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CASE REPORT Open Access

Anti-elastase ANCA-associated vasculitis: a case report and literature review

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Abstract

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) is a systemic necrotizing vasculitis that primarily affects small- to medium-sized vessels and represents a spectrum of diseases with overlapping clinical and immunopathological features. While anti-proteinase 3 (anti-PR3) and anti-myeloperoxidase (anti-MPO) are the most common ANCA autoantibodies, other ANCA specificities, such as anti-elastase, have been described, but their clinical significance remains uncertain. We report the case of a 38-year-old male presenting with chronic rhinosinusitis, nasal crusting, and epistaxis. Imaging revealed a septal perforation, and histopathological analysis showed necrotizing vasculitis. ANCA serology was positive for perinuclear-ANCA (p-ANCA), with negative anti-PR3 and anti-MPO but positive anti-elastase antibodies. A diagnosis of incomplete AAV with a sinonasal granulomatosis with polyangiitis (GPA) phenotype was established. In order to understand the prevalence of anti-elastase antibodies in AAV, a narrative review was conducted. In six retrospective studies, prevalence varied between 0% and 16.7%, suggesting limited diagnostic utility of anti-elastase in AAV. This case highlights the relevance of histopathological confirmation in AAV, particularly in atypical ANCA profiles. Anti-elastase antibodies may have limited value as a biomarker in AAV, given their low prevalence. Immunosuppressive treatment was effective, reinforcing the importance of early recognition and intervention in AAV.

Keywords Vasculitis, Rhinosinusitis, Septal perforation, ANCA, Anti-elastase

Introduction

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAVs) is a systemic necrotizing vasculitis that primarily affects small- to medium-sized vessels and represents a spectrum of diseases with overlapping clinical and immunopathological features [1]. The major clinicopathological entities include granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA) [1]. These conditions share common mechanisms of neutrophil activation and endothelial injury but may present with heterogeneous patterns of organ involvement, ranging from localized upper airway disease to life-threatening pulmonary haemorrhage or rapidly progressive glomerulonephritis [2].

Serologically, AAVs are strongly associated with ANCAs targeting proteinase 3 (PR3-ANCA) or myeloperoxidase (MPO-ANCA), which are used in clinical practice to support diagnosis and classification. However, autoantibodies against other neutrophil granule proteins have also been described [2].

Human neutrophil elastase, a serine protease predominantly expressed in polymorphonuclear neutrophils [3], plays a critical role in the innate immune response by degrading bacterial components [3, 4]. Furthermore, it is

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a recognized antigen targeted by antineutrophil cytoplasmic antibodies, and it may display a perinuclear immunofluorescence staining pattern (p-ANCA), even in the absence of antibodies against myeloperoxidase [5]. Antibodies against elastase have been reported in patients with AAVs, but data are conflicting [6], and its clinical significance remains unclear.

Here, we report a case of AAV with isolated ENT involvement and positivity for anti-elastase antibodies, and we review the available literature on the prevalence and relevance of anti-elastase in AAV.

Case presentation

We report the case of a 38-year-old male who had been suffering from yellowish nasal discharge and persistent nasal congestion for 3 years, as well as intermittent epistaxis, with no seasonal variation. He had been treated with short cycles of topical and oral corticosteroids, with a maximum dose of prednisolone 40 mg daily, with improvement of his symptoms, especially with oral steroids. Additionally, he complained of an occasional mild dry cough. There were no constitutional symptoms or other system-specific complaints. His past medical history included idiopathic pericarditis 10 years before and a giant cell tumour of the radius. He was not taking any chronic medication. There was no history of drug abuse, including cocaine, and no family history of inflammatory rheumatic diseases. Figure 1 represents the timeline of events.

He was first referred to our university hospital for an appointment with the otorhinolaryngology (ENT) department, in which clinical examination revealed thick and dry crusts in the nasal cavities, compatible with extensive ozena, as well as nasal perforation, which was confirmed by a sinus computed tomography (CT) scan. He underwent surgical debridement of the obstructive crusts, during which extensive anterior septal perforation, mucopurulent secretions, and friable mucosa were found. Cleaning of crusts and secretions was carried out, and a sample was collected for microbiological analysis. Biopsies were performed on the nasal septum and right inferior nasal turbinate.

Suspicion for vasculitis was high at this point, and blood tests were performed which revealed a mild eosinophilia (eosinophils: 1.16 g/L; normal range: 0.02–0.50 g/L) without other hemogram abnormalities (Table 1). There was no evidence for renal or hepatic impairment, and acute phase reactants including erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were within normal limits. Serological testing was positive for p-ANCA (perinuclear anti-neutrophil cytoplasmic antibodies) by immunofluorescence, with negative anti-proteinase 3 (anti-PR3) and anti-myeloperoxidase (anti-MPO) antibodies but positive anti-elastase antibodies. Viral serologies and interferon-gamma release assay (IGRA) were negative. Parasitological faecal cultures were negative. Microbiological analysis of the nasal sample revealed colonization by *Staphylococcus aureus*.

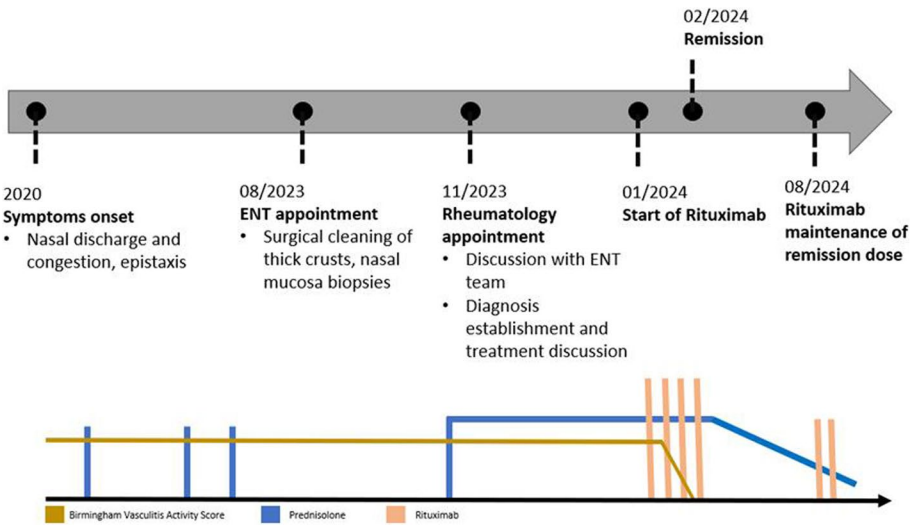

Fig. 1 Timeline of events. The *Y*-axis has relative units according to each variable. Birmingham Vasculitis Activity Score (BVAS) was used to measure disease activity and was scored with 4 points since the patient had crusts and bloody discharge; in February 2024, he achieved remission (*BVAS* = 0 point). Oral prednisolone was prescribed for short periods of time before diagnosis; at the moment of the diagnosis, it was started at 40 mg/day and tapered after remission to 5 mg/day. Rituximab was started according to the RAVE protocol (375 mg/m^2, 4 weekly infusions) to achieve remission and was kept at a maintenance dose of 500 mg every 6 months (two infusions 6 months after the induction of remission)

Table 1 Laboratorial findings for diagnostic workup. ANCA were identified by imunofluoresecence. Anti-PR3 and anti-MPO were determined by FEIA. Anti-elastase was determined by ELISA, although the specific unit of measurement was not provided in the laboratory report

	Diagnosis	Follow-up	Reference range
Creatinine	0.84	1.00	0.72–1.18 mg/dL
Urinalysis	No active sediment	No active sediment	-
Urinary protein/creatinine ratio	41	45	< 200 mg/g
B-type natriuretic peptide	5.2	-	< 100 pg/mL
CRP	0.17	0.08	< 0.50 mg/dL
ESR	6	7	1–20 mm/h
Leucocytes	8.8	8.1	3.9–10.2 g/L
Neutrophils	3.7	4.5	1.5–7.7 g/L
Eosinophils	**1.16**	0.38	0.02–0.50 g/L
Platelets	394	317	150–450 g/L
p-ANCA	**Positive**	-	-
c-ANCA	Negative	-	-
Anti-PR3	< 0.2	-	< 2.0 IU/mL
Anti-MPO	0.2	-	< 3.5 IU/mL
Anti-elastase	**Positive**	-	-
Antinuclear antibodies	Negative	-	-
Rheumatoid factor	< 9	-	< 20 IU/mL
IgG	12.03	9.23	5.40–18.22 g/L
IgA	2.49	1.71	0.63–4.84 g/L
IgM	0.56	0.31	0.22–2.40 g/L
IgE	< 16	-	< 100 IU/mL
HIV (serology)	Negative	-	-
HCV (serology)	Negative	-	-
HBV (serology)	Immune	-	-
IGRA	Negative	-	-
Parasitological faecal culture	Negative	-	-
Nasal mucosa culture	*S. aureus*[a]	-	-

c-ANCA cytoplasmic antineutrophil cytoplasmic antibody, *anti-PR3* anti-protease 3, *anti-MPO* anti-myeloperoxidase, *IGRA* interferon-gamma release assay, *HIV* human immunodeficiency virus, *HBV* hepatitis B virus, *HCV* hepatitis C virus, *Ig* immunoglobulin

[a] *S. aureus* was interpreted as colonization

Histological assessment revealed ulcerated mucosal flaps with necrosis and haemorrhage and pronounced neovascularization with polymorphonuclear infiltrate containing numerous eosinophils, an arterial vessel with some parietal thickening, luminal occlusion, and focal inflammatory permeation with histiocytic cells, as well as possible focal fibrinoid necrosis (Fig. 2).

A thoracoabdominal CT scan and pulmonary function tests were also performed, without any abnormalities.

A multidisciplinary discussion was made between rheumatology and otorhinolaryngology: ENT examination was suggestive of granulomatosis with polyangiitis, and there was no evidence of infection, despite *S. aureus* colonization. Although the patient presented p-ANCA positivity with anti-elastase antibody, being anti-MPO or anti-PR3 negative, he had clinical and histological signs of vasculitis, and there was no evidence of other mimickers of vasculitis or cocaine use. He was diagnosed with incomplete AAV with a sinonasal GPA phenotype, considering the septal perforation and the absence of an allergic background, especially asthma. There was also no evidence of systemic manifestations of ANCA-associated vasculitis.

After informing the patient about the diagnosis, rituximab was considered as the first line of treatment, with concomitant high-dose systemic corticosteroid. He started induction of remission with rituximab according to the RAVE protocol (375 mg/m^2 weekly for 4 weeks) [7] and prednisolone 40 mg per day. There was a great improvement in his nasal symptoms, including the resolution of nasal discharge, crusting, and septal ulcers. Nasal cavity patency was restored.

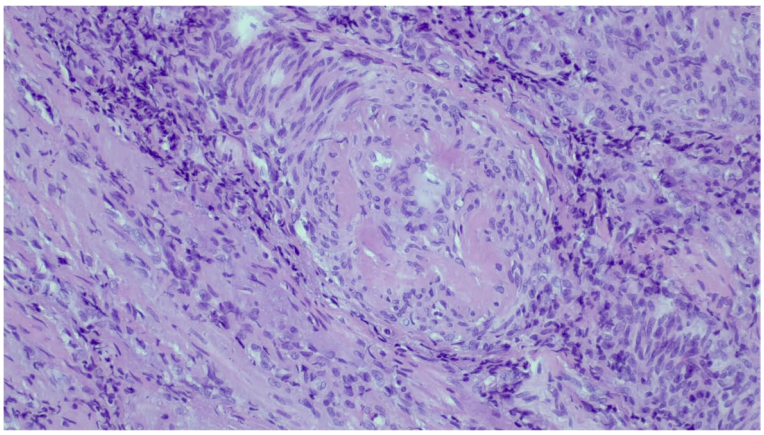

Fig. 2 Haematoxylin and eosin staining of nasal mucosa (100× magnification). An arterial vessel shows parietal thickening, luminal occlusion, and focal transmural inflammation composed predominantly of histiocytic cells, with possible focal fibrinoid necrosis

For maintenance of remission, the patient repeated rituximab 500 mg twice after 6 months, and repeat treatments at least every 6 months were planned for a total of 24 months. Prednisolone was also gradually tapered to 5 mg per day in 8 months.

The patient has remained in remission under the care of rheumatology and ENT teams, with no recurrence of ENT or systemic vasculitis symptoms. No adverse events related to rituximab were reported.

Discussion

The presented case illustrates a manifestation of incomplete AAV with a clinical phenotype of GPA limited to the upper airway, in which the histological analysis was key for the diagnosis. The patient had clear signs of chronic rhinosinusitis that only improved with systemic corticosteroid therapy, despite never achieving complete remission. A septal perforation was also detected on clinical examination and confirmed by sinus CT scan. However, there were no other manifestations suggestive of systemic vasculitis or acute phase reactants elevation. ANCAs were tested, revealing a perinuclear pattern by immunofluorescence, with a negative anti-MPO but a positive anti-elastase. Anti-elastase has also been reported in patients with cocaine-induced midline destructive lesions [8], but the patient denied any cocaine use. Although the nasal mucosa culture was positive for *S. aureus*, it was interpreted as colonization by the ENT team, which was supported by the marked clinical improvement with immunosuppressive therapy alone, without any antibiotic treatment. Furthermore, *S. aureus* carriage may be linked to the pathophysiology of GPA [9].

Combining the clinical context and histological findings, in the absence of a vasculitis mimic, a diagnosis of incomplete AAV with a clinical phenotype of sinonasal GPA was made. Interestingly, this patient fulfilled the 2022 American College of Rheumatology (ACR)/European Alliance of Associations for Rheumatology (EULAR) Classification Criteria for Eosinophilic Granulomatosis with Polyangiitis (EGPA) [10], since he had a blood eosinophil count superior to 1×10^9/L and extravascular eosinophilic predominant inflammation on biopsy, and there was no haematuria, positive c-ANCA, or positive anti-PR3 (7 points). On the other hand, he did not fulfil the 2022 ACR/EULAR Classification Criteria for GPA [11], since there was eosinophilia and a positive p-ANCA despite the nasal and cartilaginous involvement (0 points). However, perforation of the nasal septum almost never occurs in EGPA [12], and almost all patients have asthma [13]. The presence of eosinophilia and eosinophilic inflammation does not exclude a diagnosis of GPA, since it may be present in some patients [14]. Therefore, the case was defined as incomplete AAV, although the clinical presentation corresponded to a sinonasal GPA phenotype.

Treatment was based on the EULAR recommendations for the management of ANCA-associated vasculitis regarding non-organ-threatening and non-life-threatening GPA, so a combination of rituximab and glucocorticoids for induction of remission was started [15]. The rituximab regimen followed the RAVE protocol [7], but corticosteroids did not follow the PEXIVAS protocol: prednisolone was started at a dose of 40 mg/day and tapered to 5 mg/day in 8 months. Remission was achieved 1 month after induction treatment with rituximab, and there were no signs of relapse during follow-up.

The presence of anti-elastase has been reported in AAVs [5, 6, 16, 17], propylthiouracil-associated ANCA vasculitis [18], cocaine-induced midline destructive lesions [8], and levamisole-induced vasculitis [19]. It has

also been reported in other diseases such as rheumatoid arthritis [20], systemic lupus erythematosus [6, 16], mixed connective tissue disease [16], systemic sclerosis [6], and post-streptococcal glomerulonephritis [16]. However, to the best of our knowledge, there are no studies determining the diagnostic performance of anti-elastase in AAV, whereas the diagnostic performance of anti-MPO and anti-PR3 in AAVs was established [21]. Thus, we performed a narrative review of the literature using the MEDLINE/PubMed database up to December 2024, to assess the prevalence of anti-elastase in AAV patients. We used the query "("elastase") AND ("ANCA-associated vasculitis" OR "AAV" OR "granulomatosis with polyangiitis" OR "GPA" OR "Wegener granulomatosis" OR "microscopic polyangiitis" OR "MPA" OR "eosinophilic granulomatosis with polyangiitis" OR "EGPA" OR "Churg-Strauss")". A total of 131 publications were identified through our search strategy, screening their titles and abstracts for relevance. We focused on original studies that reported data from AAV cohorts, assessed the presence or prevalence of anti-elastase antibodies, and employed a defined serological method for antibody detection. Case reports, review articles, studies not published in English, animal or in vitro studies, those lacking clear AAV diagnostic criteria, and duplicate or overlapping cohorts were excluded. Six studies met our inclusion criteria, all of which were single-centre, retrospective designs.

Table 2 summarizes the prevalence of anti-elastase antibodies across the included studies. The sample size of AAV cohorts ranged from 12 to 192 patients, with a total sample of 393 AAV cases. Four studies were conducted in European populations, one in North America and one in Australia. AAV and its subtypes' diagnosis was based on established classification systems, including the International Consensus Conference on Nomenclature of Systemic Vasculitis, the American College of Rheumatology (ACR) criteria, or the Chapel Hill Consensus Conference definitions.

Among the six included studies, 30 cases of anti-elastase positivity were reported in 393 AAV patients, corresponding to an overall prevalence of 7.6%. In all included studies, anti-elastase antibodies were detected by enzyme-linked immunosorbent assay (ELISA). The reported prevalence varied widely, ranging from 0.0% to 16.7%. The largest studies reported 12.0% and 0.0% prevalence rates, suggesting a low sensitivity of anti-elastase antibodies for AAV diagnosis. Regarding the AAV subtypes, the presence of anti-elastase antibodies ranged from 0.0% to 19.2% in MPA and 0.0% to 16.7% in GPA. These antibodies were absent in EGPA in one study, as well as in nonclassified AAV in another study. Given the low prevalence of anti-elastase in AAV, these antibodies may have limited value as a disease-specific biomarker. However, these studies have small sample sizes and were not designed to assess their diagnostic performance.

Despite these findings, several limitations must be acknowledged. First, the included studies were all retrospective and conducted in single centres, introducing potential selection bias and limiting generalizability. Second, sample sizes were small, which could increase the likelihood of random variation in prevalence estimates. Third, the heterogeneity in the diagnostic criteria used to define AAV across studies may have led to differences

Table 2 Studies regarding the prevalence of anti-elastase positivity in AAV. AAV and its subtypes were diagnosed according to the International Consensus Conference on Nomenclature of Systemic Vasculitis, the American College of Rheumatology criteria, or the Chapel Hill Consensus Conference

Study	AAV sample size *N*	Number of anti-elastase cases in AAV patients *N* (%)	Study design	Methods for testing anti-elastase
S. Apenberg et al. [6]	192 [GPA: 108; MPA: 78, EGPA: 6]	23 (12.0%) [GPA: 8 (7.4%), MPA: 15 (19.2%), EGPA: 0 (0.0%)]	Single-centre study Retrospective	ELISA
J. A. Savige et al. [22]	52 [GPA: 22, MPA: 14, nonclassified AAV: 16]	2 (3.8%) [GPA: 1 (4.5%), MPA: 1 (7.1%), nonclassified AAV: 0 (0.0%)]	Single-centre study Retrospective	ELISA
M. V. Talor et al. [23]	42 [GPA: 32, MPA: 10]	2 (4.8%) [GPA: 2 (6.3%), MPA: 0 (0.0%)]	Single-centre study Retrospective	ELISA
Olaf Wiesner et al. [8]	78 [GPA: 64, MPA: 14]	0 (0.0%) [GPA: 0 (0.0%), MPA: 0 (0.0%)]	Single-centre study Retrospective	ELISA
A. C. Muller Kobold et al. [24]	17 [GPA: 17]	1 (5.9%) [GPA: 1 (5.9%)]	Single-centre study Retrospective	ELISA
S. N. Wong et al. [25]	12 [GPA: 12]	2 (16.7%) [GPA: 2 (16.7%)]	Single-centre study Retrospective	ELISA

Subtype distribution of AAV is presented in brackets beneath the total patient number

in patient selection and, consequently, in the prevalence of anti-elastase antibodies. Finally, none of the included studies was specifically designed to evaluate the diagnostic performance of anti-elastase antibodies in AAV. As such, data on sensitivity, specificity, or predictive value are lacking. This precludes any firm conclusions regarding their clinical utility.

Taken together, these limitations highlight the need for future multicentre studies with larger sample sizes and standardized assays for detecting anti-elastase antibodies, designed to assess their diagnostic performance, including their sensitivity and specificity for AAV. This would improve our understanding of their role in AAV pathogenesis and diagnostic utility, potentially clarifying whether they hold diagnostic relevance in AAV or merely reflect an epiphenomenon.

Conclusion

We report a case of incomplete AAV with a clinical phenotype of sinonasal GPA in a patient with p-ANCA positivity directed against anti-elastase. As demonstrated in our review, the role of anti-elastase antibodies in the diagnosis of AAV is unclear, since their prevalence is low and they may be present in other autoimmune and inflammatory conditions. This underscores the essential role of histopathology in confirming AAV, particularly in cases with atypical ANCA profiles. Immunosuppressive treatment with rituximab and corticosteroids led to complete remission, reinforcing the importance of early recognition and appropriate therapeutic intervention. Since there is no data regarding the diagnostic performance of anti-elastase antibodies in AAV, future studies should explore the potential clinical significance of these antibodies to determine whether they hold any diagnostic relevance.

Abbreviations

AAV	ANCA-associated vasculitis
ACR	American College of Rheumatology
ANCA	Antineutrophil cytoplasmic antibody
Anti-MPO	Anti-myeloperoxidase
Anti-PR3	Anti-proteinase 3
c-ANCA	Cytoplasmic antineutrophil cytoplasmic antibody
CRP	C-reactive protein
CT	Computed tomography
EGPA	Eosinophilic granulomatosis with polyangiitis
ELISA	Enzyme-linked immunosorbent assay
ENT	Otorhinolaryngology
EULAR	European Alliance of Associations for Rheumatology
ESR	Erythrocyte sedimentation rate
GPA	Granulomatosis with polyangiitis
IGRA	Interferon-gamma release assay
MPA	Microscopic polyangiitis
p-ANCA	Perinuclear antineutrophil cytoplasmic antibody

Authors' contributions

All authors meet the authorship criteria recommended by the ICMJE. FA: conceptualization, data collection, drafting of the manuscript, and literature review. MN, MJC, JO, BS, SC, FC, MJS: critical revision for intellectual content, clinical input, and manuscript editing. All authors approved the final version of the manuscript and agree to be accountable for all aspects of the work.

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Data availability

Data is provided within the manuscript.

Declarations

Ethics approval and consent to participate

Written informed consent to participate in this case report was obtained from the patient.

Competing interests

The authors declare no competing interests.

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