REVIEW AND CASE REPORTS

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Subglottic stenosis in granulomatosis with polyangiitis: clinical, histopathological and therapeutic considerations with presentation of two cases

BIANCA RUSU[1,2], GABRIELA CORNELIA MUŞAT[2,3], CARMEN AURELIA MOGOANTĂ[4], ALEX IULIAN MILEA[1–3], CAIUS CODRUŢ SARAFOLEANU[2,3]

[1] *Doctoral School, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[2] *Department of Otorhinolaryngology, Carol Davila University of Medicine and Pharmacy, Bucharest, Romania*

[3] *Department of Otorhinolaryngology and Head and Neck Surgery, Sf. Maria Hospital, Bucharest, Romania*

[4] *Department of Otorhinolaryngology, Faculty of Medicine, University of Medicine and Pharmacy of Craiova, Romania*

Abstract

Granulomatosis with polyangiitis (GPA), once known as Wegener's granulomatosis (WG), is an uncommon systemic vasculitic disorder characterized by inflammation of the vascular walls, particularly affecting the respiratory tract. One serious complication is subglottic stenosis (SGS), a narrowing of the airway just below the vocal cords. While uncommon, it can be life-threatening and may show up as stridor, dyspnea, or voice changes. Sometimes, it is the first or only sign that someone has GPA. Diagnosing this condition takes a combination of clinical evaluation, imaging, endoscopy, and blood tests for specific antibodies [anti-neutrophil cytoplasmic antibodies (ANCA)]. Tissue biopsy is especially valuable, it can confirm the diagnosis by showing the characteristic inflammation patterns, particularly when the disease presents in unusual ways. Treatment typically involves both systemic immunosuppressive medications and, when necessary, procedures to open up the narrowed airway. We present two cases of GPA-associated SGS that highlight how differently this condition can present, the challenges of managing a compromised airway, and the need for tailored treatment plans. These cases reinforce why early detection of airway involvement matters, why patients need careful long-term monitoring to watch for re-narrowing, and why a team-based approach across specialties leads to the best outcomes.

Keywords: granulomatosis with polyangiitis, subglottic stenosis, ANCA-associated vasculitis, airway obstruction, histopathology, multidisciplinary management.

Introduction

Granulomatosis with polyangiitis (GPA), previously termed Wegener's granulomatosis (WG), is a systemic inflammatory disorder [1]. The condition was initially documented by Klinger in 1931 [2] and subsequently characterized in greater detail by Wegener in 1936 [3]. As defined by the *2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides* (CHCC), GPA is distinguished by necrotizing granulomatous inflammation predominantly affecting the upper and lower respiratory tract, in conjunction with necrotizing vasculitis, primarily involving small to medium-sized vessels [4]. The *American College of Rheumatology* (ACR) delineates the diagnostic criteria for GPA, requiring the existence of at least two of the following features: ulcerative lesions of the oral mucosa or nasal bleeding/inflammation, abnormal findings on chest radiography such as nodules, fixed pulmonary infiltrates, or cavities, abnormal urinary sediment, and histopathological (HP) evidence of granulomatous inflammation [5]. GPA is an uncommon disease, with reported annual incidence rates ranging from 3 to 14.4 cases per million individuals, while reports of prevalence range from 23.7 to 156.5 cases per million [6]. According to Greco *et al.* [7], there are roughly three instances for every 100 000 people in the US and five cases for every 100 000 people in Europe. Nearly 90% of cases involve the head and neck, with the most common areas affected being the mouth, nose, eyes, and ears [8].

Clinical presentation

GPA typically starts as a localized condition that could spread to several tissues or organs at an unknown rate if left undiagnosed and untreated [9]. Although modest symptomatology is typically noticed at first, constitutional symptoms such as general malaise, myalgia, arthralgia, anorexia, weight loss, and pyrexia might be observed. The upper respiratory tract, lungs, and kidneys are the most frequently affected organs, with involvement reported in approximately 88.7%, 71.3%, and 61.5% of patients, respectively [10].

In rare cases, involvement of the airway may represent only clinical manifestation or the initial presentation of GPA. Laryngeal and tracheal ulcerations are reported in approximately 25% of patients, whereas subglottic stenosis (SGS) is less commonly encountered, with an estimated prevalence of 16% according to data reported by the

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National Institutes of Health (NIH) [11]. Among these patients, half required a tracheostomy [12]. If left untreated, severe SGS can lead to critical airway obstruction, which may quickly become life-threatening [13]. The underlying cause of SGS in GPA remains unclear. This anatomical region represents the junction of two embryological growth centers and is characterized by two distinct microcirculatory pathways. While the trachea above and below the stenosis are typically intact, the stenoses are typically located at the intersection of this area and the true vocal folds [14]. Hoarseness, coughing, hemoptysis, dyspnea, stridor, and wheezing are typical symptoms [15–17]. Hoarseness and cough are less frequent than dyspnea and wheezing and are noted in less than 10% of patients with airway stenosis caused by GPA [15].

Given that SGS presents with nonspecific symptoms, a heightened level of suspicion is crucial for its identification, particularly in the absence of other indicators of active GPA. The symptoms associated with SGS may overlap with those of several other respiratory disorders, and stridor may be mistaken for the wheezing commonly observed in bronchial asthma [18]. Given the potential for SGS to result in life-threatening airway obstruction, patients with suspected airway involvement should undergo prompt evaluation by an otolaryngologist [12, 19, 20]. The diagnosis must be established through the identification of vasculitis, granulomatous inflammation, and necrosis within a clinical context. Nevertheless, these HP characteristics are frequently variable, and a negative biopsy does not definitively rule out the diagnosis [16, 19, 21]. Assessment of the nasal cavity and paranasal sinuses is recommended in all patients with suspected GPA. Granulomatous lesions in these regions represent suitable sites for obtaining diagnostic tissue samples. Nevertheless, in advanced or chronic disease, extensive scarring may lead to sinus obliteration, and computed tomography (CT) or magnetic resonance imaging (MRI) findings may not reliably distinguish active granulomatous inflammation from established structural damage. Upon diagnosis of GPA-related SGS, patients require vigilant monitoring for indications of airway compromise. The severity of stenosis can be accurately assessed using endoscopic examination, radiographic determination of the stenotic segment's length, and flow-volume loop investigations [19, 21].

Diagnostic evaluation

Indirect or flexible fiberoptic laryngoscopy

When SGS is suspected, an initial endoscopic examination using indirect or flexible fiberoptic laryngoscopy should be performed by an experienced otolaryngologist. This minimally invasive method allows visualization of the laryngeal structures; however, it typically does not permit complete assessment of the trachea [20].

Fiberoptic bronchoscopy

Flexible bronchoscopy gives a closer look at the airway, allowing the estimate of how much the lumen has narrowed and to take tissue samples to confirm the diagnosis. The challenge is that in cases of severe stenosis, it may be impossible to advance the scope past the tight spot, which means it is not going to be always possible to measure the exact length of the narrowed segment. This matters because the length of the lesion plays a big role in deciding how to treat it; shorter segments tend to be better suited to less invasive approaches.

Looking at the laryngeal mucosa during endoscopy can also give us hints about whether the disease is still active. Sometimes one can tell the difference between fresh inflammatory changes and older fibrotic scarring just by appearance, though what it is seen on the surface doesn't always match what the pathologist finds under the microscope [19]. When inflammation is active, the elements that can be seen are some circumferential reddened tissue that bleeds easily, irregular granulomatous masses partially blocking the airway, or ulcerated areas.

Histopathological assessment

Biopsy of the stenotic segment is considered mandatory, despite technical challenges associated with sampling from the upper airway. Histological analysis frequently fails to reveal classical diagnostic features of GPA, such as necrotizing vasculitis of small vessels, epithelioid granuloma formation, or fibrinoid necrosis, and instead commonly demonstrates nonspecific chronic inflammation and fibrosis [16, 19]. For this reason, obtaining multiple biopsies from visibly inflamed areas is recommended to increase diagnostic yield.

Imaging studies: MRI and tracheal CT

MRI and CT scan of the trachea can be useful additions to the workup, especially when planning a bronchoscopy or therapeutic procedure. That said, these two investigations shouldn't be the first thing to achieve for they are not ideal at picking up lesions directly, and relying on them too early can lead to delays in getting to the diagnosis [22]. When imaging does show abnormalities, the elements seen are circumferential thickening of the tracheal lining, an irregular mucosal surface, or ulceration. Sometimes there are irregular masses bulging into the airway and causing partial obstruction. Cartilage involvement isn't common, but when it happens, it can distort the structure of the trachea and lead to progressive narrowing.

Spiral CT with three-dimensional reconstruction of the laryngotracheal airway, including virtual bronchoscopy, adds another layer of detail that complements what can be seen on conventional endoscopy. It allows for more precise measurement of stenosis severity and extent, clear identification of where the lesion starts and ends, and evaluation of the airway beyond the obstruction. This is particularly helpful when planning treatment, tracking response, or following airway patency over time [23–25]. If serial imaging shows that a stenosis is getting worse or becoming unstable, that's often the trigger for referring the patient for interventional bronchoscopy [24].

Virtual bronchoscopy does have its drawbacks, though. It doesn't allow for taking tissue samples, and it's not reliable for telling the difference between active inflammation and old structural damage. For these reasons, direct endoscopic examination remains the gold standard when a complete picture of what is going on in the airway is needed. Virtual bronchoscopy is best thought of as a helpful complement, not a replacement.

Biomarkers

Anti-neutrophil cytoplasm antibodies (ANCA) testing aids in diagnosis and is especially beneficial in cases with inexplicable SGS. The cause is confirmed by a positive ANCA test [26]. In active multisystem illness, the cytoplasmic staining of ANCA (c-ANCA) has a sensitivity of 91% and a specificity of 99%. Perinuclear staining is indicated by the p-ANCA, which is less often positive (between 20% and 40%).

Differential diagnosis of subglottic stenosis

Before settling on a diagnosis of SGS, it is important to rule out other possible causes, management can vary considerably depending on the underlying etiology, and missing something can lead to worse outcomes [12, 27, 28]. The differential diagnosis includes scarring from previous intubation or tracheostomy, tracheal rupture, external compression from thyroid tumors or large goiters, congenital airway abnormalities, gastro-esophageal reflux, infections, other inflammatory conditions, benign or malignant tumors, and idiopathic progressive SGS.

Congenital stenosis typically presents during childhood and is commonly linked to complete tracheal rings resulting from the fusion of the posterior cartilaginous structures. Some examples of primary benign tracheal tumors are chondromas, fibromas, papillomas, hemangiomas, and granular cell tumors. They can cause airway narrowing, but they are not very common. Other pathologies that should be considered are the infectious causes, such as tuberculosis or fungal infections like histoplasmosis and blastomycosis, especially in patients on immunosuppressive therapy, with serological tests and histopathology assisting in diagnosis.

Relapsing polychondritis, primary amyloidosis, or sarcoidosis, that are non-infectious inflammatory disorders, may also result in SGS and can generally be ruled out through a combination of clinical evaluation and HP examination [28]. A careful look at these other possible causes makes sure the right diagnosis is made, and the right treatment is given.

Treatment of subglottic stenosis

Although SGS treatment is still a therapeutic conundrum, management varies depending on the type of stenosis [12]. Nearly every non-medical treatment is an invasive operation. Treatment options include intraluminal stent implantation, balloon dilation, laser ablation, translesional corticosteroid injection, surgical resection, and reanastomosis [17, 19, 29–32]. Standard medical treatment for acute disease-related short stenosis can include inhaled topical glucocorticoids. Laser therapy may be suitable for certain patients [20]. It is utilized with radial incisions to lessen the stress on the stenosis surface since total excision of the stenosis causes further stenosis. Only in rare instances of a narrow web is dilation therapy effective [31]. To prevent chronic tracheostomy, surgical correction may be required when persistent damage develops in fixed lesions [12]. When the stenoses are localized to the cricoid, many grafting methods are possible [20, 31]. During the healing period (3–6 months), stenting may occasionally be required. Alternatively, the tracheostomy may be maintained to permit safe air circulation *via* the larynx [33]. It is important to keep in mind the three main points about tracheobronchial lesions. Initially, it is best to postpone any surgical procedures until the initial inflammation has subsided while receiving medication. Second, it is important to reduce any further tissue damage that may arise from the treatment. Third, when airway stenting is considered necessary in selected patients, only silastic (silicone) stents should be used. The use of metallic wire-mesh stents should be avoided in patients with GPA.

Therapy that suppresses the immune system

The use of systemic glucocorticoids alongside with immunosuppressive drugs has significantly lowered morbidity and mortality from active GPA that affects major organ systems [34–36]. This treatment has also been used to treat SGS caused by the condition, and in some cases, it has been shown to help [17]. However, subglottic involvement often shows little or no response to systemic immunosuppression, and up to three-quarters of patients who are impacted ultimately need interventional treatments to see significant relief in their symptoms [12, 14, 37, 38].

Moreover, disease recurrence at the subglottic level frequently occurs after a reduction or interruption of systemic therapy, typically requiring repeated treatment sessions. This elevates cumulative exposure and the risk of therapy-related side effects, including the onset or advancement of fibrotic alterations within the airway [12, 14, 37–39]. The limited efficacy of immunosuppressive drugs in isolated SGS is further highlighted by the fact that SGS can occur despite continuous systemic treatment for other indications of GPA [21, 37].

These factors collectively cast doubt on the efficacy of systemic monotherapy for subglottic lesions. Consequently, interventional airway procedures, whether used as independent treatments or in combination with traditional medical therapy, represent a more suitable therapeutic approach in numerous instances [12–14, 16, 37–44].

Interventional procedures

As mentioned earlier, there are several ways to treat SGS, ranging from endoscopic approaches to open surgery. Options include airway dilation, conservative endoscopic techniques, laser-based procedures, stent placement, and in certain cases, surgical removal of the affected segment with reconstruction [12–14, 16, 37–44]. Endoscopic dilation using a rigid bronchoscope or tracheoscope is a minimally invasive option that works well in both planned and emergency settings [38]. It is generally well tolerated, requires only a short hospital stay, and can be repeated as needed over time. Routine antibiotics or systemic corticosteroids are not usually necessary, and complications from the procedure are uncommon.

Langford *et al.* described a specific technique that combines intralesional injection of long-acting corticosteroid with dilation, sometimes referred to as intratracheal dilation injection therapy or intralesional corticosteroid injection and dilation (ILCD). This approach has been reported as both safe and effective for treating GPA-associated SGS [19]. When there is no active involvement of major organs, it can be used without the need for additional systemic immunosuppression. Although large randomized controlled

trials are lacking, several authors have suggested ILCD as a preferred treatment option for this condition [45]. The rationale is that the two components work together: the local corticosteroid injection damps down inflammation and limits fibroblast activity and scar formation, while the mechanical dilation breaks up existing fibrotic tissue. This combined effect may explain why the technique seems to work across a range of GPA-related subglottic lesions, whether inflammation or fibrosis is the dominant feature.

One of the advantages of this approach is that it reduces treatment-related toxicity and may help maintain airway patency for longer by avoiding the need for prolonged or additional systemic immunosuppression. Reassuringly, airway collapse due to laryngotracheomalacia following repeated intralesional corticosteroid injections has not been reported.

Mitomycin C, an alkylating agent known to inhibit fibroblast proliferation and extracellular matrix production [46], has also gained traction as an adjunctive treatment in selected cases of GPA-related tracheal stenosis. It has been used following ILCD [14, 16, 45] or alongside laser surgery and dilation [43, 46, 47], with the aim of reducing fibrotic remodeling and lowering the risk of restenosis. That said, evidence supporting its long-term effectiveness remains limited. A randomized, double-blind, placebo-controlled trial evaluating repeated Mitomycin C applications in patients with laryngotracheal stenosis failed to demonstrate lasting benefit in preventing restenosis [47]. Because of this, some authors recommend reserving its use for cases with active inflammatory disease rather than established fibrosis [38].

Conserving endoscopic procedures

When active tracheal involvement in WG results in airway blockage, endoscopic excision of granulomatous tissue, whether by standard surgery or laser methods, is a successful method to relieve the obstruction and often eliminates the need for stenting or bypass treatments. This technique can be used alone or in conjunction with tracheal dilatation [43, 48]. Carbon dioxide (CO_2) and neodymium-doped yttrium aluminum garnet (Nd:YAG) lasers have been used to treat patients with WG-related SGS, however the outcomes have varied in the literature [12, 16, 41]. Multiple laser sessions have been demonstrated to produce positive effects in certain studies [16, 41], however stenosis returns fast following treatment, according to another research [12].

In cases of short-segment stenosis, laser therapy appears to be more effective [12], and it is generally contraindicated during active disease stages [16]. Severe heat damage to the subglottic mucosa is one potential issue that could result in a robust fibroblastic response [45, 49]. Consequently, cicatrix development and post-laser scarring have been reported as limitations of this method [12, 49].

Stenting

There have been few studies on using stents to treat SGS, and the results are not always the same [12, 15, 50, 51]. Silicone stents are usually preferred because metal stents are hard to take out and can break down into nearby tissues. Careful thought is needed because they could make emergency airway procedures, like tracheostomy or intubation, more difficult [51].

Biological therapies

Biological therapies have been investigated as potential treatments for systemic vasculitis, but results with tumor necrosis factor-alpha (TNF-α) inhibitors have been disappointing. The addition of Infliximab to conventional treatment has been associated with a substantial risk of infectious complications, including tuberculosis, whereas Etanercept has been linked to an increased incidence of both infections and solid malignancies [52, 53]. In contrast, accumulating evidence indicates that B-cell depletion with Rituximab, a chimeric monoclonal antibody directed against cluster of differentiation 20 (CD20)-positive B-cells, may provide a beneficial therapeutic effect. Rituximab rapidly depletes circulating B-cells, reduces ANCA production, and can induce disease remission [54, 55]. However, available evidence indicates that Rituximab, like standard immunosuppressive therapies, appears to be less effective in refractory granulomatous manifestations such as SGS than in vasculitic disease manifestations [54–56]. Therefore, further research is needed to better understand the immune mechanisms involved and to develop more precisely targeted biological therapies.

Case presentations

We report two patients with GPA who developed troublesome narrowing of the airway, one at the subglottic level, the other extending further into the trachea. Both diagnoses were supported by clinical findings and positive c-ANCA serology. To get a clear picture of where the stenosis was, how far it extended, and how severe it had become, both patients underwent flexible fiberoptic laryngoscopy and contrast-enhanced CT scanning. Treatment was tailored to each patient's situation. The first patient had a short but severe segment of SGS, he needed a tracheostomy to protect her airway, followed by CO_2 laser resection of the narrowed area. We kept her on Rituximab and temporarily raised her corticosteroid dose given her active disease. The second patient had a longer stretch of narrowing, which we managed with two endoscopic sessions using electrosurgery and dilation, later clearing away fibrinous material that had built up as part of the healing process. Both patients were followed closely with clinical assessments, repeat endoscopy, and imaging to keep an eye on how well the airway was holding open, whether symptoms had resolved, and whether any further procedures might be needed.

Case No. 1

A 59-year-old man came to our Clinic with a two-month history of progressing dyspnea, dysphonia, and inspiratory stridor. He had been diagnosed with GPA four years earlier, at age 55, based on strongly positive c-ANCA levels and a nasal mucosal biopsy showing granulomatous inflammation with necrosis and necrotizing vasculitis, findings confirmed by the pathologist (Figure 1, A and B). At the time of presentation, he was on maintenance therapy with Methylprednisolone and Rituximab. His medical history was marked by several GPA-related complications: chronic rhinosinusitis, hearing loss on the left side, episcleritis, anterior uveitis and keratitis in his right eye, and glomerulonephritis. Blood tests showed he remained c-ANCA-positive (Figure 1, A and B).

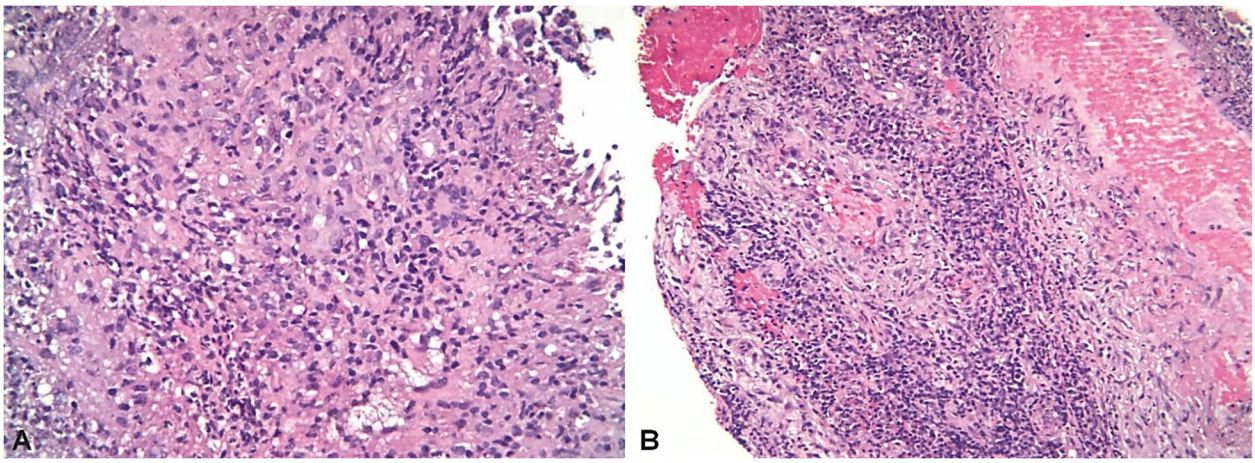

Figure 1 – ***(A and B) Biopsy fragments of mucosa lined by squamous epithelium showing reactive changes and extensive ulceration, with granulation tissue, a polymorphous inflammatory infiltrate, and focal underlying reactive fibroconnective tissue; limited granulomatous inflammation with rare multinucleated giant cells, areas of suppurative necrosis, and vasculitic lesions, focally with a leukocytoclastic pattern. Hematoxylin–Eosin (HE) staining: (A) ×100; (B) ×40.***

We obtained a contrast-enhanced CT of his neck (Figure 2, A and B), which revealed thickening of the tracheal wall just below the cricoid cartilage, partially blocking the airway. On clinical examination and fiberoptic laryngoscopy, there was significant swelling of the ventricular bands and both arytenoid cartilages, with edematous vocal cords and severe SGS, Grade III on the Myer–Cotton scale, leaving him with a critically narrow airway of just 3 mm (Figure 3A).

With the diagnosis of GPA-associated SGS confirmed, we proceeded with surgical intervention. A tracheostomy was performed first to secure the airway given the severity of the narrowing, this allowed safe ventilation and good surgical access while minimizing the risk of losing the airway during the procedure. We then used CO_2 laser to resect the stenotic segment (Figure 3B).

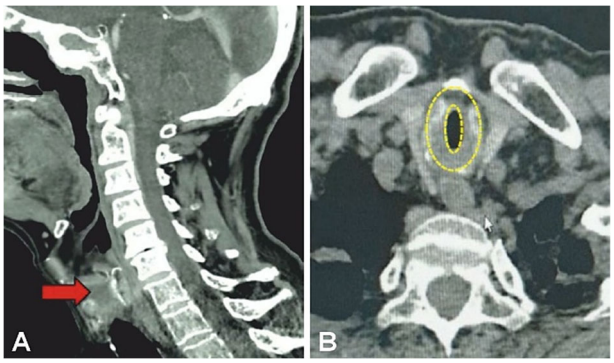

Figure 2 – ***(A and B) Contrast-enhanced computed tomography (CT) images of the larynx and trachea. The sagittal and axial images highlight the subglottic stenosis (arrow), the circumferential wall thickening of the trachea (in yellow), and the partial obstruction of the tracheal lumen.***

Case No. 2

A 58-year-old woman with a known history of GPA came to our Clinic with a one-month history of difficulty swallowing and shortness of breath at rest. She had been diagnosed with GPA seven years earlier, and over time the disease had spread to affect multiple organ systems. Her upper respiratory tract took a significant hit, chronic sinus disease, cartilage destruction, and hearing loss. Her eyes were also involved, with inflammation affecting the cornea and uvea, dry eyes, thinning of the sclera, and a granuloma behind the eye. Beyond that, she had lung disease with nodules and cavities, kidney involvement in the form of glomerulonephritis, pericarditis, nerve damage affecting the vagus nerve, and joint problems.

Over the years, she had been through a long list

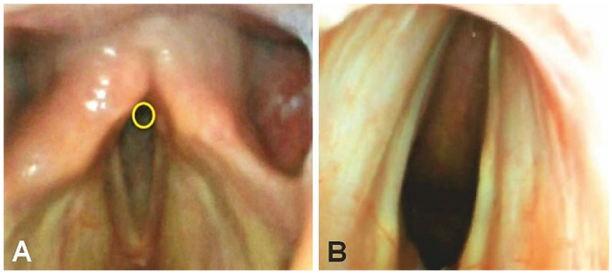

Figure 3 – ***(A and B) Laryngoscopy images showing the preoperative subglottic stenosis and the postoperative result after stenosis resection with carbon dioxide (CO_2) laser.***

Given that his disease was active [Birmingham Vasculitis Activity Score (BVAS) of 9], we decided to continue Rituximab and temporarily bump up his corticosteroid dose. The Rituximab had actually started eight months earlier when his upper respiratory tract disease flared – a sinus CT at that time showed worsening bony changes compared to previous scans, with diffuse osteosclerosis in the sinus walls, breakdown of the medial walls of the maxillary sinuses, and partial destruction of the *lamina papyracea* on both sides. He received the standard induction regimen from the Rituximab in ANCA-associated vasculitis (RAVE) study: 375 mg/m² intravenously once weekly for four weeks.

His subsequent course was encouraging. The headaches he had been experiencing improved, and follow-up examination of the subglottic region showed a normal-looking airway. At his most recent evaluation, three months out, things continue to look good, his upper airway appears healthy with no sign of restenosis, and we were able to successfully remove his tracheostomy.

of immunosuppressive treatments (Cyclophosphamide, Azathioprine, Prednisone, Methotrexate, and Rituximab) with periods of remission punctuated by flares.

A contrast-enhanced CT scan showed circumferential but asymmetrical narrowing of the airway just below the vocal cords, extending down into the trachea for about 3.5 cm. At its tightest point, the airway measured only around 4.5 mm across. We started treatment with two endobronchial sessions a month apart, using electrosurgery and dilation to open things up.

Ten days after the second procedure, we scoped her again. The larynx was moving normally, and the stenosis, now sitting high in the trachea at the level of the first cartilage ring, had widened considerably. The remaining lumen was about 13 mm but had a layer of sticky fibrinous material coating it, most likely part of the healing process after the electrosurgical work. We cleared this away using biopsy forceps, carefully working down to the tracheal wall, which left her with a healthy-looking lumen of 16–17 mm.

At three- and six-month follow-up visits, the patient remained clinically stable, with sustained improvement in respiratory symptoms and no evidence of recurrent airway obstruction or restenosis on endoscopic evaluation. The treatment outcome was therefore maintained during the six-month follow-up period, with no need for additional endobronchial intervention.

Discussions

SGS in GPA is uncommon but can be life-threatening, as our two cases show. Airway involvement can appear at any stage of GPA and varies widely, from mild narrowing to critical obstruction needing emergency treatment. In the first case, the patient had active disease with severe Grade III SGS and significant breathing difficulty. Worsening dyspnea, stridor, and voice changes called for quick intervention, first with a tracheostomy to protect the airway, then CO_2 laser resection to remove the narrowed segment. The second patient had a different story: long-standing GPA affecting multiple organs, with an extended narrowing of the larynx and trachea that responded well to electrosurgical treatment and dilation. Together, these cases show just how differently airway involvement can present and progress. Both cases remind us why it is so important to catch respiratory symptoms early in GPA patients. Shortness of breath, stridor, and voice changes should raise a red flag for subglottic involvement, especially since these symptoms are often mistaken for asthma or other respiratory conditions. Endoscopic examination and imaging are essential for mapping out the stenosis: its severity, length, and anatomy all guide treatment decisions.

HP examination helps confirm the diagnosis and sheds light on how the airways become involved. Classical findings include necrotizing granulomatous inflammation with small- and medium-vessel vasculitis, often together with areas of tissue death, giant cells, and mixed inflammatory cells. All the same, these features can be harder to spot in subglottic lesions, particularly in chronic cases where scarring and remodeling may mask active inflammation. This explains why biopsies from the subglottis sometimes come back inconclusive. In our first patient, a nasal biopsy confirmed the diagnosis by showing the hallmark granulomatous inflammation with necrosis and vasculitis. The gradual airway narrowing in both cases likely reflects a combination of fibrotic healing and ongoing inflammation, eventually determining fixed stenosis. This outlines why gathering clinical findings, blood tests, and tissue samples is so important for getting the diagnosis right.

Managing GPA-related SGS is difficult and calls for teamwork across specialties, ear, nose and throat (ENT), pulmonology, and rheumatology all play a role. Both of our patients received systemic immunosuppression alongside endoscopic procedures to open up the airway. In the first case, continuing Rituximab while temporarily increasing corticosteroids led to a good outcome, with no re-narrowing after removing the tracheostomy. The second case shows that repeated endoscopic interventions can effectively improve airway diameter and symptoms in the right patients. These findings reiterate what previous studies have shown: immunosuppression alone often is not enough to keep under control the SGS, and many patients eventually need airway procedures. Timing matters, though, operating during active inflammation can increase the risk of scarring and re-narrowing.

Taken together, these two cases illustrate how variable the course of GPA-associated SGS can be. Treatment needs to be tailored to each patient based on disease activity, how severe the obstruction is, and what treatments they've had before. Early diagnosis, appropriate endoscopic management, and coordinated care across specialties are key to achieving good outcomes and preventing permanent airway damage.

Conclusions

SGS does not happen often in GPA, but when it does, it can become a real threat to the airway if not picked up early. Getting a timely endoscopic look at the airway, along with appropriate imaging, is crucial for figuring out what you're dealing with and how to approach it. There's no single treatment that works for everyone; management needs to be shaped around the individual patient. This usually means combining medical therapy with minimally invasive procedures like CO_2 laser resection, electrosurgery, or dilation to open up the airway while keeping complications to a minimum. Surgery tends to be saved for cases where the stenosis is fixed or hasn't responded to other approaches.

None of this works well in isolation. Close teamwork between otolaryngologists, anatomopathologists, pulmonologists, radiologists, and rheumatologists is really what makes the difference, both for tackling the active disease and for managing the airway changes that develop over time.

Conflict of interests

The authors declare that they have no conflict of interests. All authors have read and agreed to the published version of the manuscript.

Informed Consent Statement

Written consent was obtained from the patients participating in this study.

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Corresponding authors

Carmen Aurelia Mogoantă, Associate Professor, MD, PhD, Department of Otorhinolaryngology, Faculty of Medicine, University of Medicine and Pharmacy of Craiova, 2 Petru Rareş Street, 200349 Craiova, Dolj County, Romania; Phone +40728–020 623, e-mail: carmen_mogo@yahoo.com

Gabriela Cornelia Muşat, Associate Professor, MD, PhD, Department of Otorhinolaryngology and Head and Neck Surgery, Sf. Maria Hospital, 37–39 Ion Mihalache Avenue, Sector 1, 011172 Bucharest, Romania; Phone +40722–505 636, e-mail: gabimusat@yahoo.com

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