Unusual presentations of AAV

R.FARROKH SERESHT

MD.RHEUMATOLOGIST

HORMOZGAN UNIVERSITY OF MEDICAL SCIENCES

AAV is a group of rare diseases with an estimated prevalence of 200–400 cases per million people

- GPA
- Microscopic polyangiitis (MPA)
- EGPA

Among all three types of AAV, GPA is the most common

GPA typically involves

- The upper respiratory tract (sinusitis , …..)
- Lungs (nodules,mass, ILD, DAH,…..)
- Kidneys (GN, TIN)

Why diagnosis is delayed

Any organ can be involved

The spectrum and severity of the disease are heterogeneous

- Indolent disease involving only one site
- Fulminant disease with multi-organ involvement

The variety and non-specificity of symptoms often contribute to diagnostic delays

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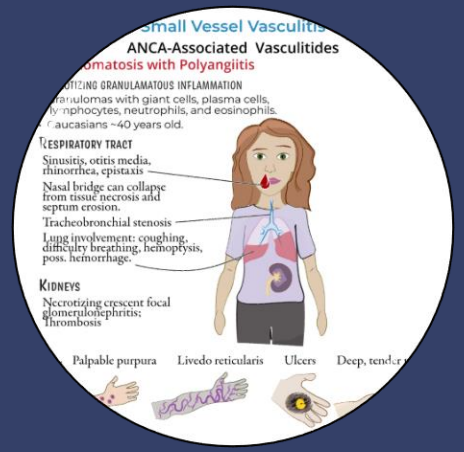
Letter to the Editor

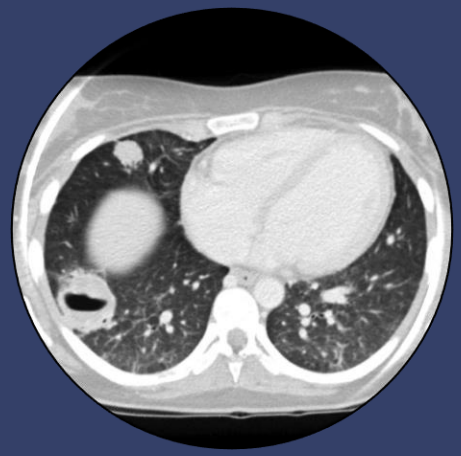
Patient-driven online survey on the clinical manifestations and diagnostic delay of granulomatosis with polyangiitis

Elaine Yacyshyn [a], Andrea Johnson [a], Marta Rode [b], Christian Pagnoux [c]

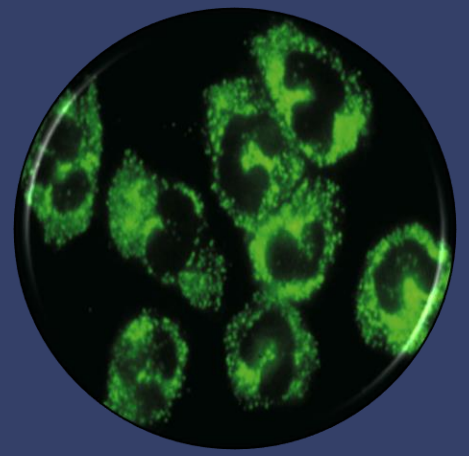
The time from initial symptoms to diagnosis varied greatly, from one month to over three years, with 36% of all symptoms reported more than one year prior to diagnosis



Diagnosis is mostly based on clinical features



Laboratory and imaging findings



ANCA and/or Biopsy

The classic form of GPA involves upper airway ear, nose, throat,lung and kidney involvement; however, any organ can be affected

- Tracheobronchial inflammation may occur anywhere in the tracheobronchial tree
- Tracheobronchial involvement may occur at any stage of the disease, even in patients being in remission
- On rare occasions, it may be the only feature of GPA, making a proper diagnosis difficult, especially in cases of ANCA negativity

The pattern of lesions is heterogeneous

Four main types are distinguished

- Mucosal abnormalities
- Masses and polyps
- Subglottic stenosis (SGS)
- Tracheobronchial inflammation and stenoses (TBISs)

Other rarer complications include tracheo- or bronchomalacia as well as bronchiectasis , with the latter occurring more frequently in patients with AAV and MPO positivity

SGS is the most common manifestation of tracheobronchial involvement in GPA occurring in 10–20% of patients

May occur in any stage of the disease

Patients may present with progressive dyspnea and stridor, dry cough, wheeze, and hoarseness

The diagnostic challenges are cases of isolated SGS, without the presence of ANCA, where the diagnosis should be based on histological examination, after exclusion of other causes

TBISs are less frequent than SGS

Often associated with GPA activity elsewhere

The inflammation may be localized or complex involving multiple tracheal or bronchial segmental stenoses

GPA affecting the airways occasionally leads to tracheo- or bronchomalacia

- Dynamic collapse during expiration or inspiration

Bronchiectasis is more prevalent in AAV with MPO-ANCA positivity and associated with distinct phenotypes

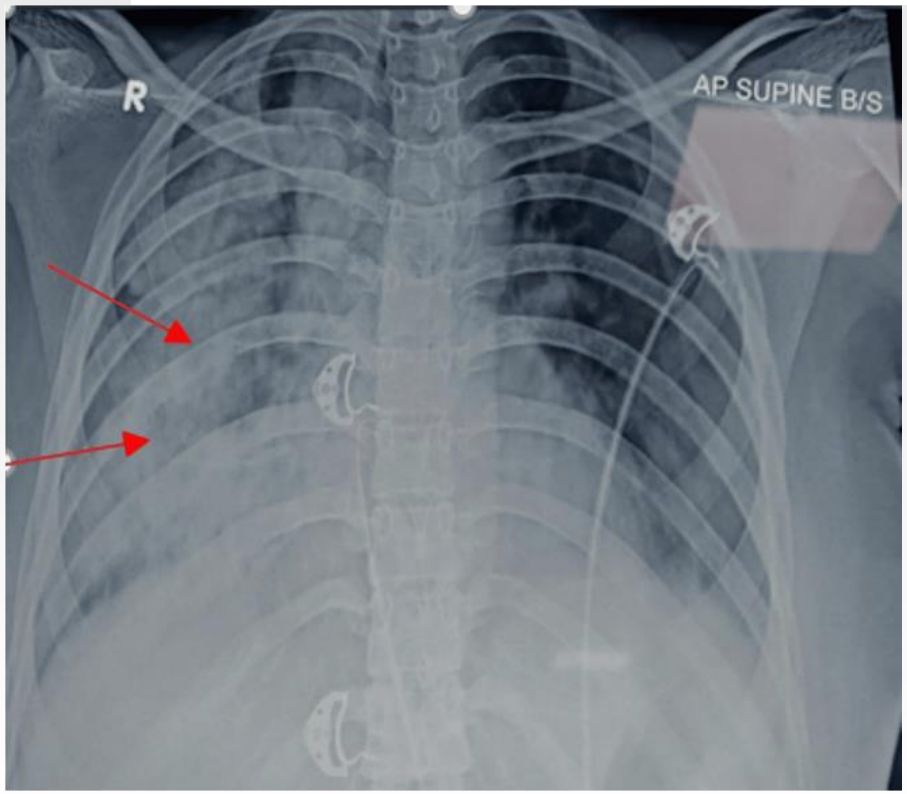
- Female, older age, common nerve involvement and renal disease

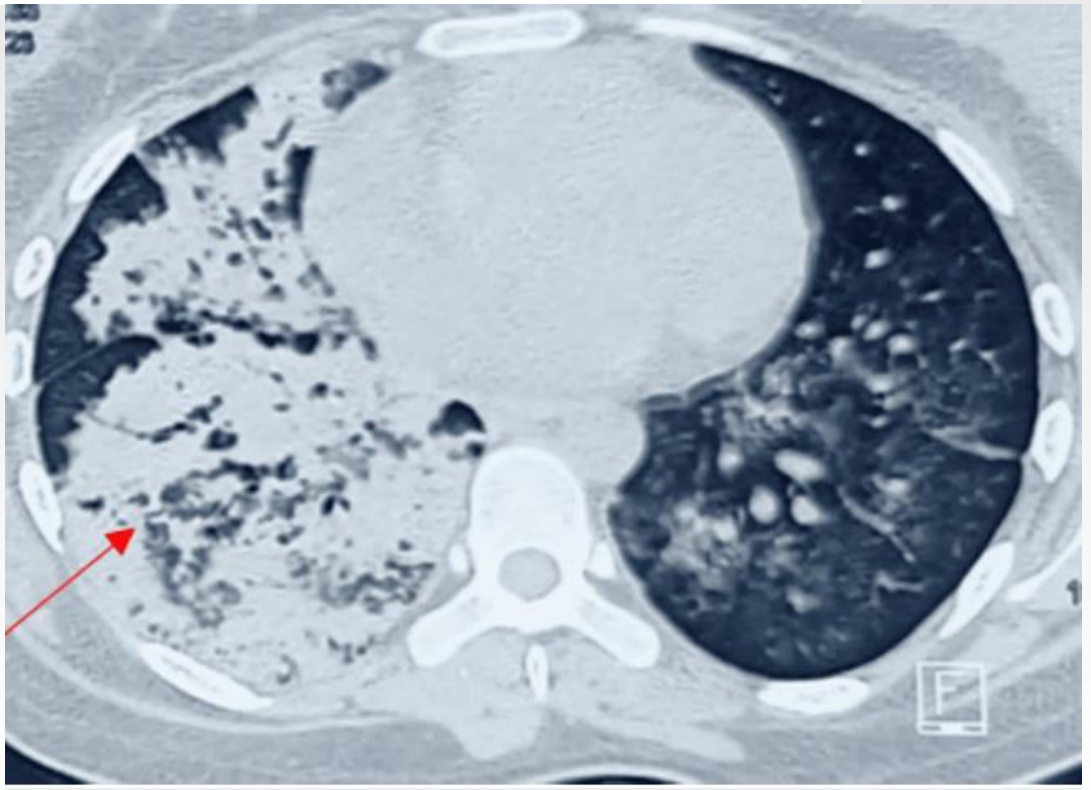
DAH

- The frequency of DAH in GPA cases ranges from 8.8 to 36% of patients
- It is frequent in MPA (25–60%) , and rare in EGPA (0–4%)

Unilateral Diffuse Alveolar Hemorrhage as an Initial Presentation of Microscopic Polyangiitis

Tapas Ranjan Behera [1] , Amninder Kaur [2] , Satyajit Acharya [3] , Prabhat Mallick [1] , Alisha Sahu [4]





ILD

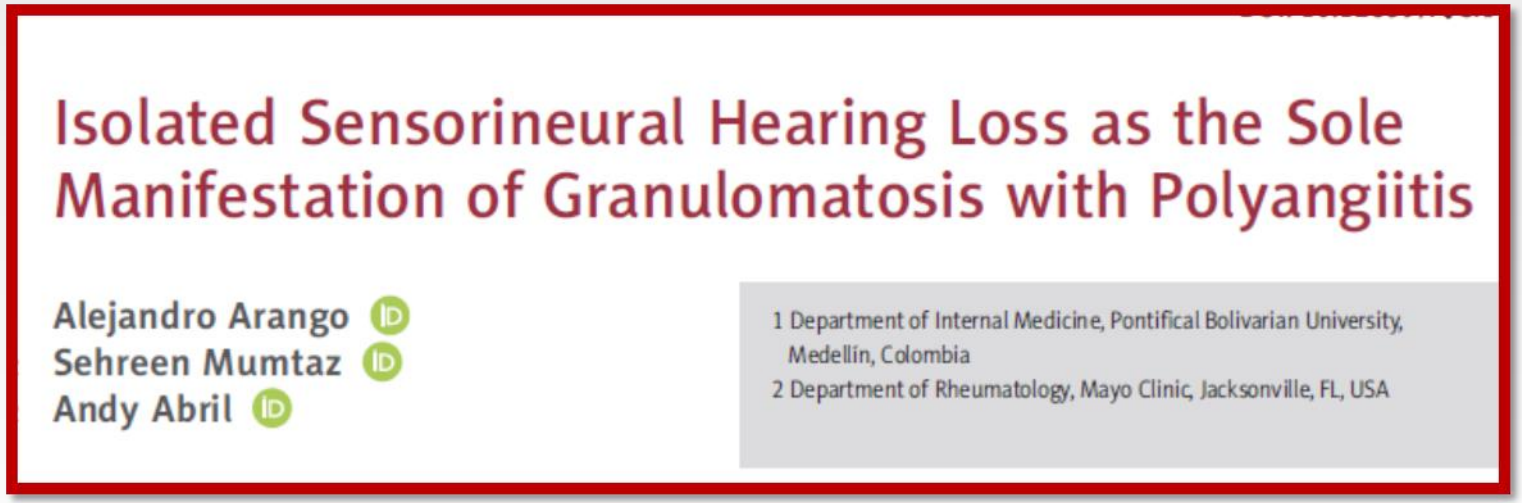
- The ILD is more likely to affect MPA than GPA
- More prevalent in MPO-ANCA than PR3-ANCA
- It is suggested that MPO-ANCA may play a direct role in the pathogenesis of ILD
- ILD may occur in any time; that is before, simultaneously or after diagnosis of AAV

Extrapulmonary involvement

- Nasal crust formation (75%)
- Excessive nose-blowing (70%)
- Nasal obstruction (65%)
- Epistaxis (59%)
- Septal perforation
- Saddle-nose deformity

Ear involvement is almost always secondary to nasal involvement

- Leads to hearing loss, which may be the first GPA symptom

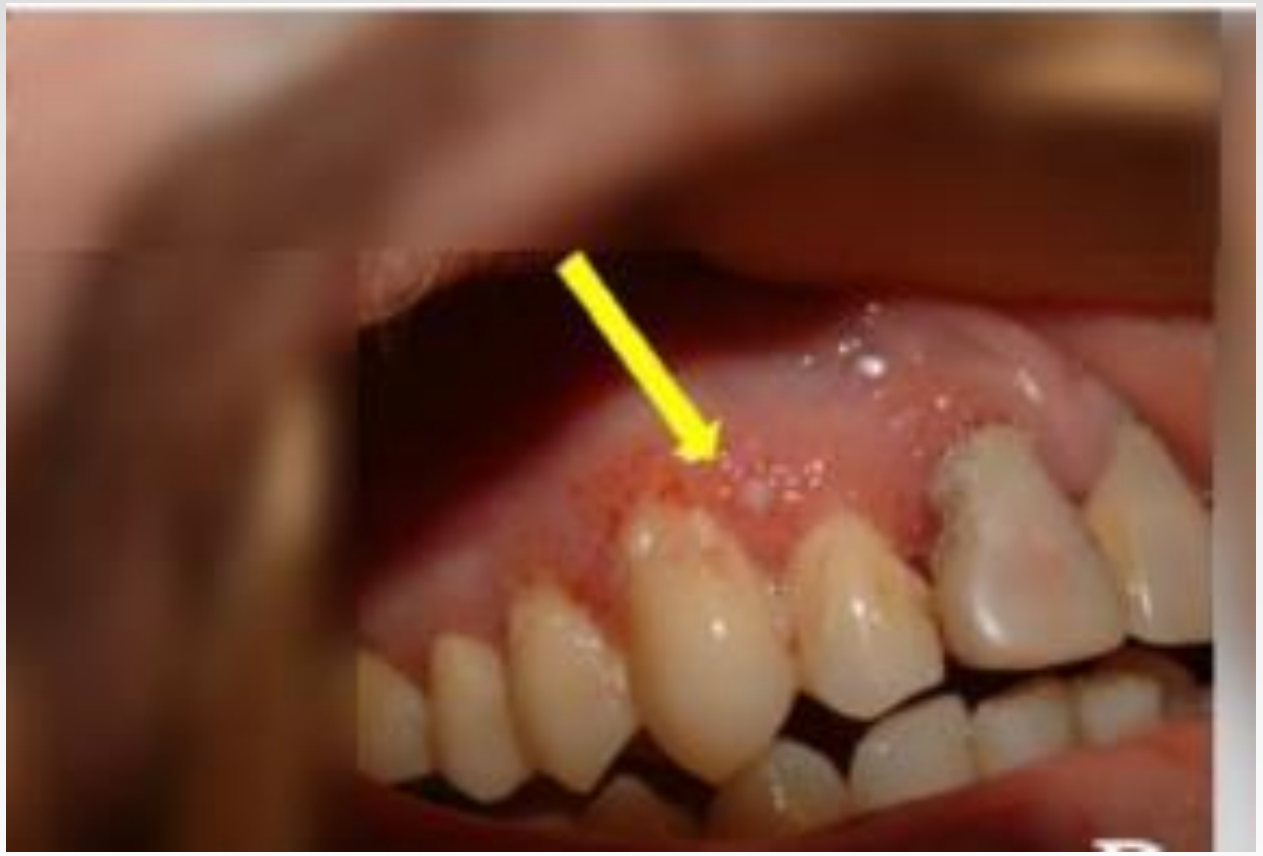


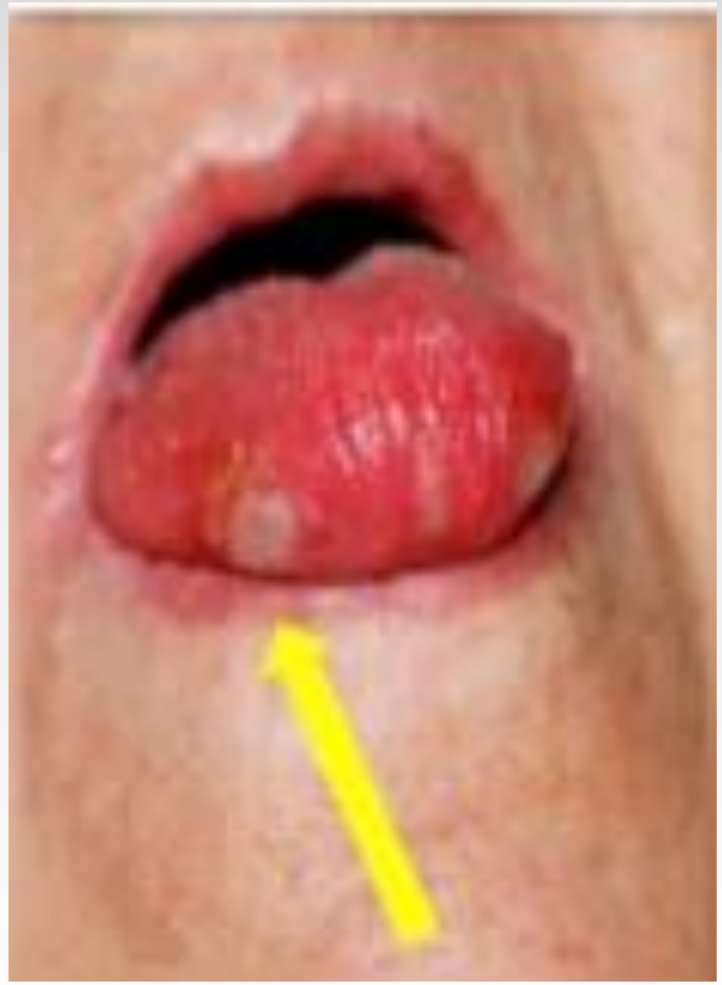
Isolated Sensorineural Hearing Loss as the Sole Manifestation of Granulomatosis with Polyangiitis

Alejandro Arango
Sehreen Mumtaz
Andy Abril

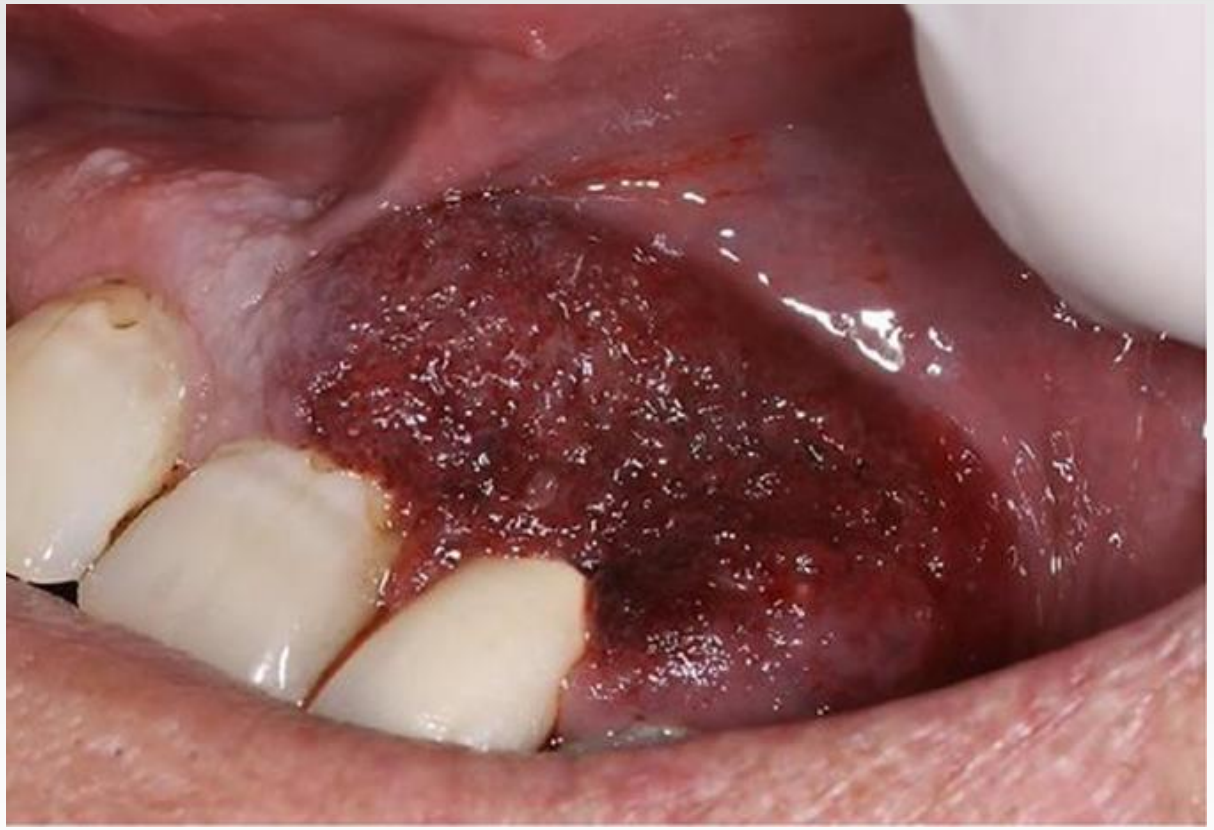
1 Department of Internal Medicine, Pontifical Bolivarian University, Medellín, Colombia
2 Department of Rheumatology, Mayo Clinic, Jacksonville, FL, USA

Involvement of the oral cavity is present in 6–13% of patients





Strawberry” gingival hyperplasia is the most characteristics sign of GPA



The frequency of cardiac involvement ranges from 1 to 61%

- Clinically overt cardiac involvement is reported in 3.3% of patients
- Pericarditis and myopericarditis are the most common cardiac manifestation

Peripheral nervous system (PNS) involvement

- The classic presentation is pain of acute onset, weakness, and sensory loss in the distribution of a named nerve (mononeuritis)
- Involvement of additional nerves in a stepwise fashion over weeks to months (multifocal neuropathy or mononeuritis multiplex)

Central nervous system (CNS) involvement is rarely seen in GPA

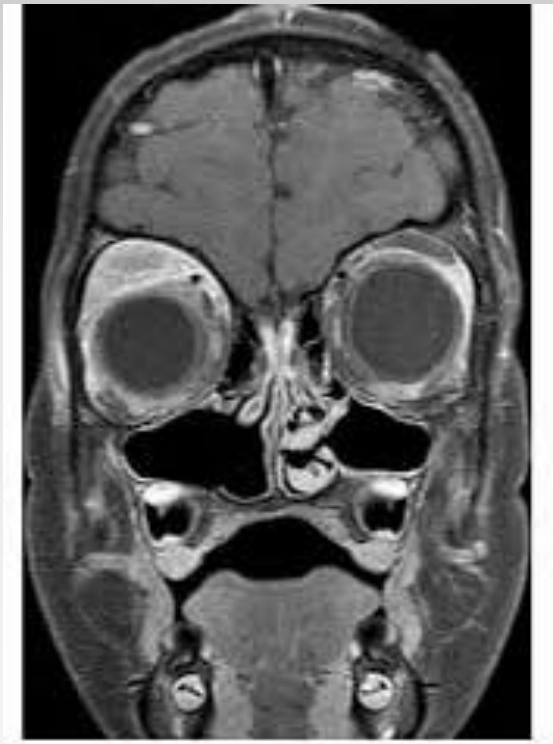
Hypertrophic Pachymeningitis: An Unusual Cause of Headache

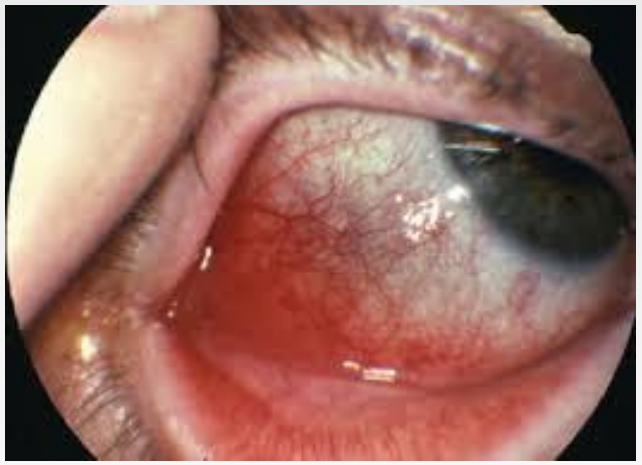
Joud Enabi [1], Muhammad Waqar Sharif [1], Raksha Venkatesan [1], Hema Kondakindi [1], Maida Faheem [2]

1. Internal Medicine, Texas Tech University Health Sciences Center, Odessa, USA 2. Internal Medicine, Midland Memorial Hospital, Midland, USA

Eye involvement

- Orbital lesions (manifesting as pseudotumor or granulomatous inflammation)
- Conjunctivitis
- Episcleritis
- Scleritis
- Dacryoadenitis





Necrotizing Scleritis as a Manifestation of Eosinophilic Granulomatosis With Polyangiitis: A Case Report

Kidney involvement

- IgA nephropathy
  - May coincide with AAV
  - May present months before diagnosis of AAV
  - MPO-ANCA positivity
- Isolated tubulointerstitial nephritis (TIN)
- Patients with high amounts of proteinuria may have a second glomerular disease
  - Mamberanous nephropathy
  - An atypical histologic pattern (full-house immune complex deposit)

Finally, gastrointestinal involvement has been reported in 1–26.5% of cases

- The most frequently symptoms include abdominal pain and bloody diarrhea

Take home messages

- AAV is a heterogeneous multisystem disease; any organ may be involved, leading to delayed diagnosis
- Airway and pulmonary involvement are often under-recognized (subglottic stenosis, DAH, ILD)
- ENT, oral (“strawberry gingivitis”), eye, GI, and CNS findings may be key diagnostic clues
- Renal disease is not always classic necrotizing crescentic GN
- Maintain a high index of suspicion in unexplained inflammatory multi-organ disease