Endoscopic Management of Subglottic Stenosis in a Child with Granulomatosis with Polyangiitis

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SUMMARY

Background: Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a rare autoimmune disorder characterized by necrotizing vasculitis of small- to medium-sized vessels, most commonly affecting the kidneys and respiratory tract. Airway involvement, particularly subglottic stenosis, is more frequently observed in pediatric patients and may present as stridor or progressive dyspnea.

Methods: We report the case of a 12-year-old girl presenting with acute upper airway obstruction. Emergency endoscopic surgery was performed without tracheotomy, utilizing CO_2 laser excision in combination with local anti-inflammatory therapy.

Results: Histopathology revealed granulation tissue without definitive granulomas. Positive atypical perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) supported the diagnosis of GPA. The patient responded favorably to systemic corticosteroid therapy, with a complete resolution of symptoms.

Conclusion: Although rare, GPA-related airway manifestations in children require prompt multidisciplinary evaluation. Early endoscopic intervention can be effective and may obviate the need for tracheostomy.

KEYWORDS

Granulomatosis with polyangiitis; Subglottic stenosis; Pediatric airway obstruction; p-ANCA; Endoscopic laser surgery

Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis, is a systemic necrotizing vasculitis affecting small- and medium-sized vessels. It frequently affects the kidneys and respiratory tract, and is associated with elevated levels of anti-neutrophil cytoplasmic antibodies (ANCA)[1].

Its etiology remains unclear but is believed to involve an interplay between genetic predisposition, environmental triggers, infections and immune dysregulation[2]. GPA is uncommon in children, with an estimated incidence of 3-14 cases per million per year[3]. Nephropathy (65%), lower respiratory tract disease (61%) and upper airway involvement (82%) are the most prevalent clinical signs of childhood GPA[4]. Recurring epistaxis, crusts, granulomata, sinus inflammation, or persistent or bloody nasal discharge are all valid criteria to assess upper airway involvement. A typical presenting characteristic of GPA in children is sinus disease, which should be differentiated from rhinosinusitis, or infectious or allergic rhinitis. Chronic illness can harm nasal cartilage, resulting in saddle-nose deformity and septal perforation. Otitis, mastoiditis, oral ulcers or granulomata, mucocele, hearing loss and subglottic stenosis are further signs of involvement of the ears, nose and throat. Pediatric patients have a higher risk of airway involvement, with subglottic stenosis reported in up to 48% of affected children[5].

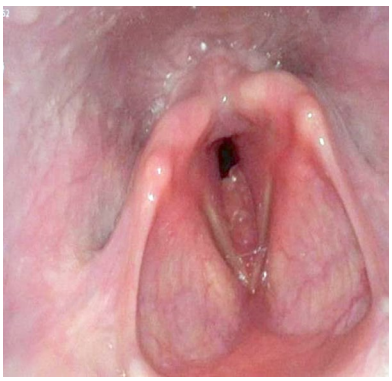
According to the EULAR/PReS/PRINTO criteria, three of the following six features should be present: 1) histopathology (granulomatous inflammation within the arterial wall or in the perivascular or extravascular area); 2) upper airway involvement (chronic purulent or bloody nasal discharge, or recurrent epistaxis/crusts/granulomata, nasal septum perforation or saddle-nose deformity, chronic or recurrent sinus inflammation); 3) laryngo-tracheo-bronchial stenosis (subglottic, tracheal, or bronchial stenosis); 4) pulmonary involvement (chest X-ray or CT scan showing the presence of nodules, cavities or fixed infiltrates); 5) ANCA positivity by immunofluorescence or ELISA (p-ANCA/MPO-ANCA or c-ANCA/PR3-ANCA); and 6) renal involvement (proteinuria > 0.3 g/24 h or > 30 mmol/mg of urine albumin-creatinine ratio on a spot morning sample, hematuria or red blood cell casts in the urinary sediment or ≥ 2+ on dipstick, or necrotizing pauci-immune glomerulonephritis)[6].

This paper aimed to emphasize that GPA is not a common cause of stridor, wheezing, or dyspnea in children, but that it should be a part of the differential diagnosis at some point, and to demonstrate successful treatment by excision of the formation and immediate extubation, without tracheotomy.

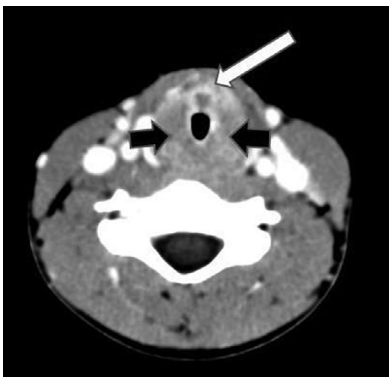
Case report

We describe the case of a 12-year-old girl who presented with 2 weeks of worsening respiratory distress, stridor and hoarseness. She had a 10-month history of a dry, nocturnally worsening cough. Due to her poor clinical condition, she was hospitalized. Oropharyngoscopy and an endoscopic examination revealed normal-appearing vocal cords. However, subglottically, a tumorous formation of soft consistency that filled the entire membranous part of the glottis was visible (Figure 1A). Although it seemed that the formation was also descending subglottically, we could enter the tracheal lumen with the endoscope. An urgent neck CT scan showed diffuse thickening of the supraglottis extending down to the subglottis (Figure 1B). In the area of the anterior commissure, a nodular lesion measuring 7 × 6 × 11 mm could be seen, spreading subglottically.

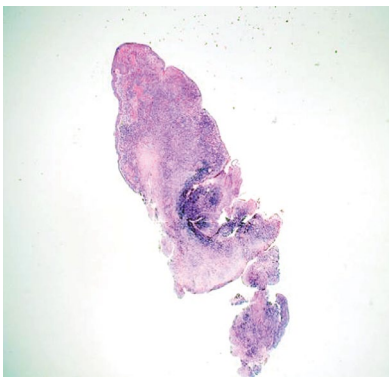
When the patient came to our Department, she could barely climb the stairs and was breathing very loudly. After the examination, the surgeon talked to her parents and they both signed a consent for tracheotomy because, at that time, a tracheotomy was very likely. The patient underwent emergency airway management surgery under general anesthesia since there was a danger of total airway obstruction. The plan was to perform a tracheotomy or to leave her intubated, but the bravest and, as it turned out, the best option was excision of the formation and biopsy, simultaneously with the application of triamcinolone. It was a delicate intubation, and we were



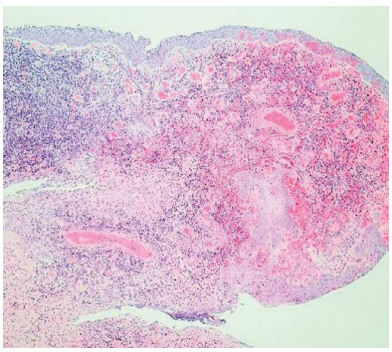
A) Preoperative findings: An endoscopic view of the larynx.



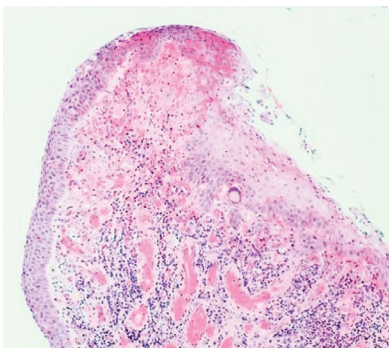
B) Preoperative findings: Axial CT image shows a focal lesion with contrast enhancement in the anterior commissure (B, white arrow) and a thickening of the vocal cords (B, black arrows).



C) A low power image of a small laryngeal biopsy specimen from a 12-year-old girl with a tumefaction of the membranous glottis. The surface of the polypoid sample is covered by respiratory and squamous epithelium. A dense inflammatory infiltrate can be seen, but no granulomas were found (H&E; x20).



D) Upon closer inspection, focal ulceration with reactive changes of the remaining surface epithelium could be seen. Small areas of necrosis, as well as hemorrhage and dense mixed inflammatory infiltrate (H&E; x100) are visible.



E) Only one multinucleated giant cell was found on serially sectioned slides (H&E; x200).

FIGURE 1 Diagnostic findings in our patient

even more concerned about extubation (any procedure, especially a laser in such a small airway, can cause edema that ends in respiratory failure).

Due to the potential for difficult intubation, THRIVE (high-flow nasal oxygen) was administered before anesthesia induction, with preparations made for emergency tracheotomy. Intubation was successfully performed. Direct laryngoscopy revealed a tumorous mass in the subglottic region, originating at the anterior commissure, accompanied by diffuse edema extending downward from the vocal cord edges. A precise excision of the formation was performed using direct microlaryngoscopy with a CO_2 laser (Lumenis Acu Pulse Duo $C0_2$ Laser System) with a micromanipulator.

Triamcinolone acetonide was applied to the lateral walls of the subglottic region, and mitomycin was administered to the anterior commissure to prevent the formation of synechiae. The patient was extubated immediately after surgery without complications, demonstrating prompt respiratory and phonatory recovery.

The histopathological examination of the biopsy material revealed a polypoid fragment of tissue with a dense mixed inflammatory infiltrate consisting of lymphocytes, plasma cells and neutrophils. The surface of the specimen was focally ulcerated and covered by respiratory and squamous epithelium, which showed reactive changes. In the underlying granulation tissue, congested small blood vessels, hemorrhage and small foci of necrosis with fibrin deposition could be seen. The specimen was serially sectioned on 15 slides. The number of sections is mentioned to emphasize that the material was thoroughly examined; fifteen sections is significantly more than is usually necessary for histopathological examination. Deeper serial sections were done in order to try to find the necessary features to confirm the diagnosis, but no granulomas were found, although a single multinucleated giant cell was discovered (Figure 1C-E).

A pediatric rheumatologist was consulted because positive atypical perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) (1:80) were detected. Inflammatory markers were elevated at admission (ESR 35 mm/h, CRP 20 mg/L, leukocytes 14.5×10^9/L). Since there was a danger of total airway obstruction, an emergency surgical procedure was performed, and glucocorticoid therapy was initiated immediately. Soon after, laboratory values decreased. Other laboratory findings were within normal ranges. Due to ANCA positivity, an ultrasound of the kidney was performed, which was normal, and a 24-hour urine collection did not show significant proteinuria. Three days after surgery, the patient was discharged in good general condition, with no voice or breathing difficulties. She received 20 mg of oral prednisone for 4 weeks.

A control MRI scan at 6 months revealed a regression of edematous changes in both vocal cords and the epiglottic area, while in the subglottic region, edematous and diffusely thickened mucosa was still present. Atypical p-ANCA after 6 months was still positive (1:40), which can fit the diagnosis of GPA. On her last clinic visit, 12 months after surgery, there were no respiratory or speech difficulties.

Discussion

In our search of the available literature, 31 pediatric cases of laryngotracheal involvement of GPA were found (Table 1, Figure 2). Out of the 14 patients in the biggest case series, 10 had subglottic or tracheal involvement at presentation, and 4 more developed similar lesions later in the disease. At diagnosis, the mean age was 14.6 years. The symptoms included dyspnea, stridor, coughing and respiratory discomfort. In 86% of the cases, cyclophosphamide was part of the

TABLE 1. Summary of reports on pediatric granulomatosis with polyangiitis with laryngotracheal involvement.

Type of report (author)	Medical treatment	Tracheostomy	Outcome	Number of patients
Retrospective series (Fowler et al.)[7]	Cyclophosphamide; 4 stabilized with medication, 10 required endoscopic surgical interventions, including balloon dilation, application of mitomycin and corticosteroid injections	No	Alive and well	14
Retrospective series (Eustaquio et al.)[8]	Corticosteroids, antineoplastic agents, rituximab, cyclosporine. 3 no surgery, 4 required surgery. Surgical procedures included balloon dilation, laser excision and/or dilation, steroid injections and cricothyroid resection in one severe case.	No	Alive and well	7
Case report (Gajic-Veljic et al.)[10]	IV dexamethasone, IV cyclophosphamide; oral prednisone, methotrexate, intranasal mupirocin ointment; prednisone, mycophenolate mofetil, vitamin D3	No	Alive and well	1
Case report (White)[11]	IV dexamethasone, oral prednisone, azathioprine, trimethoprim/sulfamethoxazole	No	Alive and well	1
Case report (Passey)[9]	IV antibiotics, oral steroids; cyclophosphamide, steroids, intensive nutritional support	Yes, decannulated	Alive and well	1
Case report (Adlakha)[12]	IV methylprednisolone, oral cyclophosphamide; oral prednisone, cyclophosphamide, trimethoprim-sulfamethoxazole; IVIG; laryngotracheoplasty with stent placement	Yes	Alive and well	1
Case report (Bohlman)[13]	IV ampicillin and hydrocortisone; ampicillin, cough and cold medications	Yes	Renal failure	1
Case report (Park et al.)[14]	Prednisone, methotrexate; dilatation, laser therapy; cyclophosphamide; manual dilation of trachea	No	Tracheal stenosis	1
Case report (Matt)[15]	None (postmortem diagnosis)	No	Death	1
Case report (Neumann et al.)[16]	IV antibiotics; prednisone, azathioprine, cyclophosphamide; multiple plasmapheresis; immunosuppressive chemotherapy	Yes	Death	1
Case report (Thomas et al.)[17]	Prednisolone, dexamethasone	Yes, decannulated	Alive and well	1
Case report (Cohen et al.)[18]	Cored-out airway lesions using bronchoscope, IV cyclophosphamide, oral cyclophosphamide and prednisone, dilatation of stenotic areas	Yes	Alive and well	1

IV = intravenous; IVIG = intravenous immunoglobulin

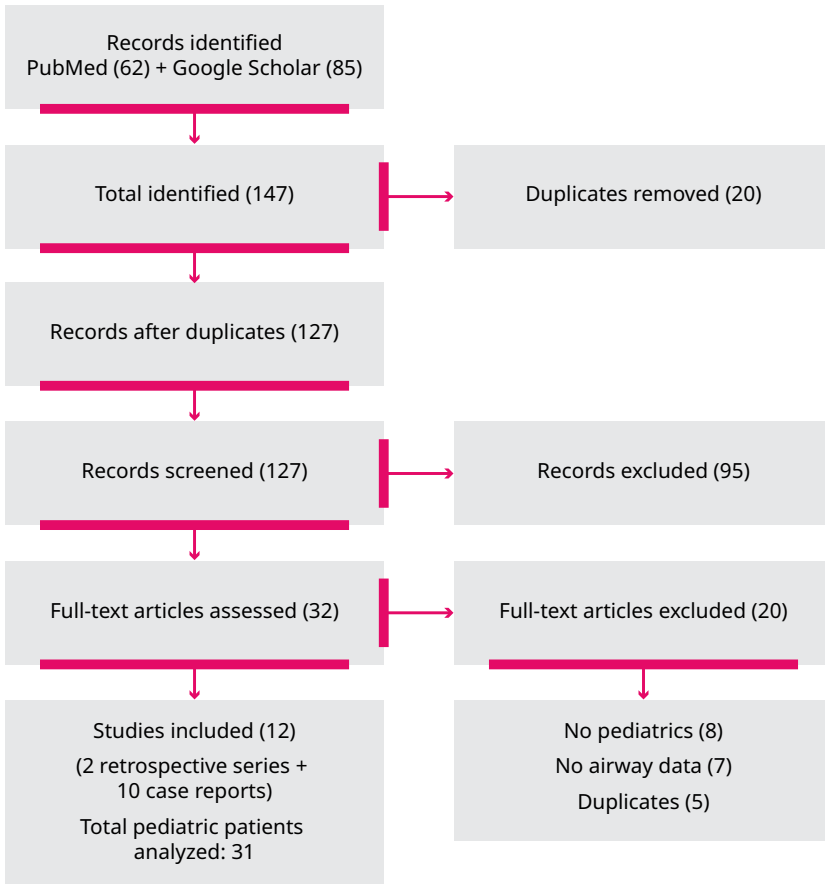


FIGURE 2. PRISMA flowchart of literature review

medical treatment. Four patients had their airway illness adequately stabilized by this treatment; however, 10 patients needed endoscopic surgical procedures, such as balloon dilation, mitomycin application and corticosteroid injections[1,7].

Seven patients with GPA-related airway lesions, such as multilayer stenosis, subglottic stenosis and vocal fold granuloma, were shown in the second case study. The average diagnostic age was 12.7 years. Otolaryngology procedures were not necessary for the isolated lesions found in three individuals, one of whom had a vocal fold granuloma, while the remaining two had subglottic stenosis. Three patients also received rituximab, while one patient received cyclosporine. The remaining four patients had multilayer involvement and were treated with corticosteroids and at least one antineoplastic drug. The surgical techniques used included balloon dilation, laser excision and/or dilation, steroid injections and cricothyroid resection in one severe case[1,8].

Among other patients, at the time of presentation, five patients had tracheal or laryngeal lesions; the remaining five patients acquired these lesions as the illness progressed. Only one patient exhibited laryngotracheobronchial symptoms, and no other organ system involvement was noted[1,8].

Seven individuals underwent surgical airway intervention. The most frequent surgical method

was placing a tracheostomy, and two case reports showed that decannulation was successful[1,9].

Our patient presented with typical endoscopic and radiological findings. The histopathological examination showed granulation tissue and p-ANCA was positive. She also had laryngotracheal stenosis, which led to the diagnosis of GPA. There was also a good response to glucocorticoid therapy. Biopsy is performed in patients with suspected GPA to confirm the disease and to rule out other diseases. Typical histopathological features of GPA are granulomatous inflammation, vasculitis, multinucleated giant cells and necrosis, but it is rare for all of them to be present in a single biopsy specimen, particularly in the head and neck region. All these features are typically evident on lung biopsy specimens and are often absent in most biopsy specimens of oral lesions. More common features in this site are acute or chronic inflammation with microabscesses, multinucleated giant cells, and pseudoepitheliomatous hyperplasia, which are considered nonspecific[19].

Granulomatous inflammation and vasculitis may be absent due to early stages of the disease or small amounts of tissue available, which occasionally necessitates a repeat biopsy to confirm the diagnosis. Sometimes the biopsy specimen contains only non-specific changes which are usually present at the periphery of the more typical areas, which can also be misleading. Histopathological findings in our patient's biopsy specimen were non-specific inflammation with a single multinucleated giant cell. No signs of vasculitis or granuloma formation were found. As mentioned earlier, "classic" histopathological features of GPA in this site (larynx) are not commonly found[20], and non-specific inflammatory changes are more common, so the discovered histopathological findings did not exclude the diagnosis of GPA. The presence of a multinucleated giant cell supported the diagnosis of GPA.

The 2022 American College of Rheumatology Classification Criteria for GPA can be used to classify GPA as a separate disease from other forms of systemic vasculitis[21]. The definition of GPA in the 2012 revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides is as follows "Necrotizing granulomatous inflammation usually involving the upper and lower respiratory tract, and necrotizing vasculitis affecting predominantly small to medium vessels (e.g., capillaries, venules, arterioles, arteries and veins). Necrotizing glomerulonephritis is common.", but it also emphasizes that histopathological features used in definitions do not mean that a diagnosis of the disease can be made only if the pathologic process is directly seen histologically in a biopsy specimen[22]. ANCA associated vasculitides include GPA, microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA). The purpose of the classification criteria — which are not the same as diagnostic criteria — is to ensure that a homogeneous population is selected for clinical trials and research studies of GPA[21].

The final criteria are as follows, along with their weights: bloody nasal discharge, nasal crusting or sino-nasal congestion (+3); cartilaginous involvement (+2); conductive or sensorineural hearing loss (+1); cytoplasmic anti-neutrophil cytoplasmic antibody (ANCA) or anti-proteinase 3 ANCA positivity (+5); pulmonary nodules, mass or cavitation on chest imaging (+2); granuloma or giant cells on biopsy (+2); inflammation or consolidation of the nasal/paranasal sinuses on imaging (+1); pauci-immune glomerulonephritis (+1); perinuclear ANCA or antimyeloperoxidase ANCA positivity (−1); and eosinophil count $\geq 1 \times 10^9$/L (−4). A patient with the diagnosis of small- or medium-vessel vasculitis could be categorized as having GPA if the cumulative score was ≥ 5 points after eliminating vasculitis mimics[21].

This patient's presentation was remarkable, given her predominant complaint of stridor. The peculiarity of this case report is that the procedure was successfully performed without a

tracheotomy, followed by instant extubation, while in most similar cases, the patients were intubated for a long time[1]. Also, a complete excision of the granulation tissue was performed immediately, not just a biopsy.

A limitation of this case report is that we did not find granulomas in the tissue sample, although there was a significant amount of histiocytes, but they were not organized into clear granulomas. The inflammatory infiltrate present in this biopsy was mixed; it consisted of lymphocytes, plasma cells, histiocytes, neutrophils and eosinophils (predominantly lymphocytes), and on deeper serial sections one multinucleated giant cell was found, which is in line with GPA. The indication of granuloma formation would be a multinuclear giant cell of the foreign body type. It can fit into the initial phase of Wegener's granulomatosis, before clear granulomas have yet formed. With positive ANCA, it can fit in the diagnosis of GPA. Since the patient had stridor and laryngeal involvement, a giant cell on histopathological analysis and positive ANCA, she fulfilled the 2022 American College of Rheumatology/European Alliance of Associations for Rheumatology classification criteria for the diagnosis.

Conclusion

GPA-related tracheolaryngeal involvement is a rare presentation of vasculitis in children, with only a few reports in the literature. Patients with subglottis and/or trachea involvement require airway evaluation and some even require surgery to preserve their airway. Even though GPA is rare, it must be considered in such cases, since children who are affected need therapy and lifetime observation. ■

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SAŽETAK

Endoskopsko liječenje subglotične stenoze u djeteta oboljelog od granulomatoze s poliangiitisom – prikaz slučaja

Mirta Peček, Andro Košec, Maja Končić, Tomislav Gregurić, Bernardica Jurić, Mandica Vidović i Siniša Stevanović

Uvod: Granulomatoza s poliangiitisom (GPA), ranije poznata kao Wegenerova granulomatoza, rijetka je autoimuna bolest karakterizirana nekrotizirajućim vaskulitisom malih i srednje velikih krvnih žila, koji najčešće zahvaća bubrege i dišne putove. Zahvaćenost dišnih putova, posebno subglotična stenoza, češće se opaža kod pedijatrijskih bolesnika i može se manifestirati kao stridor ili progresivna dispneja.

Metode: Prikazujemo slučaj 12-godišnje djevojčice s akutnom opstrukcijom gornjih dišnih putova. Hitna endoskopska operacija izvedena je bez traheotomije, korištenjem CO_2 laserske ekscizije u kombinaciji s lokalnom protuupalnom terapijom.

Rezultati: Patohistološki nalaz sadržavao je granulacijsko tkivo bez konačnih granuloma. Pozitivna atipična perinuklearna anti-neutrofilna citoplazmatska antitijela (p-ANCA) bila su sukladna dijagnozi GPA. Bolesnica je dobro reagirala na sistemsku terapiju kortikosteroidima s potpunim povlačenjem simptoma.

Zaključak: Iako rijetke, manifestacije zahvaćenosti dišnih putova povezane s GPA kod djece zahtijevaju brzu multidisciplinarnu procjenu. Rana endoskopska intervencija može biti učinkovita i izbjeći potrebu za traheotomijom.

KLJUČNE RIJEČI

Granulomatoza s poliangiitisom; Subglotična stenoza; Opstrukcija dišnih putova u djece; p-ANCA; Endoskopska laserska kirurgija