

Síndrome de Ortner: Uma causa rara de disfonia

Ortner's Syndrome: A rare cause of dysphonia

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RESUMO

A síndrome de Ortner é uma entidade rara, caracterizada por disfonia decorrente da paralisia do nervo laríngeo recorrente (NLR), no contexto de doenças cardiovasculares.

Este trabalho relata dois casos de síndrome de Ortner. Homem, 81 anos, apresentava disfonia com uma semana de evolução. Ao exame objetivo verificou-se paralisia da prega vocal esquerda. A tomografia computadorizada (TC) revelou dilatação auricular esquerda.

Homem, 76 anos, com queixas de disfonia há duas semanas. Observou-se paralisia da prega vocal esquerda. A TC demonstrou um aneurisma do arco aórtico.

O NLR inerva os músculos laríngeos intrínsecos, com exceção do músculo cricotiroideu. A paralisia unilateral do NLR está associada a disfonia mas a paralisia bilateral pode causar sintomas mais severos, como dispneia e estridor.

Muitas patologias benignas e malignas podem afetar o nervo ao longo do seu percurso, pelo que a imagiologia é fundamental no diagnóstico diferencial.

Apesar de rara, a síndrome de Ortner pode causar paralisia da prega vocal.

Palavras-chave: disfonia; nervo laríngeo recorrente; paralisia prega vocal; síndrome ortner

ABSTRACT

Ortner's syndrome is a rare entity, characterized by dysphonia caused by recurrent laryngeal nerve (RLN) palsy, secondary to cardiovascular disorders.

This work reports two cases of Ortner's Syndrome. A 81-year-old male presented with dysphonia for a week. Physical examination showed left vocal fold palsy. Computed tomography (CT) revealed left atrial dilation. A 76-year-old male, had complaints of dysphonia for two weeks. Left vocal fold paralysis was observed. CT showed an aortic arch aneurysm.

RLN is responsible for the innervation of the intrinsic laryngeal muscles, with exception of the cricothyroid muscle. Unilateral RLN palsy usually causes hoarseness. Bilateral RLN paralysis may be associated with more severe symptoms, such as dyspnea and stridor.

Many benign and malign causes can affect the nerve through its course, whereby imaging is crucial in differential diagnosis. Despite being rare, Ortner's syndrome may be the cause of vocal fold paralysis.

Keywords: hoarseness; recurrent laryngeal nerve; vocal fold paralysis; ortner's syndrome

INTRODUCTION

Ortner's syndrome or cardiovocal syndrome refers to dysphonia caused by recurrent laryngeal nerve palsy, secondary to compression between the aorta and the pulmonary artery, in the context of cardiovascular disorders. It was first described in 1897 by Norbert Ortner, an Austrian physician, in a patient with mitral stenosis and left atrial enlargement.¹

Ortner's syndrome is a rare entity, with no information on the literature about its incidence.²

The left recurrent nerve is 1.75 times more often affected than the right side and compression is usually caused by dilation of the left atrium or by dilation of pulmonary artery.^{1,2,3}

We report two cases of Ortner's syndrome. Our aim is to review the literature and discuss and highlight hoarseness resulting from recurrent laryngeal nerve paralysis in the context of cardiovascular disease.

CASE REPORT

Case 1

A 81-year-old male, with history of atrial fibrillation and stroke, was observed by an Ear Nose and Throat (ENT) doctor referring hoarseness and discrete dysphagia for

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one week. Physical examination showed left vocal fold palsy, with no other findings. In this context, cervical and thoracic computed tomography (CT) was performed and revealed left atrial dilation secondary to mitral stenosis, with compression of the left recurrent laryngeal nerve (Figures 1 and 2).

The patient begun medical treatment to improve cardiovascular risk factors and control the heart rate.

FIGURE 1

Axial CT with asymmetry of the vocal folds due to left paralysis.

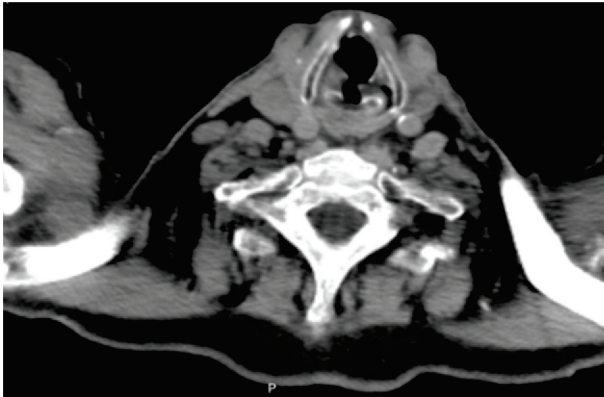
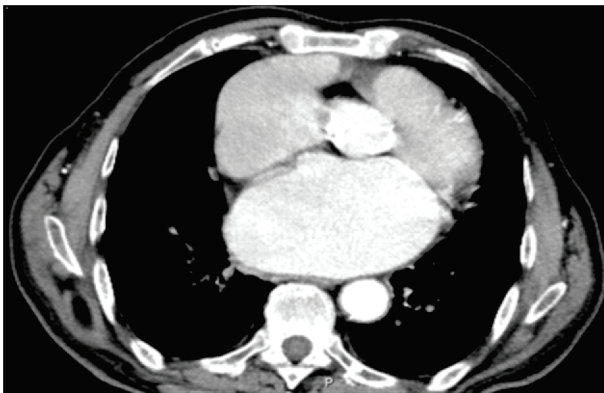


FIGURE 2

Axial CT with cardiomegaly - severe bilateral auricular dilatation (more pronounced on the left side).



Case 2

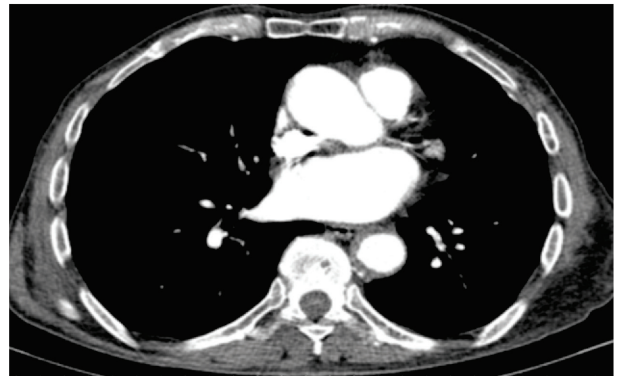
A 76-year-old male, with hypertension, hypercholesterolemia and chronic obstructive pulmonary disease, had complaints of dysphonia in the previous two weeks. At physical examination left vocal fold paralysis was observed.

Cervical and thoracic CT showed an aortic arch aneurysm with compression of the left recurrent laryngeal nerve (Figure 3).

Further investigation revealed coronary artery disease and carotid stenosis <50%. Taking into consideration the patient's comorbidities and functional status, medical treatment was chosen, with hypertension control, high-dose statin and dual antiplatelet therapy.

FIGURE 3

Angio CT showing aortic arch aneurysm



DISCUSSION

The left vagus nerve descends into the superior mediastinum between the left common carotid and subclavian arteries before traversing the left side of the aortic arch. At this level it gives rise to the left recurrent laryngeal nerve which hooks around the ligamentum arteriosum before ascending in the groove between the esophagus and trachea. It continues along this groove to supply ipsilateral motor innervation to the larynx intrinsic muscles, with the exception of cricothyroid muscle.^{4,5}

Since left recurrent laryngeal nerve has a longer course than right recurrent laryngeal nerve, several studies have identified left-sided vocal fold paralysis to be more common than right.^{4,5,6}

In cases of recurrent laryngeal nerve paralysis, vocal fold may be at midline, paramedian or lateral position.⁷ In unilateral recurrent laryngeal nerve palsy patients typically experience hoarseness and hypophonia. These can range from subtle vocal fatigue to near-total aphonia, depending on the vocal fold position and consequent degree of glottic insufficiency. Dysphagia with aspiration may also be reported but is more frequent when both superior and recurrent laryngeal nerves are affected.^{2,8,9}

In patients with hemilaryngeal paralysis observation is an option, especially if there is minimal vocal disability, no evidence of aspiration or comorbidities that discourage intervention. Treatment options vary from voice therapy, injection laryngoplasty, medialization thyroplasty, arytenoid repositioning procedures or laryngeal reinnervation.^{8,10}

Bilateral recurrent laryngeal nerve paralysis usually presents as an inability to abduct the vocal folds, which may be associated with more severe symptoms such as dyspnea or stridor. Since the number of adductor muscle fibers are approximately 4 times greater than the number of abductor muscle fibers, it is anticipated that the adductors will receive greater reinnervation and the vocal folds tend to assume a paramedian position.^{2,11}

In bilateral vocal fold paralysis, treatment is guided by the degree of airway limitation. Tracheostomy is frequently

performed in an emergency situation and may be an option for the long term as well, but generally patients prefer to avoid the inconveniences of tracheostomy. Other treatment options include lateralization of the vocal fold, resection of the arytenoid and/or vocal fold tissue to enlarge the glottic aperture or reinnervation. Some of these procedures are destructive and irreversible, so the otorhinolaryngologist should be sure that any reasonable possibility of spontaneous improvement has been exhausted before they are considered.^{8,12}

Recurrent laryngeal nerve lesion may occur in any segment of the nerve. In Ortner's syndrome, compression of the nerve is due to benign cardiovascular diseases, however other causes may affect the nerve through its course, whereby imaging is crucial in differential diagnosis (Table 1).^{13,14}

CT from the skull base through the arch of the aorta or the right subclavian, is the minimum recommended study for laryngeal paralysis. In cases where a 'high vagal' paralysis is suspected magnetic resonance imaging (MRI) may offer a more reliable means of imaging the skull base or central nervous system.^{8,15}

Tomographic findings of vocal fold paralysis include paramedian position of the vocal fold, thickening of the ipsilateral aryepiglottic fold and increased volume of the ipsilateral pyriform sinus and ventricle.¹³

Persistent voice changes justifies observation by an Otorhinolaryngologist. In cases of unilateral vocal fold paralysis, if the other vocal fold can act in compensation, hoarseness may decrease. In both cases presented, hoarseness persisted, with little improvement, despite optimized medical treatment.

CONCLUSION

Vocal fold palsy that isn't otherwise explained by other findings after an Otorhinolaryngologist examination should be studied with cervico-thoracic CT or MRI.

There are many benign and malignant causes for recurrent laryngeal nerve lesion. Despite being rare, in a patient with cardiovascular disease, Ortner's syndrome may be the cause of vocal fold paralysis.

TABLE 1

Differential diagnosis of vocal fold paralysis

Cardiovascular	Aortic dissection or pseudoaneurysm, left atrial enlargement, congenital heart disease, pulmonary artery enlargement, pulmonary embolism
Neoplastic	Bronchogenic, esophageal or thyroid carcinoma, lymphoma, neurogenic tumors, thymic malignancy, nodal retrosternal goiter (extremely rare)
Iatrogenic	Cardiac surgery or median sternotomy, patent ductus arteriosus ligation or embolization, left pneumonectomy or lobectomy, mediastinoscopy, radical esophagectomy, tracheal resection, thymectomy, thyroidectomy, central venous line catheterization, anterior approaches to the cervical spine, carotid endarterectomy, external radiation therapy
Inflammatory/Infiltrative	Sarcoidosis, silicosis, fibrosing mediastinitis, amiloidosis
Infectious	Tuberculosis, histoplasmosis, coccidiomycosis, bacterial abscess, mycotic aortic pseudoaneurysm
Traumatic	Endotracheal intubation, deceleration injuries, penetrating injuries

Conflito de Interesses

Os autores declaram que não têm qualquer conflito de interesse relativo a este artigo.

Confidencialidade dos dados

Os autores declaram que seguiram os protocolos do seu trabalho na publicação dos dados de pacientes.

Proteção de pessoas e animais

Os autores declaram que os procedimentos seguidos estão de acordo com os regulamentos estabelecidos pelos diretores da Comissão para Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

Política de privacidade, consentimento informado e Autorização do Comitê de Ética

Os autores declaram que têm o consentimento por escrito para o uso de fotografias dos pacientes neste artigo.

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Disponibilidade dos Dados científicos

Não existem conjuntos de dados disponíveis publicamente relacionados com este trabalho.

Referências bibliográficas

1. Subramaniam V, Herle A, Mohammed N, Thahir M. Ortner's syndrome: - case series and literature review. *Braz J Otorhinolaryngol*. Sep-Oct 2011;77(5):559-62. doi: 10.1590/s1808-86942011000500004.
2. Klee K, Eick C, Witlandt R, Gawaz M, Didczuneit-Sandhop B. Unilateral recurrent nerve palsy and cardiovascular disease – Ortner's syndrome. *J Cardiol Cases*. 2016 Dec 8;15(3):88-90. doi: 10.1016/j.jccase.2016.10.018.
3. Heikkinen J, Milger K, Alexandre-Lafont E, Woitzik C, Litzlbauer D, Vogt JF. et al. Cardiovascular syndrome (Ortner's syndrome) associated with chronic thromboembolic pulmonary hypertension and giant pulmonary artery aneurysm: case report and review of the literature. *Case Rep Med*. 2012;2012:230736. doi: 10.1155/2012/230736.
4. Bickle IC, Kelly BE, Brooker DS. Ortner's syndrome: a radiological diagnosis. *Ulster Med J*. 2002 May;71(1):55-6.
5. Yamada M, Hirano M, Ohkubo H. Recurrent laryngeal nerve paralysis. A 10-year review of 564 patients. *Auris Nasus Larynx*. 1983;10 Suppl:S1-15. doi: 10.1016/s0385-8146(83)80001-7.
6. Glazer HS, Aronberg DJ, Lee JK, Sagel SS. Extralaryngeal causes of vocal cord paralysis: CT evaluation. *AJR Am J Roentgenol*. 1983 Sep;141(3):527-31. doi: 10.2214/ajr.141.3.527.
7. Seyed Toutouchi SJ, Eydi M, Golzari SE, Ghaffari MR, Parvizian N. Vocal cord paralysis and its etiologies: a prospective study. *J Cardiovasc Thorac Res*. 2014;6(1):47-50. doi: 10.5681/jcvtr.2014.009.
8. Sulica L, Sadoughi B. Paralysis of the Larynx. In Watkinson JC, Clarke RW, editors. *Scott-Brown's Otorhinolaryngology and Head and Neck Surgery*. 8th Ed. p. 1101-10. Boca Raton: CRC Press; 2018.
9. Misono S, Merati AL. Evidence-based practice: evaluation and management of unilateral vocal fold paralysis. *Otolaryngol Clin North Am*. 2012 Oct;45(5):1083-108. doi: 10.1016/j.otc.2012.06.011.
10. Misono S, Merati AL. Evidence-based practice: evaluation and management of unilateral vocal fold paralysis. *Otolaryngol Clin North Am*. 2012 Oct;45(5):1083-108. doi: 10.1016/j.otc.2012.06.011.
11. Li Y, Garrett G, Zeale D. Current Treatment Options for Bilateral Vocal Fold Paralysis: A State-of-the-Art Review. *Clin Exp Otorhinolaryngol*. 2017 Sep;10(3):203-212. doi: 10.21053/ceo.2017.00199.
12. Sapundzhiev N, Lichtenberger G, Eckel HE, Friedrich G, Zenev I, Toohill RJ, Werner JA. Surgery of adult bilateral vocal fold paralysis in adduction: history and trends. *Eur Arch Otorhinolaryngol*. 2008 Dec;265(12):1501-14. doi: 10.1007/s00405-008-0665-1.
13. Dutra BL, Campos LC, Marques HC, Vilela VM, Carvalho REDS, Duque AGS. Síndrome de Ortner: relato de caso e revisão da literatura. *Radiol. Bras*. [Internet] 2015 Jul/Ago; 48(4):260-62. Available from: <https://doi.org/10.1590/0100-3984.2013.1836>.
14. Paquette CM, Manos DC, Psooy BJ. Unilateral vocal fold paralysis: a review of CT findings, mediastinal causes and the course of the recurrent laryngeal nerves. *Radiographics*. May-Jun 2012;32(3):721-40. doi: 10.1148/rg.323115129.
15. Bilici S, Yildiz M, Yigit O, Misir E. Imaging Modalities in the Etiologic Evaluation of Unilateral Vocal Fold Paralysis. *J Voice*. 2019 Sep;33(5):813.e1-813.e5. doi: 10.1016/j.jvoice.2018.04.017.