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Incidentally Diagnosed Supraglottic Neurofibroma in Adult

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Abstract **Case Report**

Neurogenic tumors of the larynx are extremely rare, especially in adults. Most cases reported to date have been in children with neurofibromatosis. Because of their slow growth and variability in location and size, laryngeal neurofibromas may remain asymptomatic for years or become symptomatic at birth. Symptoms include stridor, hoarseness, dysphagia, dysphonia, globus sensation, dyspnea, hoarseness, and feeding difficulties and are usually progressive. There is little information about these tumors, but knowing this diagnosis is very important, especially for physicians in charge of airway management, and missing it can have fatal consequences. We present a case of a laryngeal neurofibroma that was discovered incidentally in a 36-year-old adult male.

Keywords: Adult, Airway management, Larynx, Neurofibroma.

INTRODUCTION

Neurogenic tumors of the larynx are extremely rare, especially in adults [1]. Laryngeal neurofibromas are less common than schwannomas and may be solitary and non-syndromic or, most commonly, associated with neurofibromatosis (NF) type 1 or 2 [2-4]. Most cases reported to date have been in children with neurofibromatosis (also known as von Recklinghausen's disease), predominantly presenting as multiple tumors [5]. Because of their slow growth and variability in location and size, laryngeal neurofibromas may remain asymptomatic for years or become symptomatic at birth. Symptoms include stridor, hoarseness, dysphagia, dysphonia, globus sensation, dyspnea, hoarseness, and feeding difficulties and are usually progressive [3]. There is little information about these tumors [6], but knowing this diagnosis is very important, especially for physicians in charge of airway management, and missing it can have fatal consequences.

We present a case of a laryngeal neurofibroma that was discovered incidentally in a 36-year-old adult male.

CASE

A 36-year-old man fell 3 days ago and presented to the emergency department with scalp swelling and dizziness. He also has shortness of breath. He was diagnosed with plexiform neurofibromatosis type II at the age of 10 due to a mass in the chest, forehead, and abdomen, and he underwent right mastoidectomy and parotidectomy for parotid neurofibroma at the age of 20 and lost his hearing. He also had difficulty communicating due to a speech impediment.

The computed tomography (CT) scan of the brain and neck showed no brain damage, but a solitary mass in the left supraglottic area was confirmed.

$PaCO_2$ was 60 mmHg on ABGA. His respirations were short and unstable, with a frequency of 20 to 25 times per minute.

Communication was difficult and he said that his shortness of breath and persistent sputum production started after the last COVID-19 infection one year ago, so it was difficult to determine exactly when the respiratory symptoms started.

After consultation with an otolaryngologist, direct laryngoscopy revealed a large solid mass obstructing more than 80% of the tracheal lumen. It originated from the left posterior supraglottic area. The rest of the larynx appeared normal (Figure 1).

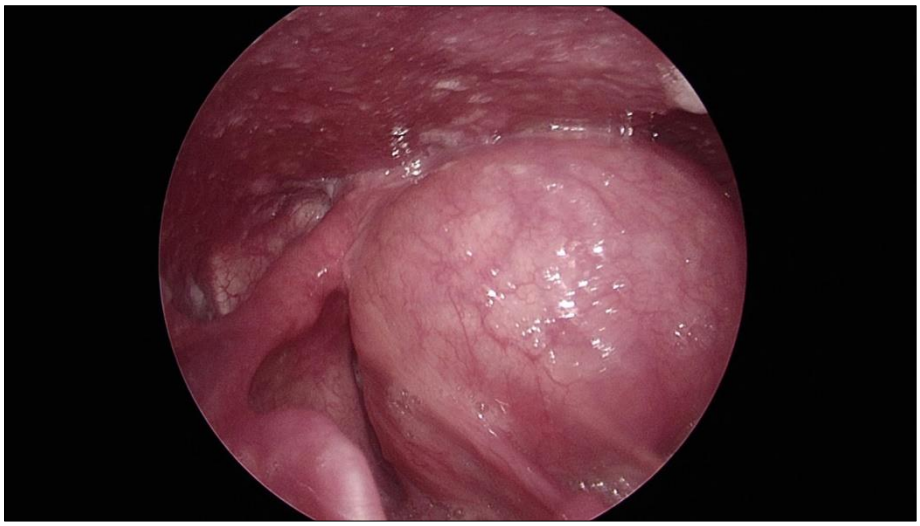

Figure 1: Direct laryngoscopic view

Brain and neck CT showed a left glottic and supraglottic laryngeal mass with smooth outer margin, heterogeneous enhancement, maximum diameter of 4 cm, multiple cystic changes (Figure 2).

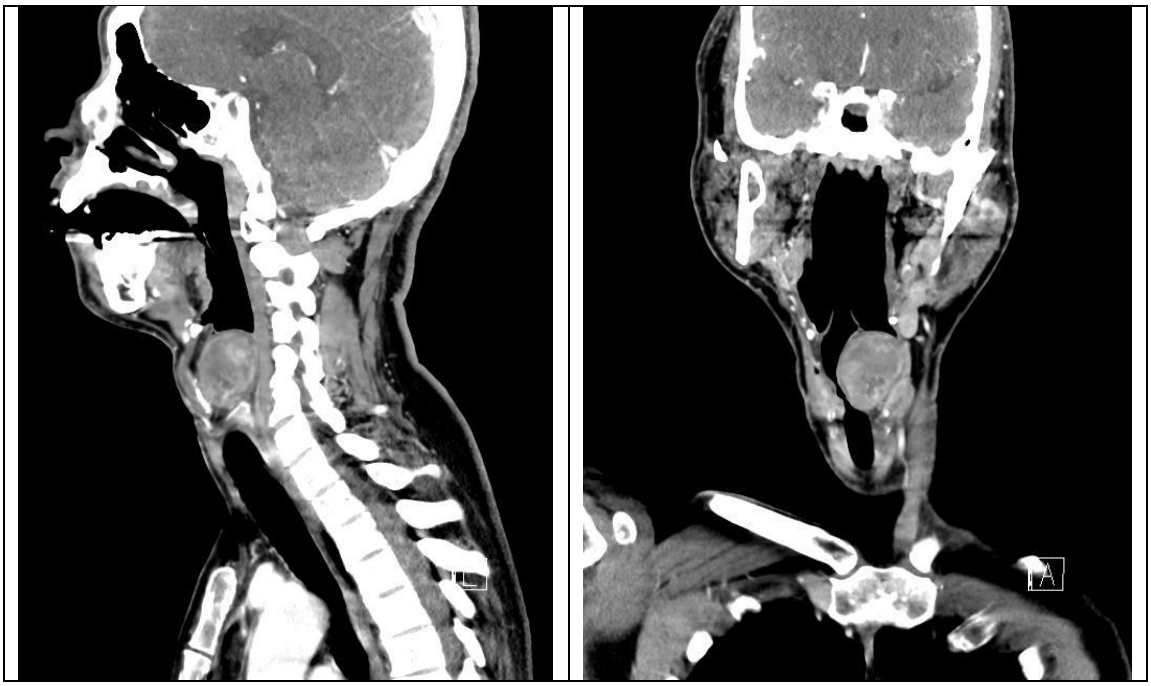

Figure 2: Brain and neck CT shows a left glottic and supraglottic laryngeal mass

After consultation with emergency medicine, otolaryngology, and anesthesiology, it was decided to perform a tracheostomy under local anesthesia to secure the airway, as there was no immediate surgical plan for airway management. There were concerns about the possibility of airway obstruction after induction of general anesthesia, the endotracheal tube entering the narrow space between the tumor and the trachea, or deterioration due to bleeding.

Because the mass was compressing the underlying structures, it seemed difficult to secure space for a tracheostomy. By explaining the situation in writing to the patient, the tracheostomy could be performed safely.

DISCUSSION

It can be difficult to suspect and diagnose a laryngeal mass because it is common to assume that a patient's difficulty breathing is simply a lung or heart condition. In addition is very rarely diagnosed in adults. Therefore, it is very important to suspect a laryngeal lesion when respiratory symptoms occur in patients with a history of neurofibromatosis. However, in this patient's case, he had a history of neurofibromatosis type II and was currently experiencing respiratory symptoms, but it

was thought to be a sequela of COVID infection, but the diagnosis was made by coincidental brain and neck CT.

Clinical findings in laryngeal tumors include hoarseness, dysphagia, odynophagia, dysarthria, globus sensation, dyspnea, and shortness of breath [7]. When located in the larynx, neurofibromas usually occur in the false cords or aryepiglottic folds [4].

Treatment of laryngeal neurofibroma depends on the location, extent, and severity of symptoms. For small solitary lesions, local resection is usually curative and is not associated with neurological deficits, except for localized intraneural neurofibroma, where resection necessarily requires sacrifice of the parent nerve [8]. Recurrence after complete excision is rare and occurs months to years later [9]. Complete excision is the therapy of choice and has two main goals: prevention of recurrence and preservation of laryngeal function. Complete resection of laryngeal neurofibroma and preservation of laryngeal function is often impossible due to the infiltrative nature of the lesion. Currently, there are no specific guidelines for the management of these patients. A conservative approach is advisable when total resection is not possible and the patient has minimal symptoms [3].

Other treatment options include tracheostomy or open surgery. However, regardless of the surgical approach, there is a significant likelihood of recurrence due to the infiltrative nature of laryngeal neurofibroma [10]. Long-term follow-up is essential because of the high recurrence rate and the possibility of malignant degeneration.

The patient in this case underwent tracheostomy to secure the airway and is being followed up.

CONCLUSION

When respiratory symptoms occur in a patient with a history of neurofibromatosis, laryngeal neurofibroma should be suspected and appropriate tests should be performed to diagnose and treat it.

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