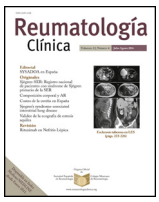
Reumatol Clin. 2016;12(5):267–273

Sociedad Española de Reumatología - Colegio Mexicano de Reumatología

Reumatología Clínica

www.reumatologiaclinica.org



Brief report

Subglottic Stenosis in Granulomatosis With Polyangiitis (Wegener's Granulomatosis): Report of 4 Cases☆

Gabriel Horta-Baas,[a,*] María Fernanda Hernández-Cabrera,[b] Rocío Catana,[c] Mario Pérez-Cristóbal,[c] Leonor Adriana Barile-Fabris[c]

[a] *Servicio de Reumatología, Hospital General Regional 220, Instituto Mexicano del Seguro Social, Toluca, Estado de México, Mexico*
[b] *Servicio de Reumatología, Centro Médico ISSEMYM, Toluca, Estado de México, Mexico*
[c] *Servicio de Reumatología, Centro Médico Nacional Siglo XXI, Instituto Mexicano del Seguro Social, Distrito Federal, Mexico*

ARTICLE INFO

Article history:
Received 24 March 2015
Accepted 30 October 2015
Available online 25 July 2016

Keywords:
Subglottic stenosis
Granulomatosis with polyangiitis
Wegener's granulomatosis

ABSTRACT

Introduction: Subglottic stenosis (SGS) in granulomatosis with polyangiitis (GPA) may result from active disease or from chronic recurrent inflammation. The objective of the study was to describe the clinical features and treatment of patients with subglottic stenosis.
Methods: We retrospectively reviewed the medical records of all patients with SGS due to GPA diagnosed at Rheumatology department between January 2000 and June 2015.
Results: We present 4 cases of SGS at our department during a period of 15 years. The interval between the presentation of the GPA and SGS varied between 2 and 144 months. The leading symptoms of SGS were dyspnea on exertion and stridor. Three patients presented SGS without evidence of systemic activity. Two patients presented SGS grade I and received tracheal dilatation; two recurred and three needed a tracheostomy due to severe airway-limiting stenosis.
Conclusion: SGS presents high morbidity. Even though subglottic dilatation provides symptomatic relief, recurrences may present. Severe airway-limiting stenosis often requires tracheostomy.

© 2015 Elsevier España, S.L.U. and Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. All rights reserved.

Estenosis subglótica en granulomatosis con poliangitis (granulomatosis de Wegener): presentación de 4 casos

Palabras clave:
Estenosis subglótica
Granulomatosis con poliangitis
Granulomatosis de Wegener

RESUMEN

Introducción: La estenosis subglótica (ESG) en la granulomatosis con poliangitis (GPA) puede ser consecuencia de la enfermedad activa o de procesos inflamatorios repetitivos. Nuestro objetivo es describir las características clínicas y el tratamiento de los pacientes con ESG.
Métodos: Estudio descriptivo retrospectivo de los casos diagnosticados durante el período comprendido entre el 1 de enero del 2000 y el 1 de junio del 2015.
Resultados: Presentamos 4 casos; la ESG se presentó entre los 2 y 144 meses del diagnóstico de la GPA, los síntomas de presentación fueron disnea de esfuerzo y el estridor laríngeo, 3 desarrollaron ESG en ausencia de actividad sistémica. Dos sujetos con ESG grado I fueron tratados con dilatación traqueal, 2 casos presentaron reestenosis y en 3 casos fue necesario la realización de traqueostomía.
Conclusión: La ESG presenta una alta morbilidad. La dilatación endoscópica proporciona alivio sintomático; sin embargo, suelen existir recidivas de la estenosis. La obstrucción grave de la vía aérea a menudo requiere de traqueostomía.

© 2015 Elsevier España, S.L.U. y Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. Todos los derechos reservados.

☆ Please cite this article as: Horta-Baas G, Hernández-Cabrera MF, Catana R, Pérez-Cristóbal M, Barile-Fabris LA. Estenosis subglótica en granulomatosis con poliangitis (granulomatosis de Wegener): presentación de 4 casos. Reumatol Clin. 2016;12:267–273.

* Corresponding author.
E-mail address: gabho@hotmail.com (G. Horta-Baas).

2173-5743/© 2015 Elsevier España, S.L.U. and Sociedad Española de Reumatología y Colegio Mexicano de Reumatología. All rights reserved.

Introduction

Granulomatosis with polyangiitis (GPA) is a systemic autoimmune disease of unknown etiology. It is characterized by necrotizing granulomatous inflammation of the respiratory tract and vasculitis that affects small- and medium-sized blood vessels.[1] Stenosis of subglottis and proximal trachea (SGS) can result from active disease or from recurrent inflammatory processes. An incidence of 8%–23% has been reported during the course of GPA, and it can be the first manifestation in 1%–6% of the patients.[1–6] The objective of this article is to describe the clinical characteristics and treatment of patients with SGS.

Materials and Methods

We conducted a retrospective descriptive study of the cases diagnosed during the period between 1 January 2000 and 1 June 2015 in the Centro Médico Nacional Siglo XXI, a tertiary care referral center in Distrito Federal, Mexico. In order to compare our findings with the results of other published series, we performed a search of the available medical literature by means of a systematic review in the PubMed and EMBASE databases. It was limited to articles published in English or Spanish, using the following search terms: "subglottic stenosis", "tracheal stenosis", "Wegener's granulomatosis" and "granulomatosis with polyangiitis". We included those studies that provided demographic data, the level of disease activity, the description of the therapy employed and the outcomes. Case reports were excluded.

Results

We present 4 cases of SGS in patients, predominantly women, with GPA; the presenting symptoms were exertional dyspnea and laryngeal stridor in every case; SGS was diagnosed between 2 and 144 months after the onset of GPA; 3 patients developed SGS in the absence of systemic activity; all the patients had chronic sinusitis. Three individuals had saddle nose, and 1 showed no evidence of antineutrophil cytoplasmic antibodies (Table 1). Two patients had grade I SGS and were treated with glucocorticoids and tracheal dilatation; restenosis developed in 2 patients, and 3 required tracheostomy due to severe airway compromise (grades III and IV SGS according to the Myer-Cotton grading system). During follow-up, decannulation was possible in 1 patient.

Patient no. 1

A 73-year-old woman who had been diagnosed with GPA at the age of 57 years was being treated with methotrexate at 15 mg/week and prednisone at 5 mg/day. She presented with dry cough, laryngeal stridor and dyspnea. Laryngoscopy revealed SGS of 50%; she had a score of 6 on the Birmingham Vasculitis Activity Score (BVAS). She was treated with 8 mg intravenous dexamethasone and 0.5 mg nebulized budesonide every 12 h, and her symptoms improved with no additional procedures. Six months later, laryngoscopy revealed SGS of 30% and no further treatment was required.

Patient no. 2

A 64-year-old woman who had been diagnosed with GPA at the age of 57 years. At the age of 59, she presented with dyspnea and stridor. Computed tomography (CT) of larynx and trachea revealed a granulomatous lesion obstructing the tracheal lumen (Fig. 1). Bronchoscopy disclosed an obstruction of 75% in subglottis, and tracheal dilatation and tracheostomy were performed. Two months later, the patient presented with restenosis of 85% requiring tracheal dilatation, and 2 weeks after that, SGS of 50% was detected, and tracheoplasty was performed using the technique of Grillo and Pearson. The patient required repeat dilatation 18 months later due to SGS of 80%. Fifteen months after that last dilatation, bronchoscopy revealed SGS of 30%, and resolution of dyspnea and stridor. As restenosis did not occur over the following 7 months, decannulation was possible.

Patient no. 3

A 44-year-old man had been diagnosed with GPA at the age of 39 years. Two months after the diagnosis, he presented with laryngeal stridor, dyspnea, foreign body sensation in his larynx and dysphonia. Fiberoptic laryngoscopy revealed SGS of 20% with systemic activity (BVAS score of 23). He was treated with 3 doses of 1 g/day of methylprednisolone and intravenous cyclophosphamide; the systemic manifestations remitted, but his dysphonia, stridor and dyspnea progressed. One month later, CT revealed concentric circumferential thickening that partially obstructed the tracheal lumen (Fig. 2 A and B). Bronchoscopy showed SGS of 70%, with erythematous laryngeal structures. He received 3 doses of 1 g/day of methylprednisolone and underwent tracheal dilatation. Subsequently, the clinical course was satisfactory, with remission of the stridor and improvement in the symptoms. However, 7 days later, his symptoms recurred, with 100% restenosis (Fig. 2C and D), which required emergency tracheostomy.

Patient no. 4

A 17-year-old woman had been diagnosed as having GPA at the age of 11 on the basis of pulmonary and renal involvement. Eighty-four months after the diagnosis of GPA, she presented with a 2-month history of dysphonia, laryngeal stridor, dry cough and dyspnea, and a BVAS score of 3. Computed tomography revealed tracheal stenosis measuring 6 cm in length, and a transverse diameter at the narrowest portion of 5 mm. Tracheostomy was performed due to airway compromise and, after 2 months of follow-up, decannulation remained impossible.

The literature search yielded 77 studies, 12 of which evaluated the treatment of SGS in patients with GPA and met the selection criteria. As the study published by Langford et al.[5] included the patients described in the study of Lebovics et al.,[7] they were grouped in a single study. Table 2 summarizes the major findings of these studies.

Discussion

Subglottic stenosis occurs more frequently in women and young individuals (Table 2).[4,7–12] The incidence of SGS was higher among patients with GPA onset during childhood or adolescence rather than during adulthood (48% vs 10%, $P<.001$).[7] The subglottic region of the trachea is particularly susceptible to narrowing because of its small diameter, its lack of distensibility, the fragility of the tissue that lines it and its poor vascularization, which, if damaged, heals concentrically, further reducing the lumen.[13]

Subglottic stenosis is the result of inflammation, edema and fibrosis that typically extends from 3 to 4 cm below the vocal chords.[13] The active phase of GPA is accompanied by involvement of the tracheal mucosa, occasionally in the form of ulcers. As the vocal chords are seldom affected, in most cases the symptoms are mild or nonexistent. If the subclinical tracheal inflammation is not treated, fibrotic scar tissue forms within a variable period of time (the average is reported to be 39–60 months); when the lumen is already significantly compromised, dyspnea, cough, voice changes and stridor appear.[2,10,14] Patients with SGS associated with GPA have been reported to have a higher incidence of paranasal sinus

Download English Version:

https://daneshyari.com/en/article/3384012

Download Persian Version:

https://daneshyari.com/article/3384012

Daneshyari.com