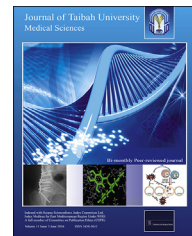




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Original Article

## Wegener's granulomatosis causing subglottic stenosis: Experiences at a tertiary care hospital of the Eastern India



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### المخلص

**أهداف البحث:** تهدف هذه الدراسة لمعرفة المظهر السريري ومعالجة تورمات "فيجنر" مع ضيق تحت المزمار في مستشفى تعليمي جامعي في شرق الهند.

**طرق البحث:** أدرجت الدراسة الحالية سلسلة حالات 6 مرضى، تم علاجهم خلال الفترة من 2007م إلى 2015م، لتورمات فيجنر مع ضيق تحت المزمار. وعرضت التفاصيل الديموغرافية للمرضى مثل العمر، والجنس، والأعراض السريرية، وتنظير الحنجرة، والتصوير الشعاعي، والفحوصات المخبرية، والخيارات الطبية والجراحية.

**النتائج:** كان لدى خمسة من بين الستة مرضى، أعراض بالحنجرة مثل بحة في الصوت أو صعوبة في التنفس في وقت المراجعة. وكان أربعة من المرضى إناثا واثنين من الذكور، تتراوح أعمارهم من 14 إلى 62 عاما. تم تأكيد التشخيص لجميع المرضى عن طريق فحص الأنسجة. وكان لدى أحدهم انتشار منغل تحت المزمار، وأربعة مرضى كان لديهم فحص المستضدات الذاتية لمضادات سيتوبلازم العدلات إيجابيا عند المراجعة. وأعطى جميع المرضى العلاجات المثبطة للمناعة والستيرويدات منذ تشخيصهم. واحتاج خمسة منهم إلى عمل قطع في القصبة الهوائية مع توسعة تحت المزمار بواسطة أدوات الصلب الباردة، يتبعها حقن الستيرويد موضعيا ووضع ميتومايسين - س. وقد أظهر أربعة من المرضى تحسنا سريريا.

**الاستنتاجات:** بالرغم من أن تورمات فيجنر هي حالة سريرية نادرة لكن غالبيتها تتشابه مع أمراض شائعة تؤدي إلى تأخير التشخيص، وتأثر تحت المزمار. تحت المزمار هو جزء حيوي للمجرى الهوائي الرغامى لأن أي انسداد خفيف قد يؤدي إلى كارثة تهدد الحياة. التشخيص الصحيح وفي الوقت المناسب لتورمات فيجنر يساعد في الوقاية من المضاعفات التي تهدد الحياة كضيق تحت المزمار.

**الكلمات المفتاحية:** الحنجرة؛ ضيق تحت المزمار؛ تورمات فيجنر؛ المجرى الهوائي الرغامى؛ انسداد مجرى الهواء

### Abstract

**Objectives:** To study the clinical profile and management of Wegener's granulomatosis (WG) with subglottic stenosis (SGS) at a tertiary teaching hospital in Eastern India.

**Methods:** This prospective study incorporated a case series of six patients treated between 2007 and 2015 for WG with SGS. The demographic details of the patients, such as age, sex, clinical presentation, laryngeal endoscopy, imaging, laboratory tests, and medical and surgical options, are described.

**Results:** Of the six patients, five had laryngeal symptoms, such as hoarseness or breathing difficulty, at the time of presentation. There were four female and two male patients ranging in age from 14 to 62 years. The diagnosis of all six patients was confirmed via histopathological examination. Of the six patients, one had isolated subglottic involvement, and four had a positive antineutrophilic cytoplasmic autoantibody (C-ANCA) test on presentation. All of the patients received immunosuppressant and steroid therapies at the time of diagnosis. Five patients required tracheostomy with subglottic dilatation with cold steel instruments followed by the local injection of steroids and mitomycin-C application. Four patients have shown clinical improvement.

**Conclusion:** Although WG is a rare clinical condition, it is often confused with common ailments, which delays diagnosis, and it may involve the subglottis. The subglottis is a vital part of the laryngotracheal airway, and mild obstruction can be life threatening. The accurate and timely diagnosis of WG helps to prevent life-threatening complications, such as SGS.

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**Keywords:** Airway obstruction; Laryngotracheal airway; Larynx; Subglottic stenosis; Wegener's granulomatosis

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## Introduction

Wegener's granulomatosis (WG), or granulomatosis with polyangiitis (GPA), is an autoimmune multisystemic inflammatory disease characterized by small- to medium-vessel vasculitis with the formation of necrotizing granulomas. Heinz Klinger first described WG in 1931; it was subsequently described by Frederich Wegener in 1936.<sup>1</sup> WG has a predilection for forming in the upper and lower respiratory tract and kidneys. WG without kidney involvement is referred to as limited Wegener's. There is no definitive cause of WG, although it is not infectious or hereditary. The diverse clinical presentations of WG contribute to difficulties in diagnosing this disease. WG most commonly presents among patients in their sixth and seventh decades of life, but it can occur at any age and occurs with equal frequency between genders among adults.<sup>2</sup>

Laryngotracheal involvement occurs in 16% cases of WG and may be missed. Laryngotracheal involvement may be symptom free, but its clinical manifestations can range from subtle hoarseness to life-threatening stridor.<sup>3</sup> Some cases may present with normal chest X-rays. The narrowing of the airway at the level of cricoid cartilage presents a challenging situation for the treating doctors. WG is an uncommon clinical condition, with a prevalence of 3 per 100,000; subglottic stenosis (SGS) occurs in approximately 16% of cases, leading to life-threatening airway obstruction.<sup>4</sup> The characteristic laboratory findings in WG are elevated ESR, leucocytosis, anaemia, elevated rheumatoid factor and positive antiproteinase-3 ANCA. The C-ANCA test is not always positive initially and should be repeated if WG is suspected. SGS in WG usually does not respond to medical treatment.<sup>5</sup> We present a series of cases of WG with SGS with an emphasis on early diagnosis and treatment.

## Materials and Methods

All cases of WG with SGS were studied at a tertiary care teaching hospital in eastern India between 2007 and 2015. All of the six cases of WG were treated over eight years, and all involved the subglottis. All were clinically assessed in an attempt to further characterize this unusual disease entity and to develop a rationale for the diagnosis and management of the compromised laryngotracheal airway. To this end, the existing literature on the subject was critically reviewed.

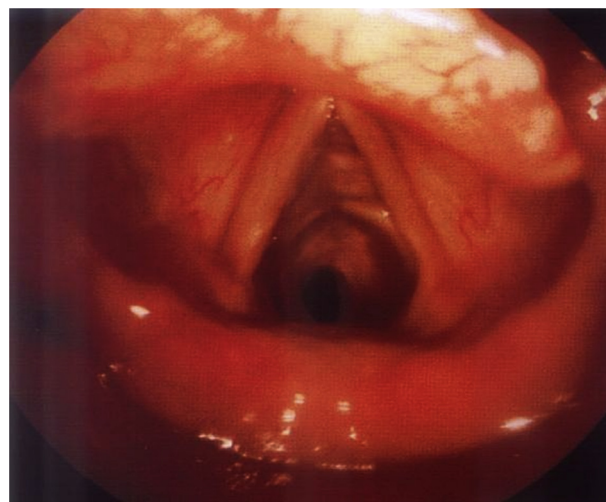
### Case-1

A 21-year-old female was admitted with a history of progressive breathing difficulty for 3 months. She had

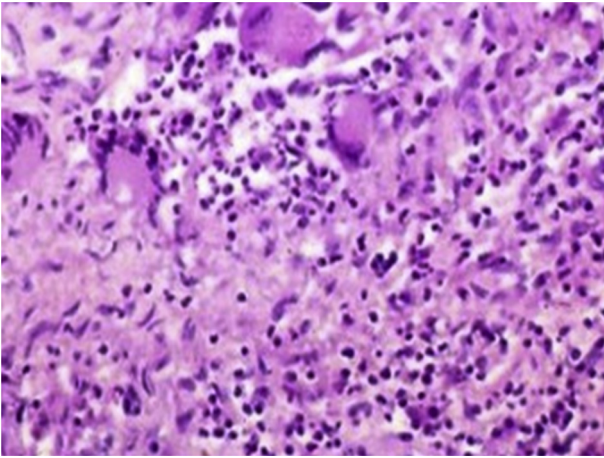
undergone a tracheostomy for the sudden development of respiratory failure on the ward. Nasopharyngolaryngoscopy (NPL scopy) showed Grade II Cotton Meyer subglottic stenosis (Figure 1), and the stenotic area had a concentric, erythematous, friable appearance. Computed tomography (CT scan) of the neck showed stenosis confined to the subglottis, with no involvement other parts of the airway. Laboratory tests showing elevated ESR, positive C-ANCA. Microlaryngeal surgery (MLS) was performed under general anaesthesia, and a biopsy of multiple fragments from the subglottic lesion revealed granulation tissues. Methyl prednisolone (80 mg) was injected at the stenotic area, followed by dilation and the local application of mitomycin-C (2 mg mitomycin-C diluted in 1 ml distilled water). The chest X-ray was normal, but the paranasal sinus (PNS) X-ray showed haziness in the right maxillary sinus. A biopsy sample taken from the right maxillary sinus using an endoscopic approach confirmed the WG diagnosis. Treatment with glucocorticoids and cyclophosphamide was initiated and lead to clinical improvement and the reduction of SGS in the subsequent months.

### Case-2

A 32-year-old man was referred for complaints of hoarseness, coughing and difficulty breathing. Indirect laryngoscopy showed narrowing of the subglottis with circumferential boggy, erythematous folds of redundant tissue. The vocal cords were normal and mobile. The results of a general physical examination of the patient were within normal limits. The blood counts and urine analysis were normal. A Montoux test for tuberculosis was negative. A chest X-ray showed a patch on one lobe; the PNS X-ray was normal. A CT scan of the neck showed concentric narrowing, with soft tissue confined to subglottis. Because of severe subglottic obstruction and breathing difficulty, a tracheostomy was performed and a biopsy was obtained. Histopathological examination showed granulation tissue infiltrated with lymphocytes, giant cells, histiocytes and a small vessel in a non-inflamed area showing granulomatous vasculitis (Figure 2). The subglottic area was injected with



**Figure 1:** Nasopharyngolaryngoscopy showing subglottic stenosis.



**Figure 2:** Histopathology image showing granulations infiltrated with lymphocytes, histiocytes, giant cells and a small vessel showing granulomatous vasculitis.



**Figure 3:** CT scan of the neck showing SGS.

steroids, and mitomycin-C was applied. The diagnosis of WG was confirmed, and the patient was treated with prednisolone 60 mg/day and cyclophosphamide 150 mg/day. Subsequently, the subglottic airway showed improvement.

#### Case-3

A 14-year-old female known to have WG and undergoing immunosuppressive therapy (prednisolone 60 mg/day and cyclophosphamide 150 mg/day) was referred to us for breathing difficulty with cough. She had undergone tracheostomy for stridor. She had a high ESR (110 mm/h), and her C-ANCA was positive. Her chest X-ray showed patchy and nodular areas with a ground glass appearance in both lung fields, which were consistent with WG. Diagnostic nasal endoscopy revealed crusts inside the nasal cavity, and an earlier biopsy of the nasal cavity showed features of WG. NPL scopy showed SGS. The CT scan of the neck confirmed circumferential narrowing confined to the subglottis (Figure 3). The patient was scheduled for MLS and dilation with cold steel instruments. Methyl prednisolone (80 mg) was injected at the stenotic area, followed by dilation and the local application of mitomycin-C (2 mg mitomycin-C diluted in 1 ml distilled water). After 3 months, the patient tolerated the blocking of tracheostomy and subsequent decannulation.

#### Case-4

A 27-year-old woman diagnosed with WG at an outside institution was referred to us for mild dyspnoea at rest and a brassy cough. NPL scopy showed SGS (Cotton-Myer Grade III). A CT scan localized the lesion at the subglottis. In the early stage, she was taking steroids and cyclophosphamide. Her C-ANCA test was positive. A tracheostomy was performed to treat stridor. Via MLS, the subglottic stenotic segment was excised, and a local injection of steroids was administered along with an application of mitomycin-C. After 4 months, the tracheostomy tube was decannulated and the subglottic area had improved.

#### Case-5

A 62-year-old woman presented with nasal obstruction and discharge. On examination, she presented mild dyspnoea at rest and a brassy cough. The chest X-ray was normal. Pulmonary function tests were consistent with extrathoracic upper airway obstruction. NPL scopy showed sluggishly moving bilateral vocal cords and a circumferentially narrowed subglottic space (Cotton-Meyer Grade I). A biopsy of the nasal granulomatous tissue showed typical necrotizing WG. The patient's C-ANCA test was positive. The patient received prednisolone and cyclophosphamide, and no tracheostomy was needed. The patient died one year after treatment from renal failure.

#### Case-6

A 55-year-old male with known WG and chronic renal failure was referred to us for breathing difficulties, a hoarse voice and nasal obstruction. NPL scopy showed Grade III Cotton Meyer SGS. He was treated with prednisolone and azathioprine. CT scan showed SGS. Tracheostomy was performed for stridor. MLS with dilation of the subglottic area, steroid injection and mitomycin-C application were performed. The patient continued with azathioprine and prednisolone. He is now under follow-up care.

#### Results

All six cases of WG were referred to our department within the last eight years. The diagnosis was confirmed via histopathological examination. Four patients had a positive C-ANCA test on presentation. Of the six patients, one had isolated subglottic involvement. There were four female patients and two male patients. The age range was from 14 to 62 years. Four patients had laryngeal symptoms at the time of presentation and subsequently developed airway obstruction. The severity of SGS was determined according to the patient's history, clinical features, and laryngeal endoscopy and imaging results. SGS in WG was diagnosed via NPL

scopy and confirmed with radiological imaging of the laryngotracheal airway. All of the patients had taken immunosuppressants and steroids since the time of diagnosis. Of the six patients, five required a tracheostomy. Via MLS, the subglottic area was injected with methyl prednisolone, and mitomycin-C was applied. Four patients improved with this treatment; one patient is in follow-up, and one patient died of renal failure. Details are provided in Table 1.

## Discussion

WG is a rare autoimmune multisystemic disorder characterized by granulomatous inflammation of the respiratory tract, necrotizing vasculitis affecting small- to medium-sized vessels, and necrotizing glomerulonephritis. WG is also known as granulomatosis with polyangiitis (GPA).<sup>5</sup> Both sexes are equally affected in WG; however, in our case series, females outnumbered males (4:2). There are three varieties of WG: Types 1, 2 and 3. Type 1 is the limited form of WG, in which the patient presents with the symptoms of upper respiratory infection for weeks and does not respond to antibiotics. This type is associated with serosanguinous nasal discharge, pain and crust formation in the nasal cavities. In Type 2, in addition to nasal involvement, other organs are involved. In Type 3 WG, there is widespread involvement of multiple organs, including airway, pulmonary, renal and sometimes dermatological lesions.<sup>6</sup> Laryngotracheal manifestations in WG are rare, with subglottic involvement being the most common. Laryngeal manifestations occur as a late complication of fully developed or previously treated WG. They are seen in 8–23% of patients throughout the disease process; they represent the first manifestation of WG in 1–6% of cases<sup>7</sup> and are common when WG is diagnosed before the age of 20 years.<sup>8</sup> Although all six patients in this case series presented with SGS, in only one was the disease confined to the subglottis. A few systemic diseases can cause SGS, including WG, relapsing polychondritis, sarcoidosis, and amyloidosis. Congenital stenosis, trauma to the larynx, high tracheostomy and the extension of tumours from another site to the subglottic area are other

causes of SGS.<sup>9</sup> The pathogenesis of SGS in WG is unclear. It is thought that during systemic flare-ups, subclinical subglottic involvement can occur, leading to healing and circumferential scarring and stenosis. SGS can progress even in the absence of systemic manifestations of WG. The subglottis is a watershed area of microcirculation; therefore, it is vulnerable to granulomatous inflammation. This watershed area is the junction of two embryologically different mucosal linings. The exposure of the respiratory epithelium to gastric fluids during laryngopharyngeal reflux is believed to play a role in the development of SGS. Initial granulomatous inflammation at the subglottis leads to circumferential scarring and narrowing of the airway.

The clinical presentations of WG are often heterogeneous and often affect the upper respiratory tract, lungs, and kidneys. Localized forms of WG in the head and neck area are not exceptional.<sup>10</sup> The upper respiratory tract is affected in 95% of WG patients.<sup>9</sup> The otolaryngological manifestations of WG are sinusitis, nasal septal perforation, saddle nose deformity, otitis media, hearing loss and SGS (16%). Other clinical manifestations are glomerulonephritis, pulmonary infiltrates, conjunctivitis, scleritis, hyperthyroidism, arthralgia, neuropathy, and pericarditis. In cases of WG with subglottic involvement, dyspnoea on exertion is the most common clinical manifestation; other symptoms include voice changes, cough or stridor. SGS can be graded using the Cotton-Myer classification, which considers circumferential narrowing of the subglottic area: Grade I: Obstruction of 0–50% of the lumen, Grade II: Obstruction of 51–70% of the lumen, Grade III: Obstruction of 71–99% of the lumen, and Grade IV: Obstruction of 100% of the lumen. The typical history and classical clinical findings on examination of mild to moderate anaemia and elevated ESR are suggestive of WG.<sup>6</sup> The diagnosis is confirmed via histopathological examination. A positive C-ANCA test is an important addition to the diagnosis of WG, but these autoantibodies may be absent in certain cases, particularly in limited type of the disease. ANCA are antibodies that target the two important components of neutrophil granulocytes: PR3 serine and myeloperoxidase. Anti-PR3 antibodies are

**Table 1: Detailed clinical profiles of the patients involved in this study.**

| SL No | Gender | Age (years) | Clinical presentation                                  | Disease location           | Surgical treatment | Immunosuppressive therapy   | Outcome   |
|-------|--------|-------------|--|----------------------------|--------------------|---|---|
| 1     | Female | 21          | Breathing difficulty                                   | Subglottis, nose and sinus | Tracheostomy, MLS  | Prednisolone and cyclophosphamide   | Improved  |
| 2     | Male   | 32          | Breathing difficulty and hoarseness of voice           | Subglottis, lungs          | Tracheostomy, MLS  | Prednisolone, cyclophosphamide, tracheostomy                                  | Improved  |
| 3     | Female | 14          | Breathing difficulty, cough, nasal crusts              | Subglottis, lungs, sinus   | Tracheostomy, MLS  | Prednisolone, cyclophosphamide, MLS with dilation with cold steel instruments | Improved  |
| 4     | Female | 27          | Breathing difficulty, brassy cough                     | Subglottis                 | Tracheostomy, MLS  | Prednisolone, cyclophosphamide, tracheostomy                                  | Improved  |
| 5     | Female | 66          | Pansinusitis, breathing difficulty                     | Subglottis, sinus, kidney  | Not applicable     | Prednisolone, cyclophosphamide  | Patient died of renal failure                   |
| 6     | Male   | 55          | Breathing difficulty, nasal obstruction, renal failure | Subglottis, nose, kidney   | Tracheostomy, MLS  | Azathioprine, prednisolone, tracheostomy                                      | Patient with tracheostomy; undergoing follow-up |

virtually pathognomonic of WG, whereas anti-myeloperoxidase antibodies are more consistent with necrotizing primary vasculitis, as seen in microscopic polyangiitis.<sup>11</sup> There are two types of tests to detect ANCA: Immunofluorescence or enzyme linked immunoassay (ELISA). The immunofluorescence test differentiates between anti-PR3 and anti-myeloperoxidase on the basis of staining patterns: the first is associated with C-ANCA, and the second is associated with P-ANCA.<sup>12</sup> Detection with ELISA (the presence of anti-PR3 or anti-myeloperoxidase) offers greater specificity. When both techniques are combined, the sensitivity and specificity for the diagnosis of WG increase to 90% and 98%, respectively.<sup>13</sup> The sensitivity of the C-ANCA test was found to be 67% with immunofluorescence and 60% with ELISA for patients with active local or regional symptomatology.<sup>14</sup> A negative C-ANCA test does not exclude the diagnosis of WG. The sensitivity of the ANCA test depends on the extent of the lesions and activity at the time of the test. In our series, C-ANCA was positive in four patients at the time of presentation, whereas the other two cases became positive later in the course of the disease. C-ANCA is not constant throughout the course of the disease and can be modified with medical treatment.

Biopsies give best information for diagnosis. Subglottic biopsies are not always sensitive as large amounts of tissue from the subglottic area are difficult to obtain. The diagnosis of WG is confirmed with a biopsy that shows necrotizing granulomatous vasculitis.<sup>9</sup> Biopsies from pulmonary lesions provide the highest diagnostic yield, but biopsies from other parts, such as the airway and kidney, can also be taken for diagnosis.<sup>9</sup> The classical laboratory findings are markedly elevated ESR, anaemia, leucocytosis, elevated rheumatoid factor and positive antiproteinase-3 ANCA.<sup>9</sup>

The head and neck manifestations of patients with WG usually respond well to immunosuppressive therapy (cyclophosphamide or methotrexate) along with glucocorticoids for several months during the active phases of the disease. Later, the patient will require maintenance therapy with azathioprine and methotrexate during the quiescent phase, along with boluses of glucocorticoids to control the reactivation of the diseases.<sup>15</sup> Monoclonal antibodies against CD20 (rituximab) can also be useful in WG.<sup>16</sup> During the early stage of laryngotracheal involvement of WG, corticosteroids and cyclophosphamide are used, but only 20% of cases respond. All of our cases received immunosuppressive therapy, but there was no long-lasting benefit to the course of the SGS in any of the cases. This suggests that SGS can occur independent of the systemic manifestations and their treatment. Intralesional steroids have been used to treat SGS and have been shown to reduce the need for systemic therapy. Intralesional corticosteroids are a safe and effective treatment when used in conjunction with dilation of the SGS. Intralesional steroid injection and the application of mitomycin-C was performed in five out of the six cases. SGS can be treated with airway dilation via silicon or metal stent placement, but the prognosis remains guarded.<sup>17</sup> In cases of resistant fixed obstructions of the airway, open reconstructive surgery, such as laryngoplasty and cricotracheal resection with primary stenosis, is useful.<sup>17</sup> CO<sub>2</sub> laser treatment of SGS has been attempted but has not proven useful in extensive cases.

## Conclusion

Although WG is a rare clinical condition, an accurate and timely diagnosis can be made on the basis of certain clinical features and laboratory tests and confirmed with biopsy. Because airway involvement may occur in WG, it is essential that the treating physicians rule out this condition; as favourable diagnosis depends on early detection and treatment. Microlaryngoscopy and dilation with mitomycin-C and steroid treatments provide good results in cases of SGS caused by WG. A combination of clinical features, immunological tests and characteristic histopathological presentation are needed to establish the diagnosis. A CT scan is performed to define the extent of the stenosis. Patients who do not respond to immunosuppressive therapy may require surgical intervention. In our series, four patients improved, one patient is in follow-up, and one patient died from renal failure. The treating physicians should always be aware of the possibility of SGS in WG, which may lead to life-threatening stridor.

## Recommendation

This study was approved by a competent authority of IMS and Sum Hospital, Siksha 'O' University, Bhubaneswar, India.

## Conflict of interest

The authors have no conflict of interest to declare.

## Authors' contributions

SKS conceived and designed the study, conducted research, provided research materials, and collected and organized data. MCS analyzed and interpreted data for drafting the manuscript. JRP wrote initial and final draft of article, and provided logistic support. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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