hereditary factors, of which obesity is the major risk factor.<sup>1,2</sup> Collapsibility of the airway can be increased by underlying anatomic alterations and/or disturbances in upper airway, neuromuscular control, or both, which play key roles in the pathogenesis of obstructive sleep apnea. Obesity narrows the upper airway secondary to deposition of adipose tissue in the parapharyngeal space. Observational and experimental evidence shows that OSA can predispose to the development of systemic hypertension, cardiovascular disease and abnormalities in glucose metabolism.<sup>2</sup> However, the sheer mass effect of the tumor leading to symptoms of OSAS is virtually rare in clinical practice.

As such primary tumors of the parapharyngeal space are rare and account for 0.5% of all head and neck tumors. Of these 70 to 80% are benign and 20 to 30% are malignant.<sup>4</sup> Most parapharyngeal space tumors are of salivary gland followed by neurogenic tumors.<sup>5</sup> Next to schwannomas, neurofibromas are the second most commonly encountered primary tumor of the nerve sheath origin. Careful preoperative planning, advanced surgical techniques and vigilant postoperative care result in minimal morbidity and resolution of tumor symptomatology.

Although complete surgical resection is ideal for all parapharyngeal tumors, the dilemma of complete vs partial

resection arises when massive size increases the possibility of neurological dysfunction from damage to adjacent cranial nerves and cervical sympathetic fibers.

Neurofibromas arising from cervical sympathetic chain are rare in any population and only few cases have been reported. This article documents the unique case of a cervical sympathetic neurofibroma mimicking as obstructive sleep apnea in a 31-year-old male.

## CASE REPORT

A 31-year-old obese gentleman consulted with a 2 years history of snoring, day time somnolence and extreme fatigability. He also gave history of difficulty in swallowing and breathing for the past 2 months. There was no history of any other specific co-morbid conditions. Patient appeared normal and alert and had normal vital signs. He exhibited all the typical features of OSAS. Subsequently, while investigating the cause and the level of mechanical obstruction in the pharynx, a left-sided mass lesion was detected in the oropharynx on clinical examination. Hence, an investigation protocol was instituted to establish the diagnosis of the space occupying lesion in the left parapharyngeal space.

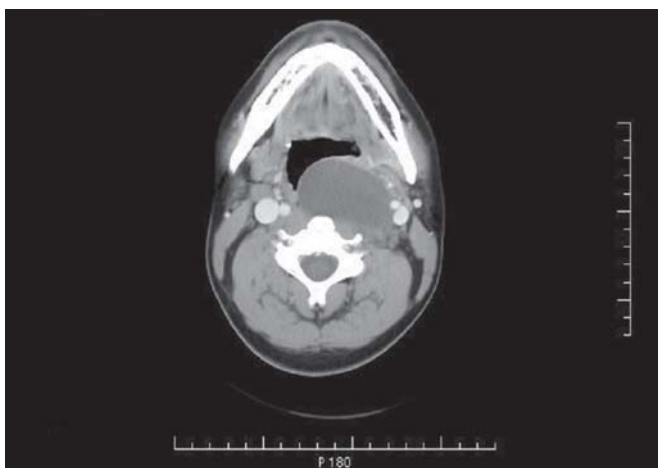
Examination of the oropharynx revealed a swelling in the left side of the posterior wall of the pharynx, behind the left posterior tonsillar pillar, crossing the midline. Mucosa over the mass appeared intact and mobile. Consistency was firm with no palpable thrill or transmitted pulsations. There were no other palpable swellings in the neck or anywhere else in the body. No focal or general neurological deficits were detected.



**Fig. 1:** Laryngeal endoscopy showing tumor almost occluding inlet of larynx (preoperative view)



**Fig. 3:** Intraoperative view of tumor arising from superior cervical ganglion



**Fig. 2:** CT scan view of circumscribed tumor with displaced carotids anteromedially



**Fig. 4:** Excised tumor *in toto*

Trachea was in the midline. Carotid pulses were equal on both sides and there was no bruit or thrill. Indirect laryngoscopy and posterior rhinoscopy were attempted but proved inconclusive. Laryngeal endoscopy (Fig. 1) showed the mass occluding a major portion of the laryngeal inlet with mobile vocal cords bilaterally.

CT scans with contrast (Fig. 2) showed a mass extending from C2-C5 levels displacing the carotids anteromedially and internal jugular vein posteriorly. MRI scans revealed a hyperintense mass in the left parapharyngeal space. On angiography, there was no significant enhancement of the tumor.

A transcervical surgical approach was planned with an option of angle mandibulotomy, if needed, for better exposure of the tumor (Fig. 3). An elective tracheostomy was performed to secure the airway. Cervical incision was used to approach the tumor. Subplatysmal flaps were raised and retracted for exposure. The great vessels were first isolated and retracted. Stylomandibular ligament was cut for ease of access and mandible was retracted anteriorly to obtain adequate exposure.

The mass was seen arising from the cervical sympathetic chain with no involvement of the vagus, hypoglossal or spinal accessory nerves. The mass was excised *in toto* (Fig. 4) and sent for histopathological examination, which was consistent with neurofibroma. He had an uneventful postoperative course. He was decannulated on the third postoperative day. Patient developed transient left Horner's syndrome which resolved by the third postoperative day.

Subsequently, a check laryngeal endoscopy and CT scans were done to assess the operated site, after a period of 3 weeks. The findings were very encouraging as there was neither evidence of any residual disease nor any functional deficit. The external scar too had healed well. Patient is completely asymptomatic as per the last follow-up (Figs 5 to 7).

## DISCUSSION

An apnea is defined as the complete cessation of airflow for at least 10 seconds. The causes of apnea can be classified as central, obstructive (occlusion of oropharyngeal airway) and mixed (central component followed by obstructive component).<sup>1,3</sup> A



Fig. 5: Postoperative endolaryngeal view of laryngeal inlet

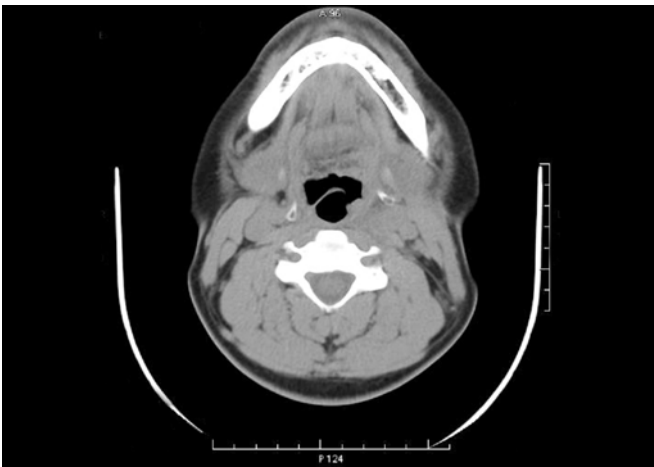


Fig. 6: Postoperative CT scan view showing complete tumor clearance



Fig. 7: Three weeks postoperatively well-healed scar

parapharyngeal tumor as such is a rare entity. There have been no case reports on cervical sympathetic neurofibroma presenting with features of obstructive sleep apnea.

Hughes reviewed 172 patients with parapharyngeal space neoplasm and found that pleomorphic adenoma was the most common neoplasm (40%), followed by paraganglioma (20%),

neurogenic tumor (14%), malignant salivary gland tumor (13%), miscellaneous malignant (7%) and benign tumors (6%).<sup>6</sup> Peripheral nerve sheath tumors of head and neck regions comprise a very small percent but an important section of all neoplastic lesions in this area. They are a complex array of both benign and malignant tumors, including neurofibroma, schwannoma and malignant peripheral nerve sheath tumors.

Microscopically, neurofibromas are formed by a combined proliferation of all the elements of a peripheral nerve: Axons, Schwann cells, fibroblasts, and (in the plexiform type) perineurial cells. The nuclei of the tumor cells of neurofibroma show a typical fascicular pattern of growth and serpentine shape.<sup>7</sup> Neurofibromas are commonly treated by surgical removal. Surgical procedures in the upper part of the parapharyngeal space, including the nasopharynx and skull base is relatively difficult to approach because of the presence of the carotid artery, internal jugular vein and cranial nerves. Many surgical techniques to approach the upper parapharyngeal space have been reported as follows; cervical approach, transparotid approach, cervical-parotid approach, transoral approach, infratemporal fossa approach, cervical transpharyngeal approach, mandibular osteotomy, mandibular swing approach and transmandibular transpterygoid approach.<sup>8</sup> Transcervical is the most common approach as it allows better access and wider scope to remove the tumor as a whole along with its capsule.<sup>9</sup> The preoperative diagnosis of peripheral nerve sheath tumors and the consideration of differential diagnoses are extremely important if one is to warn the patient about possible neurological sequelae postoperatively, i.e. Horner's syndrome, vocal palsy, etc.

## CONCLUSION

Primary parapharyngeal tumors are rare and located in a complex anatomical region. The clinical presentation of these tumors can be subtle. As in this patient a diagnosis of OSA was made and treatment protocols for the same initiated for a few months prior to the present clinical status. Therefore, radiographic study provides important information to lead to a correct diagnosis and surgical planning. Majority of the tumors are benign with salivary gland neoplasm being the most common tumor. Surgical resection is the mainstay of treatment.<sup>7,8</sup> The choice of surgical approach is guided by the extent of the tumor and the displacement of the great vessels in the neck. However, the final clinching evidence of such lesion is histopathology. One needs to be very vigilant in dealing with such conflicting clinical scenario and adopt the most effective approach in treating such rare lesions.<sup>9-11</sup>

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