

# Childhood Wegener's Granulomatosis with Subglottic Stenosis

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## ABSTRACT

Our aim is to highlight the possibility of subglottic stenosis caused by Wegener's granulomatosis. The incidence of subglottic stenosis in Wegener's granulomatosis is seen five times more often in children.

We report a case of a 12-year-old female diagnosed as Wegener's granulomatosis who was on medical treatment since six months. She presented to us with stridor. As rigid laryngoscopy revealed a pinhole opening in the subglottic region, an emergency tracheostomy was performed. This was followed by a planned microlaryngoscopy using cold steel instruments. A triradiate incision with dilatation, local steroid injection and topical application of mitomycin-c was performed. In Wegener's granulomatosis, subglottic stenosis is caused due to vasculitis therefore, the use of laser and stent is contraindicated, as this may lead to a further worsening of the stenosis.

Among various causes of subglottic stenosis, the most common cause is use of cuffed tracheostomy tube and endotracheal tube. It is essential to diagnose the etiology of subglottic stenosis to decide on the appropriate modality of treatment.

**Keywords:** Wegener's granulomatosis, Subglottic stenosis, Dilatation, Local steroid and mitomycin-c.

## INTRODUCTION

Subglottic stenosis can be defined as any narrowing of the subglottic area. The most common cause is prolonged use of cuffed tracheostomy and endotracheal tubes.<sup>1</sup> The management of subglottic stenosis has been called 'a perplexing problem which continues to tax the ingenuity of the better laryngologist'.<sup>2</sup> The systemic diseases which can cause subglottic stenosis are Wegener's granulomatosis, amyloidosis, sarcoidosis, relapsing polychondritis.<sup>1</sup> Other causes of subglottic stenosis are trauma to larynx, high tracheostomy incision, congenital stenosis and extension of neoplastic disease from other sites to the subglottic area.<sup>1</sup>

Wegener's granulomatosis is an uncommon disease with a prevalence of 3 per 100,000 and subglottic stenosis occurs in approximately 16% of patients and may result in severe airway obstruction.<sup>3</sup> Subglottic stenosis in Wegener's granulomatosis does not typically respond to medical line of treatment.<sup>3</sup> We present a case of childhood Wegener's granulomatosis in whom subglottic stenosis was managed by dilatation with cold steel instrument.

## CASE REPORT

A 12-year-old female, diagnosed 6 months earlier with Wegener's granulomatosis was referred to us for respiratory distress. She had very high ESR (i.e. 116) and her cANCA was positive. The plain CT scan of chest revealed patchy and nodular area of consolidation and ground glass appearance in bilateral lung fields suggestive of Wegener's granulomatosis (Fig. 1).



Fig. 1: CT scan chest

Anterior rhinoscopy revealed an atrophic rhinitis like picture and earlier nasal biopsy had revealed features of Wegener's granulomatosis.

A 70° Hopkins rigid laryngoscopy demonstrated a pinhole opening in the subglottis (Fig. 2). An emergency tracheotomy was performed. The plain CT scan of larynx confirmed the focal luminal narrowing at the level of glottis and subglottis region with a total length of stenosis 5 mm (Fig. 3).

Our patient was posted for microlaryngoscopy and dilatation using cold steel instruments. Methylprednisolone in the dose of 80 mg was injected at the stenotic site followed by dilatation and local application of mitomycin-C (2 mg of mitomycin-C diluted in 1 ml of normal saline) (Fig. 4). At 6 weeks

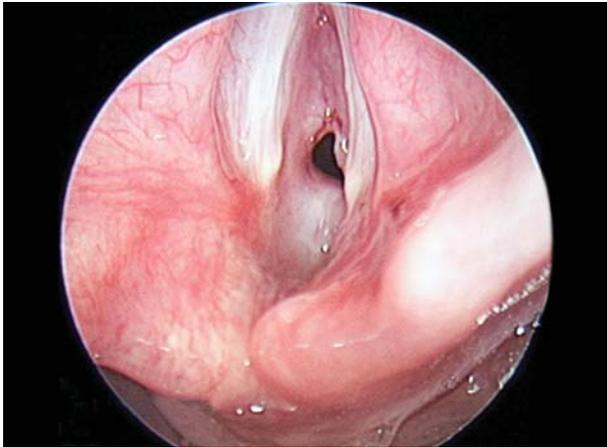


Fig. 2: Rigid laryngoscopy (70 degree Hopkins endoscope)



Fig. 3: CT scan larynx

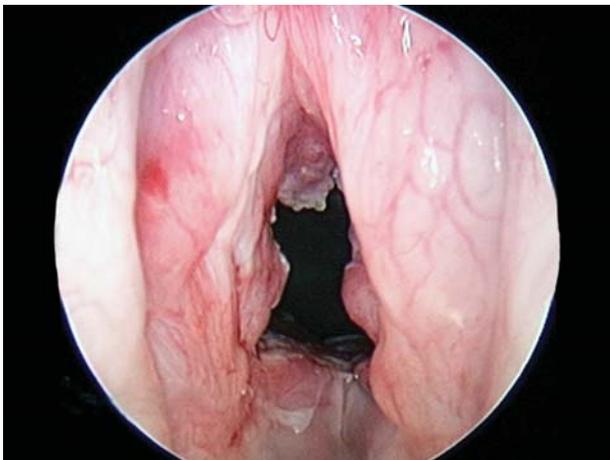


Fig. 4: After MLS and dilatation

postprocedure, patient was tolerating tracheostomy full block well.

## DISCUSSION

Wegener described the entity known as Wegener's granulomatosis in 1936 and again in 1939.<sup>1</sup> Wegener's granulomatosis is

a rare disease defined by the triad of (1) granulomatous necrotizing processes in upper or lower respiratory tract, (2) generalized vasculitis primarily of the small arteries and vein and (3) focal necrotizing glomerulitis.

Involvement of upper respiratory tract occurs in 95% of patients.<sup>3</sup> Clinical features seen by ENT specialists include sinusitis, nasal septal perforation, saddle nose deformity, otitis media, hearing loss, subglottic stenosis (16%). Other features are glomerulonephritis, pulmonary infiltrate, conjunctivitis, scleritis, neuropathy, pericarditis, hyperthyroidism, arthralgia, papule, vesicle. Diagnosis of Wegener's granulomatosis is made by demonstration of necrotizing granulomatous vasculitis on tissue biopsy.<sup>3</sup> Pulmonary tissue offers the highest diagnostic yield but biopsy can be taken from upper airway and kidney for diagnosis.<sup>3</sup>

The characteristic laboratory findings include markedly elevated ESR, mild anemia, leukocytosis, mildly elevated rheumatoid factor, positive antiproteinase-3 ANCA.<sup>3</sup>

Wegener's granulomatosis is a treatable disorder. Steroid, with or without cyclophosphamide are treatments of choice.<sup>1</sup> The treatment regimen includes daily oral therapy with cyclophosphamide (2 mg/kg body weight) and prednisone (1 mg/kg body weight).<sup>5</sup> Success rate as high as 70 to 80% has been reported by regular dilatation alone.<sup>2</sup>

## CONCLUSION

Because of diverse initial presentation of the disease, the physician should consider that the stenosis may be a manifestation of a systemic disorder and carry out an otolaryngologic and physical examination with the appropriate roentgenogram and blood, urine tests.<sup>1</sup> Since airway involvement is common, it is essential that the otolaryngologist remains alert to the possibility of this condition, because favorable prognosis depends on early recognition and treatment.<sup>1</sup>

Subglottic stenosis in Wegener's granulomatosis is due to necrotizing vasculitis, so any mucosal trauma resulting from major surgery, laser and stent leads to worst stenosis, so this should be avoided.<sup>8,9</sup> Use of controlled low pressure, large volume, nontraumatic endotracheal and tracheostomy cuffs and deflating cuff every two hourly for 5 to 10 minutes has decreased the incidence of intubation injuries.<sup>6</sup> Microlaryngoscopy and dilatation with local steroid, mitomycin-c give a good result in subglottic stenosis due to Wegener's granulomatosis.<sup>7</sup>

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