

A Systematic Review of Ortner's Syndrome

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Abstract: Ortner's syndrome, also known as the syndrome cardiovocal, is characterized by the compression, stretching or dissection of recurrent laryngeal nerve, secondarily caused by cardiovascular abnormal. Through a systematic review about the issue, on the main bases data, we named the main aspects which go around the syndrome, from its symptoms to its treatments based on its different causes, such as, valvar mitral stenosis, aortic aneurysm among others whose effect brings about a compression of the recurrent laryngeal nerve. Therefore, the left recurrent laryngeal nerve is more affected because of its anatomical position. Due to its path surrounding aortic arch and passing between pulmonary artery and aortic arch, it has a greater chance to compression. Its clinical importance is related to differential diagnosis which is also connected to many different pathologies such as neoplasia (pulmonary, thyroid, mediastinal, esophageal) and surgical iatrogenic causes which may affect the recurrent laryngeal nerve causing various symptoms related to this involvement.

Key words: Recurrent laryngeal nerve, hoarseness, paralysis, aortic aneurysm, syndrome, vocal cord paralysis.

1. Introduction

Ortner's syndrome, also known as cardiovocal syndrome, was found out in 1897 by Norbert Ortner, through the hoarseness shown in some of his patients with the same bases pathology—in this case, mitral valvar stenosis and consequently atrial dilatation [1]. Ortner postulated that the syndrome occurred through atrial dilatation due to mitral stenosis that compressed the recurrent laryngeal nerve. However, over the years, many authors in their studies, although unable to fully explain the mechanism of the syndrome, emphasized importance of other factors besides atrial dilatation [2]. Other cardiovascular abnormalities have been noted as important causes of Ortner's syndrome, including aortic arch aneurysm, persistent ductus arteriosus, pulmonary artery aneurysm, Eisenmenger's syndrome, primary pulmonary hypertension, and atrial and ventricular septal defects [2]. So this syndrome was characterized by the palsy of the recurrent laryngeal nerve secondarily caused by the cardiovascular

abnormalities. The current study aims to put together several articles about the syndrome and to emphasize the most important ones published in Brazil and in the world, related to physiopathology, symptoms, complementary exams, differential diagnoses, treatment and clinical importance belonging to Ortner's syndrome.

2. Materials and Methods

2.1 Type of Survey

Systematic review of literature was accomplished in March of 2018.

2.2 Location

Library of Santo Amaro University, São Paulo, São Paulo, Brazil.

2.3 Search for the Studies

On March 4th 2018, it was accomplished a search of the main data bases from the healthy literature (PubMed, Lilacs, Cochrane Library and Scielo) using the following keywords: Hoarseness/Rouquidão,

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Recurrent Laryngeal Nerve/Nervo Laríngeo Recorrente, Paralysis/Paralisia, Aortic Aneurysm/Aneurisma Aórtico, Syndrome/Síndrome, Vocal Cord Paralysis/Paralisia da corda vocal. Later on, we selected only published articles from the last 5 years. With all this research we could get the following results shown in Table 1.

3. Results and Discussions

The knowledge about the topography of recurrent laryngeal nerve favors the understanding of the signs and symptoms characteristic of the disease. The 10th pair of the cranial nerve (vagus nerve) gives origin to the right and left recurrent laryngeal nerve. While the right vagus nerve takes a way down to the right subclavian artery where it goes up originating the right recurrent laryngeal nerve toward the right vocal chord, the left vagus nerve takes the way down to the level of the aortic arch, which gives origin to the left recurrent laryngeal nerve, which surrounds the aortic arch in order to in this way rise and innervate the intrinsic muscles of the larynx—excepting the cricothyroid muscle—and thus vocal chord ipsilateral. This long path brings a kind of vulnerability to the left recurrent laryngeal nerve. Its anatomical location makes the left recurrent laryngeal nerve susceptible to compressions by the several organs existent in the mediastinal cavity and consequently the most incidence of the left vocal chord palsy in comparison to the right, in what is noticeable in many cases. Most of the articles suggest

one incidence of patients presenting mainly hoarseness, besides other symptoms such as a left vocal chord palsy, dysphagia, dysphonia and dyspnea. This one is proved by the laryngoscopy exam shown in Fig. 1.

There are several causes of recurrent laryngeal nerve compression, among them we can highlight abnormal such as aortic dissection, pulmonary hypertension, aortic aneurysm, cardiovascular abnormalities, chronic obstructive pulmonary disease (COPD), pulmonary artery stenosis, recurrent pulmonary embolism, and cardiac congenital defects.

One of the complementary exams that may aid in the diagnosis of several underlying cardiovascular abnormalities is chest tomography. As shown in Fig. 2, it shows the aortic arch aneurysm which is related to the compression of the recurrent laryngeal nerve and consequently to the described syndrome.

Another important complementary exam is vocal cord tomography. The tomographic findings of vocal cord paralysis are: paramedian thickening of the ipsilateral aryepiglottic fold, piriform sinus volume increase, anteromedial positioning of the arytenoid cartilage, paramedian position of the vocal cord. In the coronal plane, rectification of the subglottic arch can be observed [3].

These exams most often show the underlying causes that may be affecting the recurrent laryngeal nerve. Besides the confirmation of possible pathologies, its usefulness is also related to the different differential diagnoses.

Table 1 Results of the main data bases searches from the healthy literature.

Data bases	Strategy of searching	Results
PubMed	(((("Hoarseness" [Mesh]) and "Recurrent Laryngeal Nerve" [Mesh]) and "Paralysis" [Mesh]) and "Aortic Aneurysm" [Mesh]) and "Syndrome" [Mesh]) and "Vocal Cord Paralysis" [Mesh]	3 articles
	("syndrome" [Mesh Terms] and "hoarseness" [Mesh Terms]) and "recurrent laryngeal nerve" [Mesh Terms]	7 articles
Cochrane Library	#1 Mesh descriptor: [Hoarseness] explode all trees #2 Mesh descriptor: [Syndrome] explode all trees #3 Mesh descriptor: [Recurrent Laryngeal Nerve] explode all trees	0 articles
Lilacs	(mh:(hoarseness)) and (mh:(recurrent laryngeal nerve)) and (mh:(paralysis)) and (mh:(aortic aneurism)) and (mh:(syndrome)) and (mh:(vocal cord paralysis))	0 articles
	(mh:(hoarseness)) and (mh:(syndrome)) and (mh:(recurrent laryngeal nerve))	0 articles
Scielo	(Rouquidão) and (síndrome) and (paralisia) and (nervo laríngeo recorrente) and (cardiovocal)	1 article



Fig. 1 Laryngoscopy showing left vocal cord palsy [3].



Fig. 2 Chest tomography, sagittal, which highlighted the aortic arch aneurysm [4].

Concerning the aortic dissection about 10% are asymptomatic and there are few cases reported in the literature to correlate the syndrome. In the aortic dissection, the patient is used to having thoracic pain with irradiation to the back or abdominal area which may suggest the extension of dissection. The neurological symptoms may be presented, mainly as stroke, due to the involvement of carotid arteries in 20%-30% of the cases. Other symptoms may occur

such as syncope, transverse myelopathy, anterior spinal syndrome, quadriplegia, paraplegia [5]. There are also risk factors that include arterial hypertension, Marfan syndrome, aortic dilation, coarctation of aorta, annuloaortic ectasia, aortic arch hypoplasia, pregnancy, etc. [5].

Aortic aneurysm generally comes with chest pain, backache or epigastric ache, depending on the aneurysm location. An aortic arch aneurysm showing

hoarseness is extremely rare. Aortic arch aneurysms account for only 0.3% of cases of vocal cord paralysis [5]. Therefore, we cannot ignore this fact that also contributes to the development of the syndrome.

Aortic dissection and aortic aneurysm have similar symptoms, such as thoracic pain with irradiation to the back and epigastric region. These symptoms associated with hoarseness may lead to a clinical reasoning for Ortner's syndrome. The knowledge and distinction between them are important in the applied form of treatment and prognosis of the patient in question.

The hoarseness is a common clinical symptom dependent of local factors—in its own larynx and extra laryngeal. It was noticed that the short duration hoarseness may be a result of the local larynx causes, which in this case has benign origin in its majority. The local laryngeal causes include acute laryngitis, nodules from smoking patients, laryngeal tuberculosis, vocal polyps and carcinoma of the larynx. Many studies are just attributed to 1%-3% of the cases of extralaryngeal hoarseness as being the Ortner's syndrome [4].

Hoarseness is crucial in the diagnosis of Ortner syndrome, being its main symptom. However, since it is a very common nonspecific symptom, the distinction between its causes is extremely important, since most of them do not characterize the syndrome in question.

There are many factors in the patients with hoarseness which involve the recovering of the voice. For being a hoarseness caused by abnormalities, the first step to recover the voice, would be to solve the base pathology [6]. Therefore the totally recuperation of the voice also depends on the level, duration and primary cause of injured nerve. In general the persistence of the hoarseness symptom within 2-4 weeks requires an otorhinolaryngological evaluation to exclude the possible malignancies [7].

The vocal chord palsy refers to an extensive group of pathological processes which may cause the hoarseness. Common cases of the vocal chord palsy include malignant neoplasm of the lung affecting lung, esophagus and thyroid, iatrogenic events, traumas,

inflammatory lesions compressing the nerve, brain lesions and laryngeal tuberculosis [5]. More accurately, it is worth distinguishing their proportions: iatrogenic causes of recurrent laryngeal nerve (36.5%), cellular malignancy (18.4%), idiopathic causes (18.5%). Among the extralaryngeal causes of left vocal cord paralysis, the main cause would be bronchogenic carcinoma [7].

Epidemiologically the main causes of vocal cord paralysis are not related to cardiovascular causes. Iatrogenic causes assume a large number of this involvement due to the entire topographical location and the syntopy of the recurrent laryngeal nerve. In addition to this important cause, which does not characterize the syndrome and is found in a differential diagnosis, it is also worth pointing out the pulmonary causes that can cause nerve palsy.

Most of the cases are because of the nerve compression by the adjacent structures to the aortopulmonary window. Due to causes of the Ortner's syndrome dilatation of pulmonary artery includes primary pulmonary hypertension, recurrent pulmonary emboli, various congenital heart defects [4].

Also there are reports of the disease development from a left brachiocephalic vein stenosis, which results in the development of collateral circulation, where this one developed itself through the aortopulmonary window, causing the nerve compression [8].

A study summarized the main mediastinal differential diagnoses concerning recurrent laryngeal nerve involvement. In Table 2 below are the main differential diagnoses listed by the authors [3].

Patients with the diagnosis of the Ortner's syndrome demonstrated to present some symptoms in common that can characterize risk factors and associate pathologies that can contribute to the diagnosis. Among the patients with the syndrome were founded the following symptoms and possible risk factors: hypertension, hyperlipidaemia, active smoker, cardiovascular disease, hoarseness, left vocal cord paralysis, regurgitation, aortic regurgitation, chronic

Table 2 Differential diagnosis of medial causes of involvement of recurrent laryngeal nerve [3].

Vascular/cardiac	Aortic dissection or pseudoaneurysm, left atrium enlargement, congenital heart diseases, pulmonary artery enlargement, pulmonary embolism
Neoplastic	Bronchogenic carcinoma, lymphoma, esophageal carcinoma, neurogenic tumors (paraganglioma, schwannoma), thyroid carcinoma, malignant thymus disease, lymph node metastasis; retrosternal goiter
Surgical/iatrogenic	Heart surgery, median sternotomy, patent ductus arteriosus ligation or embolization, left lobectomy/pneumomectomy, mediastinoscopy, radical esophagectomy, tracheal resection, thymectomy, thyroidectomy, anterior approach in spine surgeries, carotid endarterectomy, external radiotherapy
Inflammatory	Sarcoidosis, silicosis, fibrosing mediastinitis
Infiltrative	Amyloidosis
Infectious	Tuberculosis, histoplasmosis, coccidioidomycosis, bacterial abscess, mycotic aortic pseudoaneurysm
Traumatic	Deceleration injuries, penetrating chest injuries

expectorant cough, aortic arch aneurysm, aortic dissection, dyspnea, COPD and chronic kidney disease [4-7].

The treatment is variable and dependent of the bases pathology. It generally consists in the implant of endovenosis prothesis in cases of aortic dissection besides the corrections of underlying abnormalities cardiovascular.

4. Conclusions

Patients with the Ortner's syndrome present common clinical symptoms of wide spectrum such as the case of hoarseness, even which is not the unique symptom, the others possible ones are not either pathognomonic, moreover they depend on base pathology in order to develop themselves, therefore, they make the early diagnosis difficult. The diagnosis is made by image exams that show clearly any abnormality cardiovascular capable to compress the recurrent laryngeal nerve, many times as well as the results of laryngoscopy exams in which it is noticed the vocal chord palsy. The clinical importance of the current review about the Ortner's syndrome is related mainly to the differential diagnosis with many pathologies of great impact, among them: pulmonary neoplasms, mediastinal neoplasms, thyroid neoplasms, esophageal neoplasms and even iatrogenic causes can affect the recurrent laryngeal nerve.

Thus, the need for in-depth knowledge about the various differential diagnoses, as well as the use of complementary tests is fundamental to characterize the

Ortner's syndrome. Furthermore, it is of great importance that further studies and research be performed, contributing to a joint identification of the main symptoms that accompany the base pathologies that will corroborate for the early diagnosis of the syndrome. For the purpose of perfecting the treatment, minimizing the symptoms and provide a better life quality for the patient.

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