Annals of Clinical Cases

ISSN: 2692-7993

Case Report

Management of a Patient with Subglottic Stenosis due to Granulomatosis with Polyangiitis

Westphal FL[1], Pereira RMA[2], Pulner JGL[2], Lima LC[1], Lima Netto JC[3], Bendahan LB[4], Fonseca MSM[5] and Westphal Filho FL[2]*

[1]Department of Surgery and Postgraduate Program in Surgery, Universidade Federal do Amazonas, Manaus, Amazonas, Brazil

[2]Medical Graduation, Universidade Federal do Amazonas, Manaus, Amazonas, Brazil

[3]Departament of Surgery, Universidade Federal do Amazonas, Manaus, Amazonas, Brazil

[4]Postgraduate Program in Surgery, Universidade Federal do Amazonas, Manaus, Amazonas, Brazil

[5]Department of Internal Medicine, Universidade Federal do Amazonas, Manaus, Amazonas, Brazil

Abstract

Objective: Present the clinical case of a patient with subglottic stenosis of etiology originating from granulomatosis with polyangiitis.

Case details: A 28-year-old female patient, with a history of dyspnea on exertion for three years, reported, during the consultation, dyspnea on small efforts, occasional dry cough, wheezing, nasal congestion, pain in the face, and epistaxis for 18 months. Spirometry showed an obstructive pattern and bronchoscopy revealed subglottic tracheal stenosis. Tracheal dilations have opted. About a month after the dilation, the patient again presented the initial symptoms, and therefore, a computed tomography scan of the neck with reconstruction was requested, which showed new subglottic stenosis. To guarantee airway permeability, placement of a tracheobronchial endoprosthesis through bronchoscopy was performed. The patient has been with the orthosis for five years.

Final considerations: Endoprostheses allow the maintenance of airway patency, they can be placed endoscopy in a minimally invasive way. They are an effective treatment option for airway stenosis, resolving patients' respiratory symptoms immediately, showing good results in subglottic strictures.

Keywords: Granulomatosis with polyangiitis; Laryngostenosis; Prostheses and implants; Stents

Abbreviations

GPA: Granulomatosis with Polyangiitis; ANCA-c: Anti-Neutralophilic Anti-Cytoplasmic Body

Introduction

Granulomatosis with Polyangiitis (GPA) - formerly called Wegener's Granulomatosis- is a small vessel necrotizing vasculitis, which mainly affects kidney blood vessels, upper and lower airways [1-5]. Vasculitis manifests itself without sex predilection, with a higher incidence in the fifth decade of life, although it can occur at any age [1,5,6]. Previous estimations of incidence and prevalence have a wide variety due to the rarity of cases documented in studies; besides, there is a small amount of data available, regarding how ethnicity, gender, and age influences the development of the pathophysiology of the disease [7].

Immunopathogenesis has not been fully elucidated yet, however, granulomatous vasculitis shows signs of being promoted mainly by type 1 CD4 T lymphocytes [8].

Citation: Westphal FL, Pereira RMA, Pulner JGL, Lima LC, Lima Netto JC, Bendahan LB, et al. Management of a Patient with Subglottic Stenosis due to Granulomatosis with Polyangiitis. Ann Clin Cases. 2021; 2(2): 1031.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Feb 17th, 2021

***Corresponding author:** Fernando Luiz Westphal, Universidade Federal do Amazonas, Rua Afonso Pena 1053, Manaus, Amazonas, Brazil, Zip: 69020-160; E-mail: f.l.westphal@gmail.com

The clinical manifestations are broad, with the main involvement in the paranasal sinuses [9]. Renal involvement, through necrotizing focal segmental glomerulonephritis, occurs in 80% of patients [1]. Pulmonary involvement is characterized by cough, hemoptysis, dyspnea, chest pain, and the presence of nodules, infiltrates, and excavations, evident in 50% to 90% of patients [1,5]. Symptoms related to the upper airway are observed in more than 70% of patients, namely: sinusitis, rhinorrhea, epistaxis, chronic otitis media, saddle nose, perforated nasal septum, and nasal obstruction [1,5]. The presence of tracheal or endobronchial subglottic stenosis is observed in 16% of patients [1-3,5].

This study aims to present the clinical case of a patient with subglottic stenosis of etiology originating from Granulomatosis with Polyangiitis.

Case Presentation

Female patient, 28 years old, single, born, and living in Manacapuru - AM. The patient came to the pulmonology outpatient clinic with a history of dyspnea on exertion three years ago, at the time of the consultation she reported dyspnea on small exertions, occasional dry cough, wheezing, nasal congestion, pain in the face, and epistaxis for 18 months. The patient denied episodes of fever or weight loss in that period. Moreover, underwent drug therapy with the association of Tiotropium, Fluticasone, and Salmeterol, besides, reported to have been treated for sinusitis several times. Injuries, surgical procedures, orotracheal intubation, other comorbidities, and transfusions were denied as well. Regarding family history, the parents are alive and without comorbidities. On physical examination, tachypnea, audible laryngeal stridor, and saddle nose were noted. On pulmonary auscultation, the presence of physiological breath sounds without an adventitious pattern, with an audible extension of the laryngeal stridor.

The dosage of ANCA-c (Anti-Neutralophilic Anti-Cytoplasmic Body) was reactive, therefore positive. Spirometry showed an obstructive ventilatory disorder with a fixed obstruction pattern that did not respond to the bronchodilator. The tomography showed diffuse thickening of the walls of the subglottic larynx and could be related to the inflammatory/infectious process. Bronchoscopy showed subglottic tracheal stenosis with a diameter of 7.0 mm and, given the change, successive dilations were performed through the tracheal dilator starting with a diameter of 6.5 mm to 11.0 mm. Thus, the diagnosis of Granulomatosis with Polyangiitis was obtained, and the patient was referred to the rheumatology service.

About 1 month after the bronchoscopy, the patient presented, once again, the same initial symptoms; therefore, computed tomography of the neck with reconstruction was requested, which showed new subglottic stenosis. To guarantee airway patency, we opted for the placement of a tracheobronchial endoprosthesis through bronchoscopy. The patient has been in follow-up for 5 years with the orthosis. The stent was replaced 4 times during this period (Figure 1A and B, Figure 2).

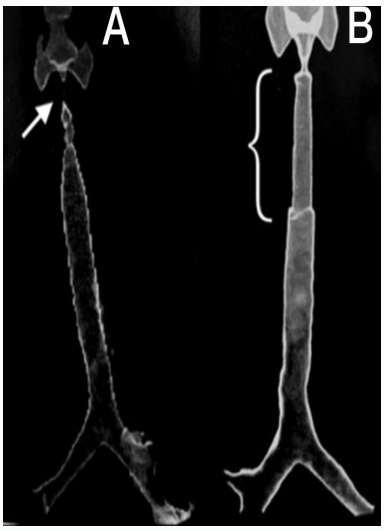

Figure 1: Computed tomography with tracheobronchial reconstruction A: Subglottic stenosis area (arrow). B: Endoprosthesis maintaining a patent airway (key).

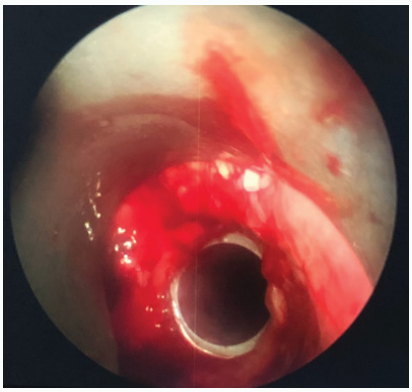

Figure 2: Bronchoscopy exam showing tracheal endoprosthesis.

Discussion

Approximately 16% to 23% of patients with GPA develop subglottic stenosis [1-3,5]. This is unusual, however, potentially lethal, literature does not have a well-explained etiology yet. Several suppositions could explain the predilection of the inflammatory reaction for this specific area, such as the fact that the subglottic area represents the junction of two different embryological centers. Besides, this area is the preferred location for the development of acute laryngitis in children, which would suggest a primary infection of this tissue as an initial event, but still without scientific evidence [10]. The involvement of the respiratory mucosa, as a single manifestation of the disease, occurs in approximately 25% of cases, with female patients, under the age of 30 years old, more prone to such presentation [11]. Subglottic stenosis tends to present at an earlier age, ranging from 26 to 40 years old, and to occur more frequently in the female sex (73%), differing from the epidemiological aspects of GPA [12]. Thus, the patient in the study would fit into the expected group.

The diagnosis of GPA is based on a combination of the clinical picture of systemic disease, which suggests a diagnosis of vasculitis; ANCA positive serology, and histological evidence of necrotizing vasculitis, necrotizing glomerulonephritis or granulomatous inflammation through organ biopsy usually affected such as skin, lung or kidneys. The classification criteria of the American College of Rheumatology involve nasal or oral involvement, presence of microhematuria with or without hematic cylinders, histological changes compatible with granulomatous inflammation in the arterial or perivascular wall, and typical radiographic changes (infiltrates, cavities, or pulmonary nodules). The presence of at least 2 of the criteria brings specificity of 92% and sensitivity of 88% [13]. The patient had several indications of the disease, including epistaxis, saddle nose, positive c-ANCA, and histology compatible with GPA.

To reduce tissue damage in affected organs, treatment was divided into an induction phase (to promote remission of the disease) and a maintenance phase (to maintain remission without relapse) [4,5].

The prognosis is directly related to the drug intervention, however, patients who progress to hemodialysis, due to renal impairment, have less prognostic value [14,15].

The recommended induction treatment consists of cyclophosphamide combined with systemic glucocorticoid. An option to cyclophosphamide in the case of severe and recurrent disease is Rituximab, which can be used in patients with contraindications to cyclophosphamide, such as in cases of high cumulative dose or young children of childbearing age without constituted offspring. In the case of both localized and systemic disease, but at an early stage, methotrexate can be used [4]. The patient underwent treatment with Rituximab and corticosteroids, with no result due to subglottic stenosis.

Subglottic stenosis does not follow GPA activity, evolving independently from other manifestations. Most recurrence episodes occur in periods of lower disease activity. Also, these relapses are common and frequent, which suggests a lack of response to classic GPA treatment [16]. The stenosis can be confirmed by the persistent decrease in the tracheal lumen in imaging exams, such as computed tomography, and by direct visualization by bronchoscopy [17]. This patient underwent bronchofibroscopy and dilation in the initial phase, but without success.

Tracheostomy was the solution to the problem until the mid-1980s, but with the modernization of endoscopic techniques, its use was restricted to emergencies and cases of severe laryngotracheal destruction. Thus, the therapy generally employed is intratracheal

dilation associated with long-term intralesional glucocorticoid injection, aiming to prevent scarring and restenosis. Such dilation can be achieved through blunt dilators, balloons, or lasers, the latter having the advantage of avoiding periods of intermittent apnea. Other treatment options are surgical reconstruction, the use of Mitomycin C, and the use of endoprostheses. In surgical reconstruction, the stenosed part is removed and an end-to-end anastomosis is performed, but due to frequent recurrences, there is a limitation in the possibility of tracheal resection. The application of Mitomycin C has presented contradictory results in different studies, requiring more research to define its role in treatment [10].

Endoprostheses allow airway patency to be maintained, in addition to further increasing its caliber, and can be placed by endoscopy in a minimally invasive manner. They are effective treatment options for airway stenosis, resolving patients' respiratory symptoms immediately [18]. Endoprostheses are most used in tracheobronchial strictures, however, positive results have been achieved in subglottic stenoses, and their use is an option to be considered in patients refractory to conventional treatments. The patient in this study has been using an endoprosthesis for 5 years [11,16,19,20].

The use of endoprostheses brings risks, for instance, the formation of granulation tissue, displacement, or perforation, however, these seem to have decreased with the advent of new technologies [18]. Endobronchial prostheses are more effective in the long term than laser ablation treatment or dilation, although their patency in this period is uncertain [21,22].

Thus, it is not yet possible to define an ideal course of treatment until the publishes of novel studies. However, this case study highlights the possibility of prolonged success of the endoprosthesis in the treatment of refractory patients.

References

1. Almouhawis HA, Leao JC, Fedele S, Porter SR. Wegener's granulomatosis: a review of clinical features and an update in diagnosis and treatment. J Oral Pathol Med. 2013;42(7):507-16.
2. Basu N, McClean A, Harper L, Amft EN, Dhaun N, Luqmani RA, et al. Explaining fatigue in ANCA-associated vasculitis. Rheumatology (Oxford). 2013;52(9):1680-5.
3. Cocho L, Gonzalez-Gonzalez LA, Molina-Prat N, Doctor P, Sainz-de-la-Maza M, Foster CS. Scleritis in patients with granulomatosis with polyangiitis (Wegener). Br J Ophthalmol. 2016;100:1062-5.
4. De Souza AWS, Calich AL, Mariz HDA, Ochtrop MLG, Bacchiega ABS, Ferreira GA, et al. Recomendações da Sociedade Brasileira de Reumatologia para a terapia de indução para vasculite associada a ANCA. Rev Bras Reumatol. 2017;57(Suppl 2):484-96.
5. Comarmond C, Cacoub P. Granulomatosis with polyangiitis (Wegener): Clinical aspects and treatment. Autoimmun Rev. 2014;13(11):1121-5.
6. Alba MA, Moreno-Palacios J, Beça S, Cid MC. Urologic and male genital manifestations of granulomatosis with polyangiitis. Autoimmun Rev. 2015;14(10):897-902.
7. Pearce FA, Grainge MJ, Lanyon PC, Watts RA, Hubbard RB. The incidence, prevalence and mortality of granulomatosis with polyangiitis in the UK Clinical Practice Research Datalink. Rheumatology (Oxford). 2017;56(4):589-96.
8. dos Santos SKJ, da Silva Raupp A, de Carvalho AAP, Bottega TS, Rogelin M, di Pietro D. Granulomatose de Wegener: importância do diagnóstico precoce. Relato de caso. Rev Bras Clin Med. 2009;7:427-33.
9. Santana LFE, Rodrigues MDS, Silva MPDA, Duarte RMSDC, Gomes OV. Granulomatose com poliangiite. Rev de Med. 2019;98(3):208-15.
10. Rasmussen N. Local treatments of subglottic and tracheal stenoses in granulomatosis with polyangiitis (Wegener's). Presse Med. 2013;42(4 pt 2):571-4.
11. Rodrigues AJ, Jacomelli M, Baldow RX, Barbas CV, Figueiredo VR. Comprometimento da árvore respiratória na granulomatose de Wegener. Rev Bras Reumatol. 2012;52(2):231-5.
12. Del Pero MM, Jayne D, Chaudhry A, Sivasothy P, Jani P. Long-term outcome of airway stenosis in granulomatosis with polyangiitis (Wegener granulomatosis): an observational study. JAMA Otolaryngol Head Neck Surg. 2014;140(11):1038-44.
13. Lutalo PMK, D'Cruz DP. Diagnosis and classification of granulomatosis with polyangiitis (aka Wegener's granulomatosis). J Autoimmun. 2014;48-49:94-8.
14. Araújo HFDA, Vilela DM, Laranjeira JVF, Fernandes BRC, dos Reis OL, dos Santos LI. Granulomatose com Poliangiite (Doença de Wegener): Apectos Relevantes de uma Doença Recém-Nomeada. Rev Interdis Ciênc Méd. 2020;4(1):62-8.
15. Franco CML, de Oliveira GMP, de Alencar Fidelix TS, Vieira LA, Trevisani VFM. Nodular scleritis and granulomatous polyangiitis (Wegener) mimicking tuberculosis. Rev Bras Oftalmol. 2015;74(2):106-9.
16. Girard C, Charles P, Terrier B, Bussonne G, Cohen P, Pagnoux C, et al. Tracheobronchial stenoses in granulomatosis with polyangiitis (Wegener's): A Report on 26 Cases. Medicine (Baltimore). 2015;94(32):e1088.
17. Morshed K, Trojanowska A, Szymański M, Trojanowski P, Szymańska A, Smoleń A, et al. Evaluation of tracheal stenosis: comparison between computed tomography virtual tracheobronchoscopy with multiplanar reformatting, flexible tracheofiberoscopy and intra-operative findings. Eur Arch Otorhinolaryngol. 2011;268(4):591-7.
18. Minamoto H, Terra RM, Cardoso PFG. Estenoses benignas da via aérea: tratamento endoscópico. Pulmão RJ. 2011;20(2):48-53.
19. Costantino CL, Niles JL, Wright CD, Mathisen DJ, Muniappan A. Subglottic Stenosis in Granulomatosis With Polyangiitis: The Role of Laryngotracheal Resection. Ann Thorac Surg. 2018;105(1):249- 53.
20. Terrier B, Dechartres A, Girard C, Jouneau S, Kahn JE, Dhote R, et al. Granulomatosis with polyangiitis: Endoscopic management of tracheobronchial stenosis: Results from a multicentre experience. Rheumatology (Oxford). 2015;54(10):1852-7.
21. Anjos DMD, Junqueira JJM, Gomes DBD, Terra RM, Minamoto H, Jatene FB. Endopróteses: opção terapêutica para estenoses traqueobrônquicas adquiridas. Rev de Med. 2007;86(3):174-84.
22. Tiernan J, Shah C, McGuigan J, Elborn JS. Successful stenting in endobronchial Wegener's granulomatosis. Ulster Med J. 2006;75(2):155-57.