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Gerhardt Syndrome after Reoperation for a Recurrent Plunging Goiter - A Case Report

*Gaudêncio FC, Hasenauer A, Perentes J Y, Bouayed S and Christodoulou M**

Department of Thoracic Surgery, Sion Hospital, Switzerland

Abstract

Thyroid surgery comes with several risks for complications. One of them being a vocal cord paralysis which may be uni- or bilateral. Most frequently this paralysis is due to an iatrogenic injury of the laryngeal recurrent nerve. Less frequently other etiologies are possible such as traumatic injury, malignancy, or neurologic degenerative diseases. In more than 1 case out of 5 the etiology remains unknown. We report a case of bilateral vocal cord paralysis in an immediate postoperative context for a recurrent thyroid goiter requiring invasive therapies to secure airways. An iatrogenic lesion of recurrent laryngeal nerve was excluded, and evolution shows a persistent paralysis. We conclude to the diagnosis of Gerhardt Syndrome.

Keywords: Gerhardt Syndrome; Laryngeal recurrent nerve; Recurrent goiter; Thyroidectomy; Vocal cord paralysis

Introduction

A number of complications are expected after a thyroidectomy, namely preoperative hemorrhage, hypothyroidism or vocal cord paralysis [1,2]. The differential diagnosis of a vocal cord paralysis in a postoperative context is vast. The extent of this differential diagnosis is explained by the long anatomic course of the vague nerve which renders it susceptible to a plethora of physio-pathological findings that may lead to a vocal cord paralysis. Some rare syndromes such as Gerhardt Syndrome can also result in a vocal cord paralysis in a postoperative context of thyroidectomy.

We present the case of a 68-year-old patient reoperated for a recurrent plunging goiter which presented postoperatively a bilateral vocal cord paralysis diagnosed as a Gerhardt syndrome. This led to acute and severe respiratory distress which required an orotracheal intubation and tracheostomy.

Gerhardt syndrome is a vocal cord paralysis defined by an abduction deficit. This syndrome is polyetiological and, to this day, its physiopathology has not been fully elucidated. Infectious and neurodegenerative disease, as postoperative etiologies have been described. Postoperative Gerhardt syndrome is mainly described after classical thyroidectomy [3,4], but has never been reported after reoperation of a mediastinal goiter with thoracotomy approach, to our knowledge. Thus, diagnosis can be time consuming and costly as well as urgent because of an elevated risk of acute respiratory distress.

This article describes a case of Gerhardt Syndrome in a postoperative context after reoperation for a recurrent plunging goiter.

Case Presentation

We report the case of a 68-year-old patient known for active smoking estimated at 100 UPA. He was treated for a tonsil squamous cell carcinoma by radio and chemotherapy with a full oncological response.

Furthermore, the patient is known for a multinodular plunging goiter, a serendipity found at an imaging control in the ENT oncologic context. This compressive goiter was addressed by isthmus-thyroidectomy by cervicotomy in august 2021, revealing a micro and macrofollicular constitution in histopathology. Follow-up CT scanner of July 2023 described a new massive goiter measuring 8.5 cm in axis with an estimated volume of 208 ml, exerting a mass effect on the trachea which is deviated on the right side as well as a mass effect over the esophagus which presents an important aerial dilation upwards (Figure 1, 2). We conclude to the recurrence of the thyroid goiter. A surgical management was decided and accepted by the patient.

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Correspondence:
Michel Christodoulou, Department of Thoracic Surgery, Sion Hospital, Avenue du Grand-Champsec 80, 1951 Sion, Switzerland,

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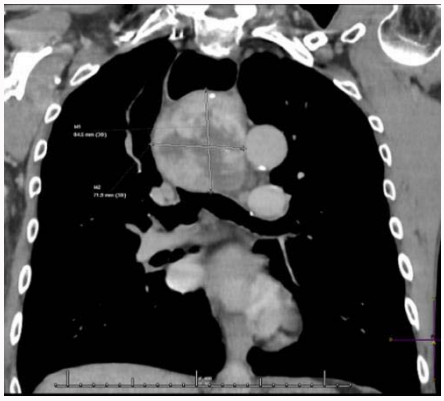

Figure 1: CT scanner coronal view, recurrent plunging goiter in retrosternal position exerting a mass effect in the trachea and esophagus.

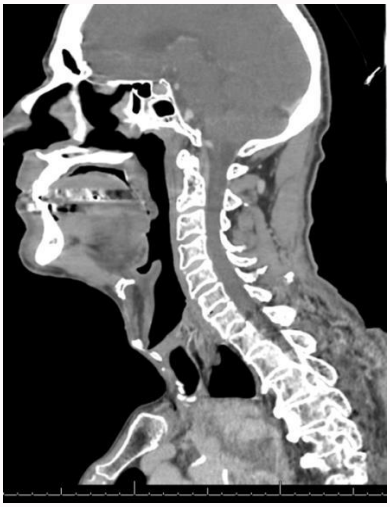

Figure 2: CT scanner sagittal view of the recurrent thyroid goiter seen behind the manubrium.

The surgical intervention took place in December 2023 at our institution in an elective manner. The patient beneficiated from a resection of the mediastinal plunging goiter associated to a lymph node dissection of the 7th station by a posterolateral thoracotomy. Surgery and anesthesia were uncomplicated, but it should be notified that the right recurrent nerve was not formally identified during intervention. Histopathological analysis showed hyperplastic thyroid tissue, without signs of malignancy.

At POD (Postoperative Day) 1, the patient developed an inspiratory stridor which rapidly deteriorated. A fibroscopy was performed which showed a symmetrical hypomotility of both vocal cords, positioned in a paramedian position with an abduction deficit. At this stage, we concluded that the postoperative edema was responsible for the symptoms. Thus, an intravenous high-dose corticosteroid treatment was initiated. Despite the medical treatment, respiratory distress deteriorated, and the patient was transferred to the Intensive Care Unit (ICU) at POD 1.

In the ICU, the patient rapidly beneficiated from a noninvasive ventilation therapy. A new fibroscopy was performed showing a paresis of the vocal cords and the absence of laryngeal oedema with a persistent abduction deficit, which are finding compatible with a diagnosis of a Gerhardt syndrome. Due to the rapidly evolutive

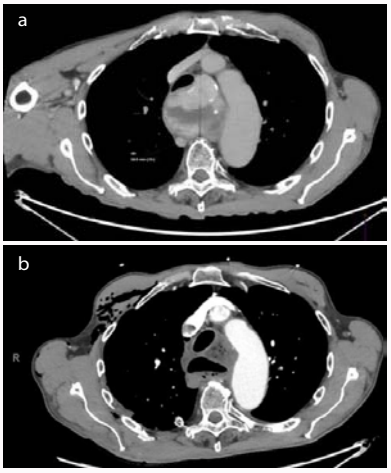

Figure 3: CT scanner transverse view a) preoperative CT scanner showing the recurrent plunging goiter at the level of the aortic arch b) postoperative CT scanner at POD2.

symptoms and the risk of respiratory failure, the patient was intubated. A CT scanner is repeated at POD 2 which did not reveal any complications namely no hematoma or post-operative edema, which refuted the first diagnosis hypothesis (Figure 3). The trajectory of the recurrent nerves shows no abnormalities, namely no hematoma or oedema, reinforcing the hypothesis of a Gerhardt syndrome. Due to the non-improvement of the respiratory and ENT symptomatology a tracheostomy was performed. This intervention took place at POD 4, without further complication. Following this intervention, invasive ventilation was rapidly stopped. At POD 5, the internal canula of the tracheotomy was replaced with a fenestrated one allowing the patient to hydrate and nourish himself by mouth with a good tolerance. A fibroscopy was repeated showing a persistence of the vocal cords' abduction deficit, motivating the introduction of an empiric new high-dose corticosteroid therapy which was conducted for a total of 5 days.

Afterwards, the evolution was favorable, and the patient was transferred to the surgical ward. The patient was discharged at POD 7. The ambulatory follow-up from the ENT team describes a persistence of a vocal cords' paralysis in a paramedian position. Nevertheless, the patient was decannulated at POD 14 and is symptomless.

Discussion

We report the case of a 68-year-old patient who presented a severe respiratory failure post mediastinal recurrent goiter resection due to bilateral vocal cords paralysis. The rapid onset and the severity of the symptoms, the spontaneous recovery and the unclear origin deserve a comprehensive review of the case.

Multiple factors could have contributed to the development of this symptomatology and differential diagnosis is large including postoperative neuropraxia, iatrogenic lesion of the recurrent nerves, previous surgery and radiotherapy, infectious causes or neurodegenerative diseases. In spite of the large differential diagnosis, literature describes a rate of vocal cord paralysis remaining idiopathic of 23% [5,6]. We retained a diagnosis of Gerhardt syndrome which will be further discussed.

A vocal cord paralysis is a frequent complication after a thyroid surgery [2,5-7]. The most frequent etiology for such an episode is

undoubtedly an iatrogenic injury of the recurrent laryngeal nerve. Based on Dankbaar et al. these injuries are responsible of 40% of unilateral paralysis and 50% of bilateral vocal cord paralysis [5]. In a more recent study, the incidence of laryngeal recurrent nerves' iatrogenic lesion, with a systematic perioperative anatomical identification of the nerve is reported at 4.1%. The article includes all types of thyroid surgery and is performed by a single specialized surgeon. The same study, reports a 21.7% rate of injury when a reoperation for a recurrent goiter is concerned [2]. A general rate of 10% to 14% of recurrent nerve paralysis is reported in literature after a surgery for a recurrent goiter.

This important rate is connected to the technical difficulty of the intervention mostly due to the presence of scar tissue in an altered anatomy. Other risk factors described in literature include malignancy due to the anatomical distortion and the possible invasion of surrounding tissues; and hyperthyroidism due to an important hypervascularity of the gland [1,2,6,8,9].

Our patient presented a recurrent goiter first addressed by cervical surgery which was reoperated by a thoracic open approach. Undoubtedly a hostile terrain which was altered by scar tissue, which constitutes an important risk factor for the development of a postoperative vocal cords' paralysis [2]. This difficulty was addressed by changing the surgical approach for the goiter resection. However, the goiter was in a retrosternal position and far of the normal anatomical course of the laryngeal recurrent nerves. Moreover, imaging assessment allowed us to exclude any complication in its trajectory. Thus, an iatrogenic injury was not retained. Furthermore, malignancy and hyperthyroidism were excluded in this case.

Some rarer etiologies for a recurrent nerve paralysis have been described, such as extrinsic tumors exerting a mass effect (as Pancoast tumors), infections such as poliomyelitis, inflammation and radiotherapy [5]. A paralysis might also be encountered in neurodegenerative diseases such as Parkinson or after a stroke. In this clinical case, the patient didn't present any symptoms making us suspect a neurodegenerative disease or an infection, thus these hypotheses were not retained.

Mediastinal postoperative oedema may lead to an inspiratory stridor and be confused with vocal cords paralysis. However, postoperative edema often resolves spontaneously and often doesn't require any further intervention. For this case, the complementary radiologic and endoscopic exams allowed us to exclude such etiology [6,10].

Gerhardt syndrome is another differential diagnosis for a bilateral vocal cords' paralysis. It manifests by an inspiratory dyspnea associated with a stridor and a discreet dysphonia, there is an elevated risk of aspiration and therefore pneumonia. The severity of dyspnea is variable depending on the degree of paralysis and the position of the vocal cords [2,3,5,6,8]. Nevertheless, in more extreme cases, the passage of air is too restrained leading to an acute asphyxia and a rapidly onset of respiratory distress [2,6,11].

Frequently, surgical intervention is needed to prevent acute asphyxiation. Diagnosis often requires imaging assessment and endoscopic examinations [11]. This syndrome was first described in 1863 [10] and has multiple etiologies and its physiopathology is not fully understood to this day [3,10]. In the beginning of the 20th century, Gerhardt syndrome was mostly described after syphilis infection, which became a much rarer etiology since the introduction of penicillin and the public health improvement in syphilis containment [10]. Nowadays, Gerhardt syndrome is often described following neurodegenerative diseases such as Shy-Drager syndrome or multisystemic atrophy or following a herpes simplex infection, causes that were excluded in this clinical case [3,10,12,13]. Indeed, as discussed, our patient didn't present any symptoms sustaining an infection or the occurrence of a neurogenerative disease.

More recent studies describe surgery as the leading cause for the development of Gerhardt syndrome [3,4,11]. Benninger et al. report up to 26% of cases of bilateral paralysis of vocal cords in adduction being related to surgery and the majority (70%) after thyroidectomy. The authors also mention other causes such as malignancy, intubation maneuvers, neurological diseases and previous radiotherapy [4].

Literature reports percentages as high as 59% for surgery as the cause of Gerhardt syndrome [3,11]. As far as our patient is concerned, multiple factors mentioned above are present, namely surgery, intubation and radiotherapy. Gerhardt syndrome being polyetiological, probably the combination of those factors contributed to the development of this abduction deficit. Our patient presented the typical clinical signs of Gerhardt syndrome which are a dyspnea, an inspiratory stridor and a slight dysphonia. The clinical presentation of an abduction deficit of vocal cords is very similar to any other cause of paralysis, making a clinical diagnosis of Gerhardt syndrome almost impossible. In fibroscopy it translates by a normal vocal cord morphology in a paramedian position in the glottis as a result of an abduction deficit, as described in the several endoscopic exams performed for our patient [3,10,11]. After exclusion of an iatrogenic lesion of the vocal cords and post operative edema by imaging assessment and endoscopic examinations, we could hypothesize the diagnostic of Gerhardt syndrome.

Being a rare syndrome not well described in literature, it is hard to make this diagnostic with full confidence, especially in rapidly evolutive situation with a threat of acute asphyxia. Indeed, it is a life-threatening condition that may lead to full airway obstruction needing surgical intervention and tracheostomy is still the gold standard for securing airways [3,10,11]. Less than 15% of patients suffering from Gerhardt can tolerate this condition without further intervention, but even for this minority, symptoms can be exacerbated by a simple intercurrent viral infection [3].

Chirila et al., report a rate of spontaneous recovery after a Gerhardt syndrome ranging from 40% to 86%, other studies report rates from 50% to 100% [3]. The expectation for a full spontaneous recovery varies in literature from 2 to 24 months [2,8,9]. A "watch and wait" attitude should be considered in this context, however, in some cases such as our patient, the threat of acute respiratory failure is imminent and serious, forcing a surgical intervention to prevent any pulmonary consequences of a chronic airway obstruction, or in extreme cases, death [2,3,14-16].

Conclusion

In conclusion, a bilateral vocal cord paralysis is a frequent complication after a thyroidectomy, mostly in a case of a recurrent goiter which is reoperated. Although iatrogenic injuries to the laryngeal recurrent nerves are the most frequent etiology, other causes must be considered even if they are hard to diagnose.

In this case, the Gerhardt syndrome corresponds to the first clinical presentation, fibroscopy findings and evolution. Nevertheless,

at the time of the events and at the urgency of the risk of an airway obstruction, other etiologies were first addressed. Knowledge of the diagnostic probably wouldn't have had an impact on medical care and would not have prevented a tracheotomy. The search for a diagnostic is time consuming and costly and can have a psychological impact for the patient, thus the importance of considering the vast differential diagnosis. It is our opinion that more studies are needed to have a better comprehension of its physiopathology and allow a faster diagnosis. In our case, even if the hypothesis of Gerhardt syndrome was mentioned during treatment of symptoms, the confirmation of this diagnostic was made after patient discharge. The delay diagnostic is probably related to a lack of information about this possibility because of the scarcity in literature. The aim of this case report is to put to light other diagnostic possibilities for a vocal cords' paralysis in a postoperative context.

References

1. Wagner HE, Seiler C. Recurrent laryngeal nerve palsy after thyroid gland surgery. Br J Surg. 2005;81(2):226-8.
2. Zakaria. Recurrent laryngeal nerve injury in thyroid surgery. Oman Med J. 2011;26(1):34-8.
3. Chirilã M, Mure R. Surgical management of Gerhardt syndrome. Chirurgia. 2010;105(3):327-30.
4. Benninger MS, Gillen JB, Altaian JS. Changing etiology of vocal fold immobility. Laryngoscope. 1998;108(9):1346-50.
5. Dankbaar JW, Pameijer FA. Vocal cord paralysis: Anatomy, imaging and pathology. Insights Imaging. 2014;5(6):743-51.
6. Holinger LD, Holinger PC, Holinger PH. Etiology of bilateral abductor vocal cord paralysis: A review of 389 cases. Ann Otol Rhinol Laryngol. 1976;85(4):428-36.
7. Gunn A, Oyekunle T, Stang M, Kazaure H, Scheri R. Recurrent laryngeal nerve injury after thyroid surgery: An analysis of 11,370 patients. J Surg Res. 2020;255:42-9.
8. Rovó L, Jóri J, Brzózka M, Czigner J. Airway complication after thyroid surgery: Minimally invasive management of bilateral recurrent nerve injury. Laryngoscope. 2000;110(1):140-4.
9. Chiang FY, Wang LF, Huang YF, Lee KW, Kuo WR. Recurrent laryngeal nerve palsy after thyroidectomy with routine identification of the recurrent laryngeal nerve. Surgery. 2005;137(3):342-7.
10. Dupuch V, Saroul N, Aumeran C, Pastourel R, Mom T, Gilain L. Bilateral vocal cord abductor paralysis associated with primary herpes simplex infection: A case report. Eur Ann Otorhinolaryngol Head Neck Dis. 2012;129(5):272-4.
11. Sapundzhiev N, Lichtenberger G, Eckel HE, Friedrich G, Zenev I, Toohill RJ, et al. Surgery of adult bilateral vocal fold paralysis in adduction: History and trends. Eur Arch Otorhinolaryngol. 2008;265(12):1501-14.
12. Bawa R, Ramadan HH, Wetmore SJ. Bilateral vocal cord paralysis with Shy-Drager syndrome. Otolaryngol Neck Surg. 1993;109(5):911-4.
13. Bannister R, Gibson W, Michaels L, Oppenheimer DR. Laryngeal abductor paralysis in multiple system atrophy: A report on three necropsied cases, with observations on the laryngeal muscles and the nuclei ambigui. Brain. 1981;104(2):351-68.
14. Rubin AD, Sataloff RT. Vocal fold paresis and paralysis: What the thyroid surgeon should know. Surg Oncol Clin N Am. 2008;17(1):175-96.
15. Palesse N, Marelli A, Legge MP. Bilateral abductor paralysis of the vocal cords in the course of neurological diseases: Report of 5 cases. Ital J Neurol Sci. 1988;9(1):59-62.
16. Eissler M, Holocher R, Lindenstrauss M, Wild K, Braun B. Autonome Dysfunktion mit nächtlicher Dyspnoe (Gerhardt-Syndrom) im Rahmen einer multiplen Systematrophie. Med Klin. 2001;96(10):626-31.