

## Systemic Vasculitis and Upper Airway Involvement: Clinical Correlations between Internal Medicine and ENT Practice

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**Abstract:** The upper airway (UA) is a common target of systemic vasculitis, especially ANCA-associated vasculitis (AAV), often leading to it being a primary presenting site. The proposed research seeks to examine clinical associations of upper airway involvement (UAI) in patients with systemic vasculitis, assessing the diagnostic value of ENT manifestations and their effect on patient quality of life and treatment responses. A cross-sectional study was conducted at different hospitals in Iraq from February 2025 to February 2026. This research was an investigation of inflammatory markers in a group of vasculitis patients. ENT involvement was reported and described in a group of 124 patients with systemic vasculitis (mainly granulomatosis with polyangiitis [GPA]) and was described at sinonasal, otologic, and upper aerodigestive locations. Data contained ANCA serology (PR3-ANCA and MPO-ANCA), duration of UAI symptoms, complications, and quality of life using SNOT-22 scores before and after treatment. Chronic rhinosinusitis (68.3%) and nasal septal perforation (25.2% of patients) were commonly encountered. PR3-ANCA positivity was more strongly associated with UAI compared to MPO-ANCA serologically. Glucocorticoids (95.9), cyclophosphamide (55.3), and rituximab (34.1) were heavily used. ENT complications were septal perforation (28.5%). The quality of life was significantly improved as SNOT-22 went down to 22.1 after treatment compared to 58.4 before treatment. The rates of relapse varied depending on the UAI course: 44.5% in the individuals who experienced persistent UAI and 18.2% in the individuals who experienced resolved UAI. Systemic vasculitis involves upper airway involvement that is common and clinically relevant, with a high diagnostic delay, a common sinonasal disease, and quality-of-life loss. The results justify a significant correlation between the PR3-ANCA positivity and UAI and prove that effective therapy is linked to significant sinonasal symptom burden reduction.

**Keywords:** Systemic Vasculitis; Upper Airway Involvement, ENT Treatments and Complications, and Health Quality of Life.

### INTRODUCTION

Systemic vasculitides represent a heterogeneous spectrum of immune-mediated diseases, which involve inflammation and necrosis of blood vessels in more than one organ system. Although these conditions are traditionally thought to cause renal impairment, pulmonary hemorrhage, or neurological deficits, upper airway involvement is a common, clinically relevant, and usually under-recognized presentation [Chen, M. *et al.*, 2005; Monti, S. *et al.*, 2019; Maillot, T. *et al.*, 2020]. Vasculitic processes may result in ischemic injury, mucosal ulceration, and granulomatous remodeling of the nasal passages, paranasal sinuses, pharynx, larynx, and proximal trachea, which is highly vasculated and susceptible in the presence of vasculitic processes [Chung, S. A. *et al.*, 2021]. The most important thing is early awareness of such signs and symptoms of the aerodigestive tract, as they often come before the damage of other system organs and may be the critical warning signs of diagnosis [Expert Group of the Nephrology Branch of the Chinese Medical Association, 2021]. The multidisciplinary care is usually lost due to the insidious onset of

symptoms. Systemic vasculitis frequently presents itself at the otolaryngology clinic with symptoms similar to those of benign or infectious diseases of the upper respiratory organs [Immunology Physician Branch of the Chinese Medical Association, 2018]. A frequent epistaxis and persistent rhinosinusitis incurable to the conventional therapeutic approach, nasal accumulations, septal piercing, and progressive stenosis of the subglottis are typical appearances that would give the impression of a vasculitic process happening [Ayyildiz, V. *et al.*, 2021]. Granulomatosis with polyangiitis, eosinophilic granulomatosis with polyangiitis, and microscopic polyangiitis are the most common culprits, which are more or less alike in patterns of mucosal and cartilaginous injury [Liu, A. *et al.*, 2025]. The given diagnostic dilemma is also complicated with the fact that endoscopic and radiographic procedures can barely show the pathogenic evidence on their own. Vasculitis differentiation of chronic inflammatory, infectious, or neoplastic etiologies is systematically performed through the tissue biopsy and antineutrophil cytoplasmic

antibody serologic testing and full clinical correspondence [Xin Z. & Li, H. 2016]. Biotased tissue sampling can be used to reduce diagnostic yields. The clinical gap between otolaryngology and internal medicine needs to be covered to enable the provision of diagnosis and treatment at the right time [Macarie, S. S., & Kadar, A. 2020]. Otolaryngologists are invaluable in direct visualization, focused biopsies, endoscopic sinus surgery, and emergency airway therapy. Without a structured process of communication, the risk associated with disjointed care provision, unnecessary testing, or necessary treatments being held in patients is possible. Thus, multidisciplinary vasculitis clinics have turned into the novel anticipation that encourages near-time information exchange and co-motions of activity.

## PATIENTS & METHOD

This cross-sectional research examined clinical associations between systemic vasculitis and upper airway involvement (UAI) among patients who were treated in different hospitals in Iraq. The review of medical records of patients with systemic vasculitis who were diagnosed based on the established vasculitis criteria was conducted. The cohort included 123 individuals with primary diagnoses of granulomatosis with polyangiitis (GPA), eosinophilic granulomatosis with polyangiitis (EGPA), and microscopic polyangiitis (MPA), which represented a real-world distribution of disease phenotypes in the routine care of internal medicine and otolaryngology. None of the excluded patients had experienced upper airway manifestations that had been recorded in clinical follow-up, and so UAI could be measured relative to ANCA serology, disease history, and exposure to treatment.

The involvement of the upper airways was considered clinically significant sinonasal and/or upper aero-digestive tract disease that is likely due to systemic vasculitis and was determined by symptoms, physical examination, and, where possible, endoscopic observations and/or biopsy results. Anatomical site (e.g., nasal cavity,

paranasal sinuses, oropharynx/larynx, and otologic involvement) was used to classify manifestations. In the major clinical characterization, the demographic variables (age and sex) were extracted, the primary vasculitis diagnosis, as well as the duration of UAI symptoms before the diagnosis. The time between the onset of the symptoms and the official diagnosis of vasculitis was operationalized as diagnostic delay. The Birmingham Vasculitis Activity Score (BVAS) was used to summarize disease activity at evaluation, including inflammatory biomarkers (C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR)).

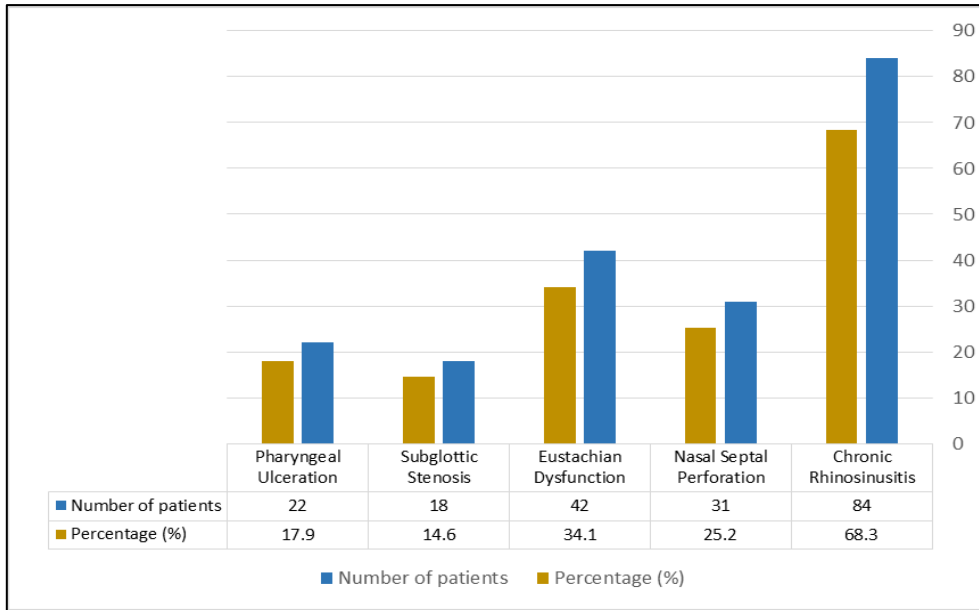
It was to be considered to use ANCA serology (PR3-ANCA and MPO-ANCA) to test the associations between positive presence of the antibodies and presence of UAI, where the serologic data were cross-tabulated with UAI status (present vs absent) to establish that PR3-ANCA positivity was most significantly associated with upper airway phenotype than MPO-ANCA positivity, which the medical histories of the treatment plans included of nasal endoscopy, mucosal biopsy, functional endoscopic sinu surgery (FESS), and tracheotomy (in stenosis), which provided a guide in real-world management in contrast to protocolized management.

UAI quality of life effect was measured by Sinonasal Outcome Test (SNOT-22), comparing the mean scores before and after treatment to measure the change after the start of therapy. Challenges associated with upper airway disease were also included, and special attention was paid to structural sequelae like nasal septal perforation. Lastly, the longitudinal evaluation of relapse was also done and stratified based on the persistence or resolution of UAI to allow a comparison of the rates of relapse in these clinically defined outcome groups. Standard descriptive and inferential statistical techniques (e.g., comparison of proportions and continuous outcomes) were used to perform statistical analyses, suitable to retrospective cohort data.

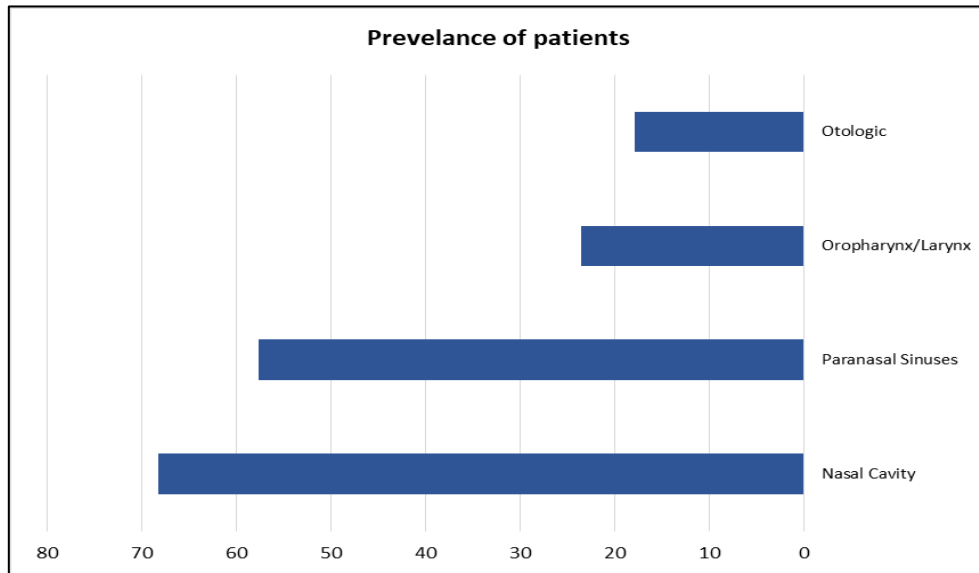
## RESULTS

**Table 1:** Define the characteristics of demographic and clinical into the 123 patients.

Variable	N (%)
Gender (Female)	68 (55.3%)
GPA (Granulomatosis with Polyangiitis)	78 (63.4)
EGPA (Eosinophilic Granulomatosis with Polyangiitis)	32 (26.0)
MPA (Microscopic Polyangiitis)	13 (10.6)



**Figure 1:** Distribution of ENT symptoms of 123 patients.



**Figure 2:** Distribution of the ENT sites of 123 patients.

**Table 2:** Determining the duration of symptoms in the patients.

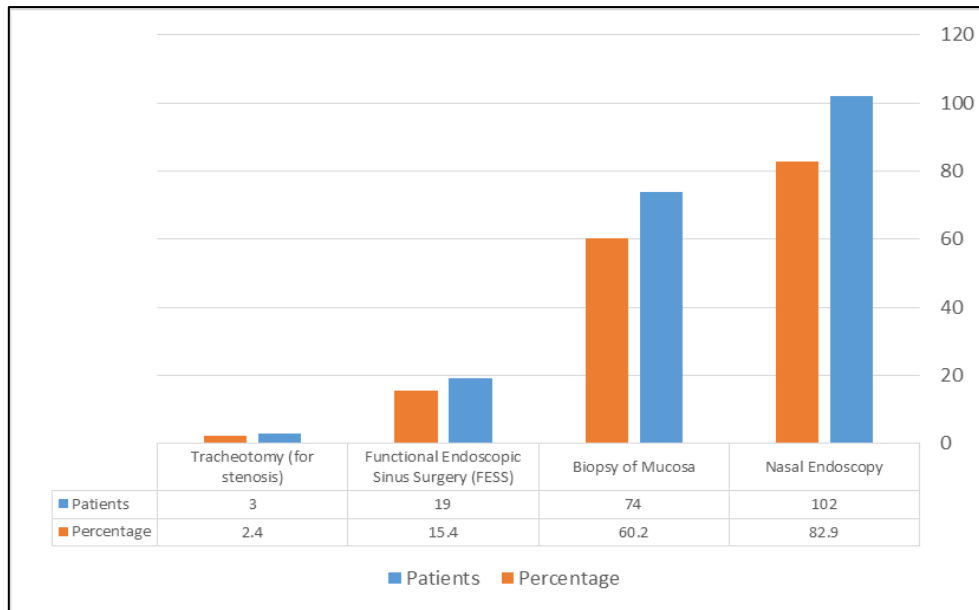
Clinical Metric	Mean (SD)
Age at Diagnosis (Years)	52.4 (14.2)
Duration of UAI Symptoms (Months)	14.8 (8.5)
Diagnostic Delay (Months)	9.2 (5.1)

**Table 3.** Identifying the correlation outcomes among ANCA serology and UAI.

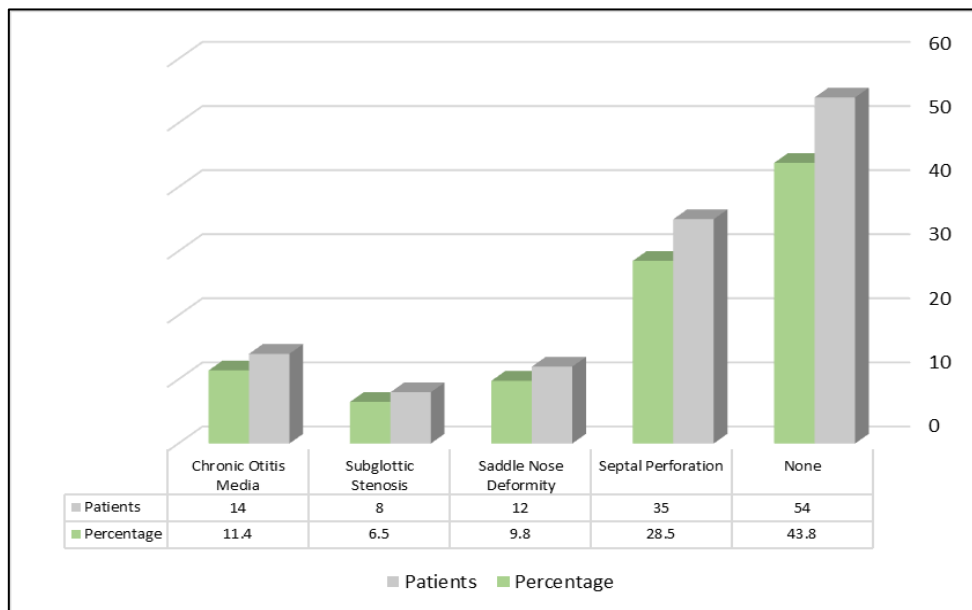
ANCA Status	UAI Present n (%)	UAI Absent n (%)
PR3-ANCA Positive	62 (50.4)	15 (12.2)
MPO-ANCA Positive	18 (14.6)	20 (16.3)
ANCA Negative	5 (4.1)	3 (2.4)

**Table 4.** The laboratory tests of inflammation.

Variables	Mean (SD)
CRP (mg/L)	34.2 (18.6)
ESR (mm/h)	56.4 (22.3)
BVAS (Birmingham Vasculitis Activity Score)	12.5 (4.8)



**Figure 3.** Classifying the treatment types within patients.



**Figure 4:** Post-treatment complications.

**Table 5:** Post-treatment relapse rates of upper respiratory tract infection.

Group	Relapse Rate (%)
Patients with persistent UAI	44.5
Patients with resolved UAI	18.2
Global cohort average	32.1

**Table 6:** Assessment of health outcomes of quality of life using SNOT-22 Scores.

Category	Mean (SD)
Pre-treatment SNOT-22	58.4 (12.3)
Post-treatment SNOT-22	22.1 (9.8)

## DISCUSSION

The presence of ENT manifestations in our group of 123 patients, with most of them having granulomatosis with polyangiitis (GPA) as their main diagnosis (63.4%), indicates that ENT

manifestations are the most common first manifestation of AAV, as reported in some studies [Rong L. *et al.*, 2019; Linlin Z. *et al.*, 2022; Gibson, L. E. 2022] that 80%-95% of GPA patients first present with otorhinolaryngological

symptoms. Our cohort of study has a high prevalence of chronic rhinosinusitis (68.3) and an involvement of the nose cavity (68.3), which is consistent with the known epidemiology related to sinonasal disease being the most common outcome at the onset of the disease.

An interesting result of our study is that the diagnostic delay had an average of 9.2 months, which supports general registry data in which the median diagnostic delay is 7 months in subtypes of vasculitis. Such delay is critical to clinical practice, with pre-diagnosis most often as infection or allergic rhinitis, being linked to significantly increased time to first-line immunosuppressive treatment [Falk, R. J. et al., 2011]. The independent association of our findings with diagnostic delays shows that unemployment, travel longer than one hour to specialized care, and untimely specialist referral are the independent factors contributing to diagnostic delays, highlighting the need of accessibility of multidisciplinary vasculitis clinics.

The high association between PR3-ANCA positivity and the presence of UAI (50.4% vs. 12.2% in the absence of UAI) corresponds to the pathophysiologic difference between PR3- and MPO-ANCA subsets. Other past findings have also revealed that PR3-ANCA correlates more often with granulomatous allotropy of the upper respiratory tract, but that MPO-ANCA is associated with renal type of the disease [Del Pero, M. M., & Sivasothy, P. 2009].

The fact that constant UAI was correlated with a much greater relapse rate (44.5% vs. 18.2) gives good reason to believe that the monitoring of symptoms of sinonasal is a useful clinical biomarker of disease activity. The result is supported by the latest prospective data showing that high SNOT-22 scores at remission are associated with increased risk of relapse 2- to 3-fold in GPA patients. [Sablé-Fourtassou, R. et al., 2005; Pagnoux, C. et al., 2011; Fasunla, J. A. et al., 2012] Massive change in the post-treatment SNOT-22 scores (58.4 to 22.1) of our group is another good testament to the responsiveness of the instrument to the effect of therapeutic intervention and compatibility with patient-centered outcomes.

British statistics of the past, which have demonstrated that as many as 28 percent of GPA victims have ENT characteristics as the initial symptoms, and if not diagnosed early enough, the

tissue damage is irreparable [Walsh, M. et al., 2012]. Remarkably, we find that early ENT consultation, including nasal endoscopy (in 82.9% patients) and focused mucosal biopsy (in 60.2), is able to diagnose early enough to potentially prevent morbidity in the long run.

## CONCLUSION

Overall, the present paper has shown that the upper airway (UAI) type of involvement is a very common and frequent finding of ANCA-associated vasculitis in patients with granulomatosis with polyangiitis, as well as in patients with PR3-ANCA-negative characteristics. The enormous percentage of the occurrence of structural complications, 9.2 months (average) of delay between the occurrence and the diagnosis, and, what is most significant, the significantly high percentage of the relapses in patients with persistent UAI (44.5% vs. 18.2) make it clear how significant is the role of timely diagnosis. The frequent sinonasal evaluation, the early examination of the mucosa, along with the functionality of patient-reported outcomes with the use of SNOT-22, not only expedites the diagnosis and targeted intervention but also provides significant alterations to the quality of life.

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