Rev. Otorrinolaringol. Cir. Cabeza Cuello 2026; 86: 67-72
doi: 10.4067/S0718-4816202600011624 CLINICAL CASE

Laryngeal manifestations of microscopic polyangiitis in a pediatric patient: a case report

Manifestaciones laríngeas de poliangeitis microscópica en pediatría: reporte de caso clínico inusual

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Resumen

Microscopic polyangiitis is a rare small-vessel vasculitis associated with antineutrophil cytoplasmic antibodies. Otorhinolaryngological manifestations are uncommon, particularly in pediatric patients, which may delay diagnosis and appropriate treatment. We report the case of a 16 year- old female with a history of Graves disease on methimazole who presented to the emergency department with nonspecific otorhinolaryngological symptoms, followed by rapid clinical deterioration that required advanced resuscitation and admission to the pediatric intensive care unit. Clinical evaluation revealed epiglottic edema, yellowish lesions on the oropharyngeal mucosa, and right vocal fold paralysis. Laboratory findings demonstrated positive anti-myeloperoxidase antibodies, confirming the diagnosis of microscopic polyangiitis. The patient received two cycles of rituximab with marked clinical improvement and preserved vocal fold mobility. This case underscores the diagnostic challenge of microscopic polyangiitis with initial laryngeal involvement in a pediatric patient and highlights the importance of close observation and an interdisciplinary approach.
Keywords: Microscopic Polyangiitis, ANCA associated vasculitis, rituximab, child, larynx.

Abstract

La poliangeítis microscópica es una vasculitis rara de vasos pequeños asociada a anticuerpos anticitoplasma de neutrófilos. Las manifestaciones otorrinolaringológicas son poco comunes, especialmente en pacientes pediátricos, lo que puede retrasar el diagnóstico y el tratamiento adecuado. Presentamos el caso de una paciente femenina de 16 años con antecedente de enfermedad de Graves en manejo con metimazol, quien acudió al servicio de urgencias por síntomas otorrinolaringológicos inespecíficos, seguidos de un rápido deterioro clínico que requirió maniobras avanzadas de reanimación e ingreso a la unidad de cuidados intensivos pediátricos. La evaluación clínica reveló edema epiglótico, lesiones amarillentas en mucosa orofaríngea y parálisis de cuerda vocal derecha. Los estudios de laboratorio mostraron anticuerpos antimieloperoxidasa positivos, confirmando el diagnóstico de poliangeitis microscópica. La paciente recibió dos ciclos de rituximab con marcada mejoría clínica y preservación de la movilidad de los pliegues vocales. este caso resalta el desafío diagnóstico de la poliangeitis microscópica con compromiso laríngeo inicial en una paciente pediátrica y destaca la importancia de la observación estrecha y el abordaje interdisciplinario.
Palabras clave: *Poliangitis microscópica, vasculitis asociada a ANCA, rituximab, niño, laringe.*

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The authors declare no conflicts of interest.

Received 16 January 2026.
Accepted 25 February 2026.

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ISSN 0718-4816 versión en línea - ISSN 0716-4084 versión impresa

Introduction

Vasculitis constitutes a heterogeneous group of diseases characterized by immune-mediated inflammation of the vascular wall. The antibody-induced response leads to structural damage, ischemia, stasis of the tissues supplied by affected vessels, and extravasation of blood[1]. When combined with the location, the extent, whether local or systemic, and the size of the affected vessel, as described in the chapel hill classification, these factors provide a broad spectrum of clinical manifestation across the different classes of vasculitis, posing a true diagnostic challenge for clinicians[2].

Given their immunological nature, vasculitides fall within the field of rheumatology; however, they often require an interdisciplinary approach depending on the organ systems involved. Although vascular inflammation can occur at any age, the underlying etiology tends to vary according to the patient's age group. In the pediatric population, ANCA-associated vasculitides are rare, with a significantly lower incidence compared to adults, and microscopic polyangiitis represents an uncommon entity within this age group. In otorhinolaryngology, the most frequently encountered vasculitides are those that affect small vessels, among which granulomatosis with polyangiitis (GPA) is the most common and typically presents with otothinolaryngological symptoms, followed by microscopic polyangiitis and Churg-Strauss granulomatosis, among others[3].

For the diagnosis and differentiation of these types of vasculitis, biopsy of the affected area is essential, complemented by laboratory testing. Diagnostic imaging may reveal suggestive findings, for example, on computer tomography (CT) or magnetic resonance imaging (MRI). but is not confirmatory[4].

Microscopic polyangiitis (MPA) is characterized as a necrotic small-vessel vasculitis belonging to the group of antineutrophil cytoplasmic antibody (ANCA) - associated vasculitides, along with GPA, with the distinction of being a pauci-immune disease[5]. It typically presents between the sixth and seventh decade of life [6], generally with renal (80%-100%), pulmonary, and cutaneous involvement[7]. Otorhinolaryngological manifestations have been reported in 9% - 30% of cases, most commonly affecting the nasal and paranasal regions and producing symptoms such as recurrent epistaxis, purulent rhinorrhea, and septal perforation, among others. Some case series have described pharyngeal involvement in approximately 17% of patients with head and neck manifestations[7]. Primary laryngeal involvement, particularly as an initial manifestation of the disease, is exceedingly rare and scarcely described in the literature, especially in pediatric patients. Therefore, otorhinolaryngological symptoms are considered infrequent at the onset of this disease.

For this reason, we present the case of a pediatric patient diagnosed with MPA at a tertiary care hospital, whose initial clinical presentation was characterized by laryngeal involvement, highlighting the diagnostic challenge posed by this atypical manifestation and its potential to delay appropriate immunosuppressive treatment. This case contributes to the limited literature on pediatric MPA with airway compromise and underscores the importance of early multidisciplinary evaluation in order to prevent airway deterioration and systemic progression.

Clinical Case

We present the case of a 16 year-old female adolescent with a history of Grave's disease diagnosed at 13 years of age, who was receiving outpatient medical treatment with methimazole and had no other significant past medical history. She presented to the emergency department of a tertiary pediatric hospital with a four -day history of neck pain, mild dysphonia, odynophagia, headache, and fever. On Physical examination, she was in poor general condition, with an enlarged palpable thyroid gland and tender bilateral cervical nodules. Initial laboratory studies revealed pancytopenia affecting all three hematologic lineages, evidence of consumptive coagulopathy and proteinuria. Based on these findings, empirical antibiotic therapy was initiated for suspected pharyngeal infection; however, her condition rapidly deteriorated, resulting in cardiopulmonary arrest that required advance resuscitation maneuvers and admission to the pediatric intensive care

unit (PICU), establishing an initial working diagnosis of septic shock.

During hospitalization, the patient was evaluated by pediatric surgery and pediatric rheumatology services. Further studies, including a neck ultrasound and contrast-enhanced CT of the neck, showed cervical lymphadenopathy without malignant features **(Figure 1).** However, immunologic and thyroid laboratory tests yielded abnormal results, with positive antinuclear antibodies (ANA), decreased complement levels (C3 and C4), and suppressed thyroid-stimulating hormone (TSH) with elevated free thyroxine, suggesting an inflammatory response secondary to macrophage activation from an adverse reaction to methimazole and thyrotoxicosis due to poor control of her underlying disease. The laboratory results obtained during hospitalization are summarized in **Table 1** and **Table 2.** Consequently, a

Table 1. Admission diagnostic labs

Lab Exam	Lab Result	Reference range
WBC	0.4 (10^3/uL)	4500-13000 (10^3/uL)
Hb	10.7 g/dL	12.1-15.1 g/dL
Hct	29.8%	35-44%
PLT	73 (10^3/uL)	150-350 (10^3/uL)
UA	63.8 mg/dL	30 mg/dL
UACR	47	< 0.2
TP	22.4	11-13.5
INR	1.9	0.8-1.2
TPT	30.7	25-35

WBC: White blood Cells, Hb: Hemoglobin, Hct: Hematocrit, PLT: Platelets, UA: Protein Albumin, UACR: Urine albumin-creatinine ratio, TP: Prothrombin time, INR International normalized ratio, PTT: Partial thromboplastin Time.

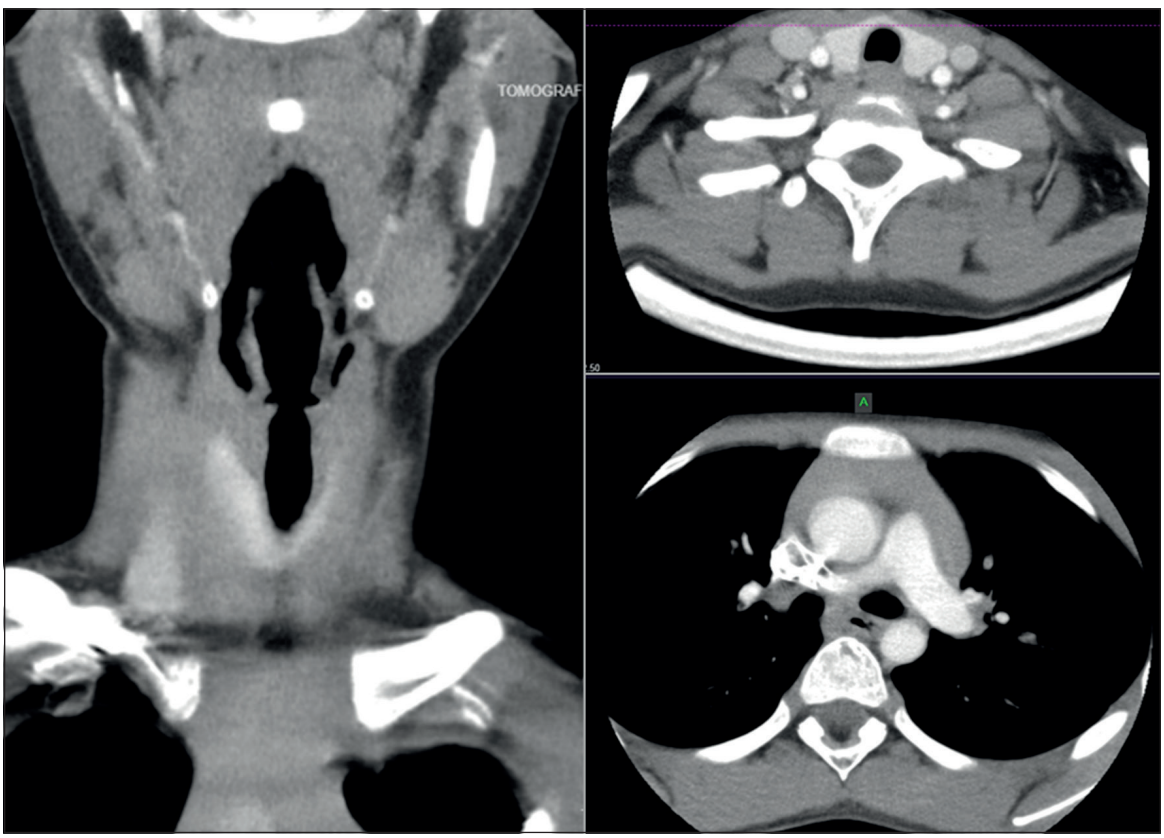

Figure 1. Contrast-enhanced neck computed tomography in axial, coronal, and sagittal planes demonstrating nonspecific cervical lymphadenopathy without evidence of masses, collections, stenosis, or other structural abnormalities. No lesions or findings are observed that compromise the patency of the upper airway.

Table 2. Inpatient diagnostic labs

Lab Exam	Lab Result	Reference range
TSH	0 mu/L	0.5-4.3 mu/L
Thyroxine	2.57 ng/dL	0.8-1.9 ng/dL
ANA	1:320	< 1:80
Complement 3	72 mg/dL	80-150 mg/dL
Complement 4	12 mg/dL	15-45 mg/dL
Anti-MPO	8.7 U/L	< 5 U/L

TSH: Thyroid-stimulating hormone, ANA: antinuclear antibody, Anti-MPO anti-myeloperoxidase antibodies.

total thyroidectomy was performed, resulting in improvement of her general condition and resolution of her initial symptoms.

Concurrently, the patient developed progressive dysphagia and dysphonia during her stay in the PICU. A consultation was requested with the pediatric otorhinolaryngology service, which performed a nasofibrolaryngoscopy revealing edema of the epiglottis and yellowish lesions on her lingual surface. The vocal folds displayed intact free edges and mobility was preserved (**Figure2 A**)**.** Consequently, the patient underwent airway exploration, which confirmed the previously described lesions and revealed right vocal fold paresis, initially suspected to be secondary to the total thyroidectomy performed by pediatric surgery. A biopsy sample was taken, but histological evaluation could not be performed due to the presence of abundant mucoid material; therefore, no additional interventions were undertaken at that time.

Subsequently, the patient developed cutaneous hyperpigmentation and motor impairment predominantly affecting the lower limbs, with laboratory confirmation of positive anti-myeloperoxidase antibodies. She was reevaluated by the pediatric rheumatology team, who confirmed findings consistent with systemic vasculitis due to microscopic polyangiitis. Among the manifestations observed in the head and neck region, this condition explained the supraglottic findings described previously. Medical management with rituximab was initiated, and after two treatment cycles, follow-up by the otorhinolaryngology service was performed. A control nasofibroblancoscopy showed persistence of partial amputation of the epiglottis with normal laryngeal mucosa and preserved mobility of both vocal folds (**Figure 2B**)**.**

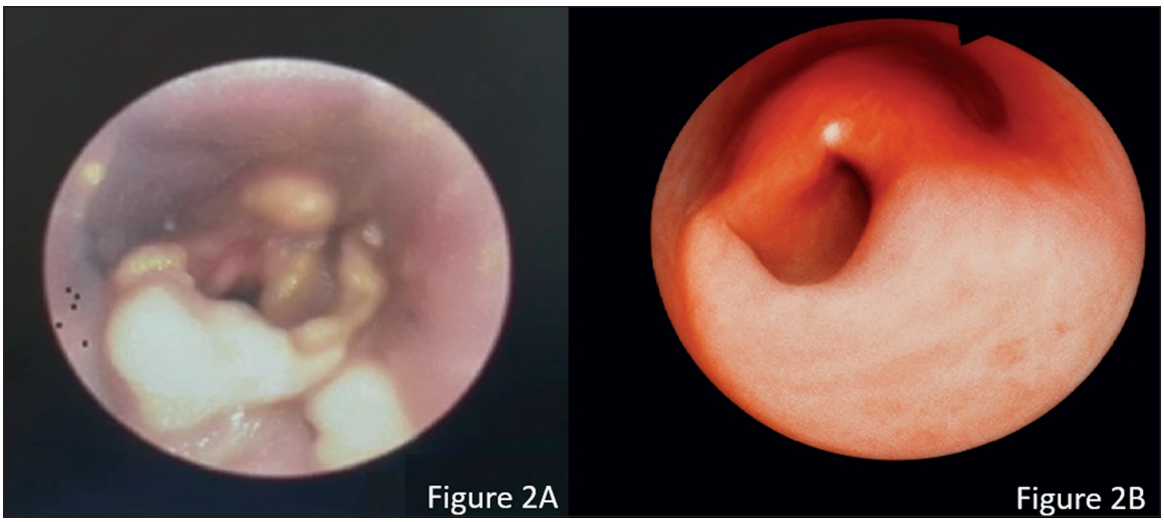

Figure 2. A) Nasofibrolaryngoscopic view showing epiglottic amputation with yellowish plaque-like lesions, edema, and erythema involving the epiglottis and hypopharynx. **B)** Follow-up nasofibrolaryngoscopic view after treatment with anti-CD20 biologic therapy, demonstrating normal laryngeal mucosa and preserved vocal fold mobility.

At present, the patient demonstrates good control of her underlying conditions and satisfactory clinical response to the medical management provided by the treating services.

Discussion

The case presented here illustrates an atypical manifestation of MPA due to its occurrence in an uncommon age group, its initial infectious-like presentation, and the patient's rapid clinical deterioration. The coexistence of a background of autoimmune disease exacerbated upon admission further contributed to the diagnostic challenge. Considering that otorhinolaryngological involvement has been described in approximately 9% to 30% of patients with MPA, its appearance as an initial symptom is rare, in contrast with GPA, where upper airway involvement is reported in 80% to 95% of cases, both belonging to the group of ANCA- associated vasculitides (AAV)[7].

Although pediatric cases of ANCA-associated vasculitis with laryngeal involvement are exceptionally rare, a previously reported case by Flores-Suárez et al. described a 16-year-old patient who initially presented with isolated unilateral vocal cord paralysis and positive MPO-ANCA antibodies, later progressing to rapidly progressive glomerulonephritis requiring renal replacement therapy. In that report, laryngeal symptoms preceded systemic involvement by several months, highlighting the potential for an indolent yet ultimately severe disease course. Similar to our case, the initial presentation mimicked more common otolaryngological conditions, delaying definitive diagnosis. However, unlike that report, our patient demonstrated early systemic progression accompanied by additional autoimmune overlap, further emphasizing the heterogeneity of pediatric MPA presentations.

The initial odynophagia and dysphonia, followed by right vocal fold paresis, raised the possibility of an unusual neurological manifestation of MPA, such as recurrent laryngeal nerve neuropathy, which has been reported in a similar case[8], rather than a postoperative complication. Although the laryngeal biopsy could not confirm vasculitis due to the predominance of mucoid material, the absence of granulomas, along with the patient's rapid systemic decline, may be explained by the systemic involvement characteristic of MPA, with renal dysfunction expected in 80% to 100% of cases[7].

The autoimmune overlap observed in this patient, given her history of Grave's disease and the new diagnosis of MPA, is noteworthy. The literature supports a genetic association between Hashimoto´s thyroiditis and an increased risk of developing MPA, but not with Grave's disease[9]. However, the presentation of this case could suggest a different, non-genetic causal relationship between thyrotoxicosis secondary to Grave's disease and the subsequent development of a new autoimmune condition in genetically predisposed patients.

Finally, the diagnostic suspicion was reinforced by the emergence of more typical manifestations of MPA, such as cutaneous involvement, reported in 30% to 60% of cases, and mononeuritis multiplex (manifested as lower lib motor impairment), observed in up to 72% of reports[8]. These findings, together with the presence of anti-myeloperoxidase antibodies, characteristic of MPA as opposed to other vasculitides (5), confirmed the diagnosis.

From an otorhinolaryngological perspective, this case reinforces the critical role of the otolaryngologist in the early recognition of atypical airway manifestations of systemic vasculitis. Progressive dysphonia, odynophagia, or vocal fold dysfunction without an identifiable structural or iatrogenic cause should prompt consideration of autoimmune etiologies, particularly when accompanied by laboratory evidence of autoimmunity. Early suspicion and timely referral to rheumatology may allow closer surveillance and earlier initiation of immunosuppressive therapy, potentially preventing irreversible organ damage.

Regarding treatment, the decision to use an anti-CD20 monoclonal antibody, rituximab (RTX), is strongly supported by high-level evidence. The RAVE-INT clinical trial demonstrated that remission induction and maintenance in severe AAV with RTX is non-inferior to conventional therapy with cyclophosphamide followed by azathioprine. Moreover, RTX regimen was associated with fewer episodes of leukopenia and pneumonia[10], lower cumulative toxicity, and reduced risk of

long-term infertility, factors of great relevance when considering treatment in pediatric patients, as in the present case.

In this context, close otorhinolaryngological follow-up is also essential during remission, as airway sequelae such as persistent vocal fold dysfunction may require rehabilitation and long-term monitoring, further underscoring the multidisciplinary nature of care in pediatric ANCA-associated vasculitis.

Conclusions

The case illustrates the rare laryngeal presentation of microscopic polyangiitis in a pediatric patient. It also highlights the importance of an interdisciplinary approach, the need to consider systemic vasculitis in atypical otorhinolaryngological presentations, and the potential role of biological therapies in complex clinical contexts.

Ethical considerations

This case report adheres to the fundamental principles of human research established in both national and international regulations, including resolution 8430 of 1993 of the Colombian ministry of health ("Scientific, Technical and Administrative Standards for Health Research") and the Declaration of Helsinki. It is classified as a study involving minimal risk. Informed consent was obtained from the patient's legal guardians for the development and publication of this work.

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