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CASE REPORT

Ortner's Syndrome: Aortic Aneurysm as a Cause of Unilateral Vocal Cord Paralysis

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ABSTRACT

Hoarseness is a symptom of Ortner's syndrome, a rare disorder caused by mechanical left recurrent laryngeal nerve compression by enlarged cardiovascular structures. This is a case of Ortner's syndrome in an elderly 74-year-old male with an aortic arch aneurysm presenting with hoarseness. We believe the aneurysm causes the left recurrent laryngeal nerve compression, resulting in unilateral vocal cord paralysis. We emphasise the significance of Ortner's syndrome as hoarseness of voice, a significant differential diagnosis in an elderly patient despite the absence of cardiac symptoms.

INTRODUCTION

Both benign and malignant aetiologies can result in recurrent laryngeal nerve paralysis. Vocal cord paralysis is easily detected by laryngoscopy; however, determining the cause can be difficult. Neoplasia is the leading cause of vocal cord paralysis, followed by iatrogenic surgical intervention and aortic and intracranial diseases (Madhuraj et al., 2021; Mulpuru et al., 2008).

Ortner's syndrome is not a common cause of left recurrent laryngeal nerve (RLN) palsy due to inflammatory or mechanical strain caused by enlarged cardiovascular structures. The three initial cases recorded by

Ortner in 1897 involved individuals with mitral valve stenosis and dilated left atrium causing left RLN compression cases were attributed to aortic aneurysm, dilated left ventricle and dilated left pulmonary artery (Ortner, 1897).

A thoracic aortic aneurysm (TAA) is a rare cause of vocal cord paralysis, occurring in less than 5% of individuals with this vascular disorder, and involvement of the ascending aorta and aortic arch is equally infrequent (Madhuraj et al., 2021). In addition, the left RLN is more susceptible to compression than its right counterpart due to its long, winding route and proximity to major mediastinal vessels, the oesophagus, the trachea, and the lung apex (Leoce et al., 2021; Subramaniam et al., 2011).

There is a strong correlation between the rising incidence of TAA and the elderly population (Isselbacher et al., 2022). Asymptomatic patients may benefit from conservative therapy to reduce aortic stress and slow the progression of aortic dilation. Primary interventions for symptomatic patients include stent graft repair and open surgery with aorta replacement (Ismazizi et al., 2016; Isselbacher et al., 2022). TAA should be considered when other common causes of hoarseness of the voice have been ruled out.

CASE PRESENTATION

This is a case of a 74-year-old male who presented for investigation of hoarseness for the past month. He denies upper respiratory tract infection symptoms, hemoptysis, dysphagia, chest pain, or shortness of breath. There are no associated constitutional symptoms. He is a chronic smoker with underlying ischemic heart disease, chronic kidney disease and hypertension. He denies any allergies, history of trauma or surgery to the neck or upper thorax, gastroesophageal reflux, or underlying malignancy. On physical examination, he was malnourished and thin-structured. The oral cavity was normal. A laryngoscope examination showed left vocal cord paralysis with no evidence of laryngeal inflammation, infection, ulceration, or neoplasm. There is no cervical lymphadenopathy or other neurological deficit. His blood pressure, pulses, respiratory rate, and oxygen saturation were normal. His lung and heart sounds were unremarkable.

Patient first proceeded with a contrast-enhanced computed tomography (CECT) scan of the neck, which showed an anteromedial rotation of the arytenoid cartilage and air distention of the left laryngeal ventricle demonstrating 'sail sign' compatible with left vocal cord paralysis (Figure 1). No enhancing lesion was seen at the glottic region to suggest laryngeal malignancy or cervical lymphadenopathy. An aortic arch aneurysm was detected. However, the image was suboptimal as its extension and relations were not included in a neck scan's limited field of view. Subsequently, a computed tomography angiography (CTA) of the thorax was performed, which shows a wide neck 3.2 x 3.6 x 1.5cm saccular aneurysm arising from the aortic arch with peripheral mural thrombus, protruding into the aortopulmonary window and mildly compressing the left proximal main pulmonary artery (Figure 2). There are no direct or indirect signs of impending aortic aneurysmal rupture. There was no other pathology along the course of left RLN. There was no mediastinal lymphadenopathy or features of lung malignancy.

This led us to conclude that the hoarseness the patient was experiencing was caused by Ortner's syndrome, which is caused by saccular aneurysmal dilatation of the aortic arch compressing on the left recurrent laryngeal nerve. As a result, the patient was recommended for endovascular aortic repair. However, he declined surgical intervention because of financial constraints and the considerable risk associated with the procedure.

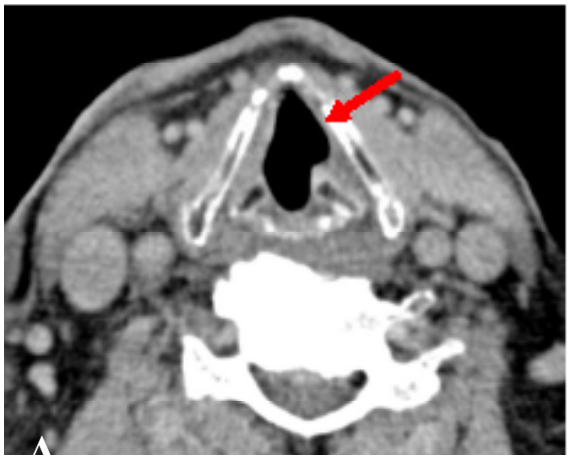
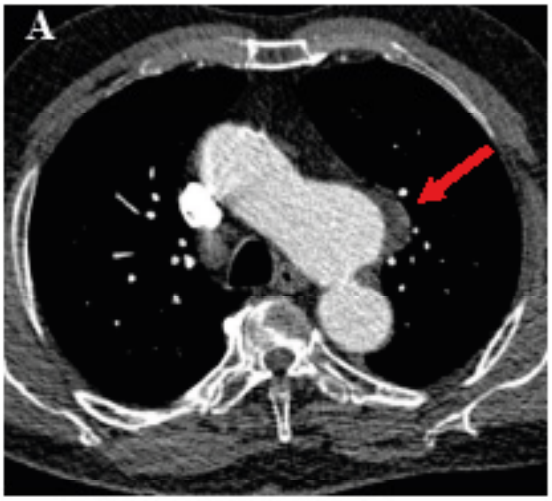


Figure 1 Axial image of CECT Neck at the glottis level shows paralysis of the left vocal cord evident by unilateral dilatation of the laryngeal ventricle demonstrating a 'sail sign' (red arrow)



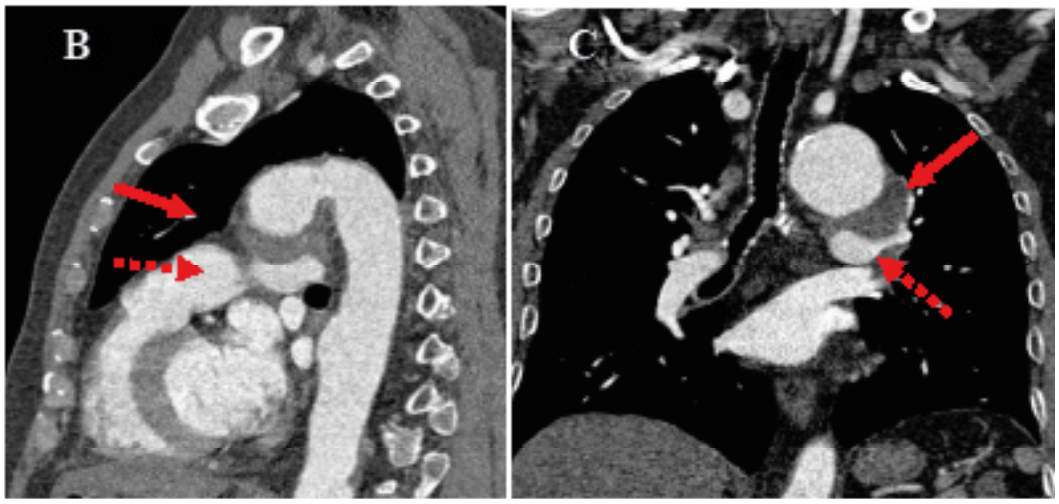


Figure 2 (A) Axial, (B) sagittal, and (C) coronal reformatted images of CTA thorax showing a wide neck aortic arch saccular aneurysm with peripheral mural thrombus (solid red arrow) and its dome pointing inferiorly protruding to the aortopulmonary window along the course of the left recurrent laryngeal nerve and compressing onto the left proximal main pulmonary artery (red dotted arrow).

DISCUSSION

An aortic arch aneurysm is a known risk factor for Ortner's syndrome, and occurrences of this illness as a subsequent complication have been reported in the medical literature (Madhuraj et al., 2021; Mulpuru et al., 2008). In addition to thoracic aortic aneurysm causing hoarseness due to vocal cord paralysis, patients presenting symptoms are dysphagia for oesophageal compression, difficulty breathing due to tracheal compression, and either superior vena cava or innominate vein compression resulting in superior vena cava syndrome. In addition, chest discomfort, fatigue, and jaw, neck, or back pain are nonspecific symptoms related to this condition (Isselbacher et al., 2022).

The immobility of the vocal colds characterises Ortner's syndrome. It is linked to damage or injury to the left RLN or vagus nerve along its course, extending from the jugular foramen until the aortopulmonary window (Agarwal et al., 2020; Zhang et al., 2022). Due to its long meandering route and proximity to the significant mediastinal vessels, oesophagus, trachea, and lung apex, the left RLN is more susceptible to compression than its right-side counterpart. Therefore, understanding the pathophysiology of Ortner's syndrome mandates a review of the anatomy of the left RLN. The vagus nerve's branch is left RLN that begins in the superior mediastinum and descends inferiorly, where it courses anterolateral to the aortic arch and aortopulmonary artery through the aortopulmonary window, distal to the ligamentum arteriosum, before it ascends in the tracheoesophageal groove (Wang et al., 2016; Zangirolami et al., 2015).

The vocal cord palsy treatment relies on the condition's underlying causes, symptoms, severity, and duration. Options for treatment include voice therapy, surgical procedures such as bulk injections, structural implants, vocal cord repositioning, and nerve replacement. A tracheotomy may be necessary if both vocal cords are paralysed. Voice therapy consists of exercises and activities to strengthen the vocal cords, improve breath control, and safeguard the airway during swallowing. Surgical interventions aim to enhance the ability to speak and swallow by repositioning the paralysed vocal cord, repositioning the vocal cord, or replacing the damaged nerve. Emerging electrical stimulation treatments may also be considered (Williamson & Shermetaro, 2022).

Most patients with TAA are asymptomatic (Isselbacher et al., 2022). Therefore, hoarseness as a symptom in the absence of any cardiac symptoms in the case of TAA is an unusual presentation. In this case, our patient presented with hoarseness due to a saccular aneurysm developing from the aortic arch compressing the left RLN. Imaging revealed that the aneurysm was near the point where the left RLN loops around the aortic arch, suggesting either stretching of the nerve or compression of the nerve at the aortopulmonary window likely caused the nerve palsy.

Aneurysms of the aortic root and ascending thoracic aorta are often heritable and occur at younger ages. In contrast, aneurysms of the descending thoracic aorta are typically degenerative and occur at older ages (Isselbacher et al., 2022). Therefore, CT angiography is ideal for depicting TAA, measuring its size and neck diameter, delineating its morphology, evaluating extension to the adjacent great vessels, and its effect on the neighbouring structures. However, a chest radiograph should be performed as a baseline screening to avoid suboptimal scanning resulting in scan repetition and exposing the patient to double radiation dose, as happened in our patient. Furthermore, should a chest radiograph be performed prior, the aortic aneurysm could be detected early, and a CT neck and thorax can be performed together.

Surgical or endovascular intervention in TAA is intended to lower the risk of unfavourable aortic events, such as aortic rupture, dissection, and aortic-related death. The AHA guidelines for thoracic aortic aneurysm dictate that an aneurysm more than or equal to 5.5 cm at the time of diagnosis, an aneurysm with an expansion rate of more than 0.5 cm per year and the onset of symptoms (which includes hoarseness) are the indications for surgical intervention (Isselbacher et al., 2022).

When the TAA diameter is more than 6 cm, there is an increased prevalence of aortic-related events such as dissection or rupture. This justifies intervention when the diameter exceeds or exceeds 5.5 cm. In addition, previously identified high-risk features of TAA rupture support the decision to intervene at a smaller diameter threshold when specific parameters are met. These parameters include rapid aortic growth (≥0.5 cm/y), symptomatic or infected aneurysm, saccular aneurysm morphology, underlying connective tissue disorder or heritable thoracic aortic diseases, and female gender (Isselbacher et al., 2022).

Therefore, determining the appropriate timing of intervention necessitates a detailed anatomic evaluation, followed by a consideration of the relative risks of intervention versus unfavourable aortic events. Early detection of Ortner's syndrome may aid in initiating immediate treatment to prevent permanent left RLN damage and restore vocal cord function. Voice improvement is expected within a few weeks of surgery, and hoarseness is reported to resolve entirely within four months (Zhang et al., 2022). In addition, endovascular stent grafting of the underlying TAA may result in progressive shrinkage of the thrombosed aneurysm, reducing nerve compression and subsequent resolution of the left RLN palsy.

Our patient's age and multiple comorbidities precluded a high-risk open thoracic repair. Hence, a thoracic endovascular aneurysm repair (TEVAR) is the best option. However, he refused any surgical intervention due to financial constraints and the high risk associated with the procedure and opted for surveillance imaging follow-up.

CONCLUSION

Ortner's syndrome is an uncommon condition that occurs as a result of a variety of cardiopulmonary disorders. Nerve compression between the pulmonary artery and the aorta is a constant factor. This report highlights the significance of conducting a cardiovascular workup in unilateral vocal cord palsy cases when no other obvious aetiology can be identified.

CONFLICT OF INTEREST

The authors declare that they have no competing interests in publishing this case.

CONSENTS

Written informed consent was obtained from the patient to publish this case. A copy of the written consent is available for review by Chief Editor.

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