

Mimics of primary systemic vasculitides¹¹



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Medications associated with drug-induced vasculitis

Antibiotics

Cephotaxime
Minocycline
Anti-thyroid drugs
Benzylthiouracil
Carbimazole
Methimazole
Propylthiouracil

Anti-tumour necrosis factor- α agents

Adalimumab
Etanercept
Infliximab

Psychoactive agents

Clozapine
Thioridazine

Miscellaneous drugs

Allopurinol
D-Penicillamine
Hydralazine
Levamisole
Phenytoin
Sulfasalazine

True vasculitis or mimics of vasculitis

A number of drugs, may be associated with positive ANCA testing, and may induce a true vasculitic syndrome.

This presentation is only focused on drugs that mimic vasculitis.

Report of a rare case

A 30 year-old lady with a history of chronic sinusitis and rhinorrhea was referred to Rheumatology clinic with a two month history of epistaxis and necrotizing lesions of her nose.

ENT examination of the patient indicated a defect in her nasal septum.

CT of nasal soft tissues and para-nasal sinuses

- Mucosal thickening in the right maxillary sinus
- Defect in the anterior aspect of the nasal septum with an approximate diameter of 13 mm



Laboratory tests

WBC=14000/ μ l

ESR=56

CRP=3+

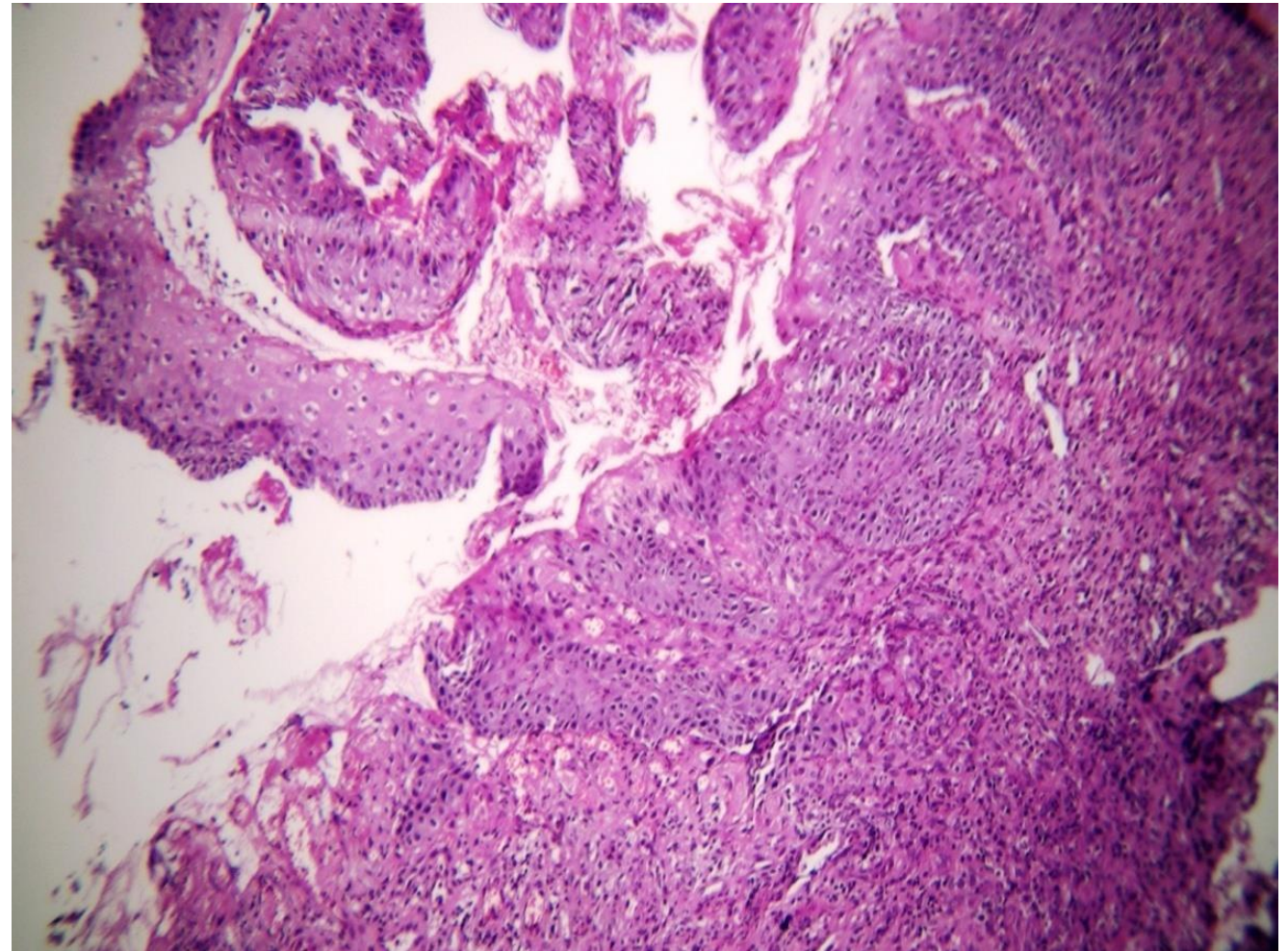
p-ANCA = 1/320

Anti PR3= +

C-ANCA ,ANA ,anti MPO = negative

Biopsy of the nasal septum

- Ulceration
- Acute inflammation
- Granulation tissue formation



Starting medical treatment with Dx of Limited GPA

Prednisolone (50 mg/day), Sulphamethoxazole and Trimethoprim was started, followed by the addition of Methotrexate (15 mg/week).

After 3 months, having noticed no improvement in the serological and clinical manifestation of the disease and considerable weight gain due to high dose steroid, Rituximab was administered for the patient. However, this treatment only slightly improved the clinical symptoms. At this point, the patient admitted continued nasal use of cocaine.

Cocaine-induced vasculitis

The use of cocaine continues to grow worldwide

Two distinct vasculitic syndromes have been described due to cocaine:

- Secondary to a direct vasoconstrictor effect of cocaine result in nasal mucosal necrosis, intense vasoconstriction leading to nasal septal perforation with a saddle-nose deformity.
- ANCA-associated vasculitis ,compounding the diagnostic confusion with WG.

Mechanism of ANCA positivity in CIMLD

- Nasal carriage of *S. aureus* .
- Levamisole-induced vasculitis. 70% of the available batches of cocaine are contaminated with the levamisole component .

Characterization of ANCA in patients with cocaine-induced midline destructive lesions (CIMLD)

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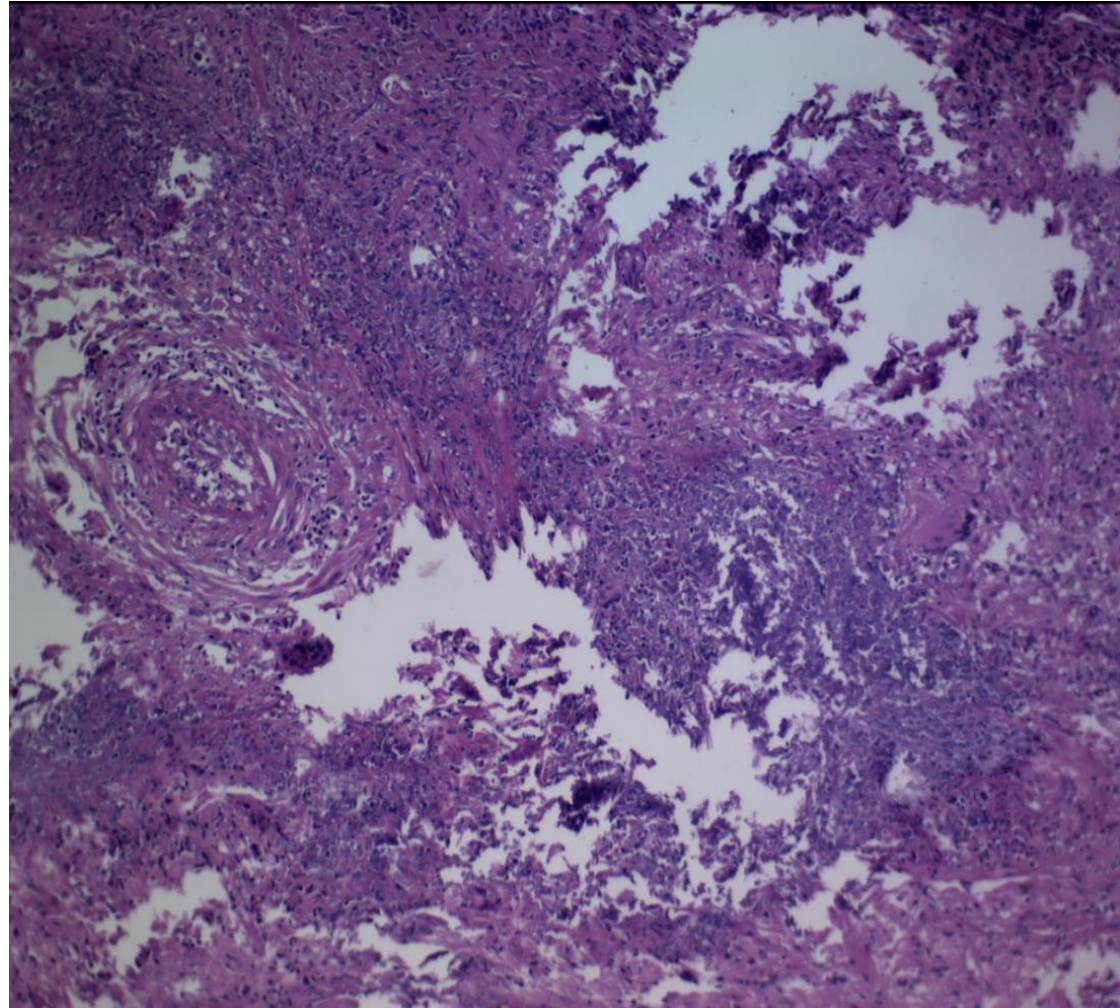
- PR3-ANCA has been reported in up to 50-84% of patients with CIMLD.
- The majority of GPA patients present with nasal signs and PR3 positivity lead to prompt diagnosis.
- The antigen for ANCA in patients with CIMDL is human neutrophil elastase. This antigen was not observed in patients with GPA.
- HNE and PR3 belong to the same family of serine proteases , The antibody reacting with both HNE and PR3 demonstrate a P-ANCA pattern on IIF.

Khasnis & Molloy. Int. J. Clin. Rheumatol. (2009) 4(5), 597–609

Peikert T, Finkielman JD, Hummel AM. Arthritis Rheum. 2008; 58: 1546–51

Histopathological difference of CIMLD and GPA

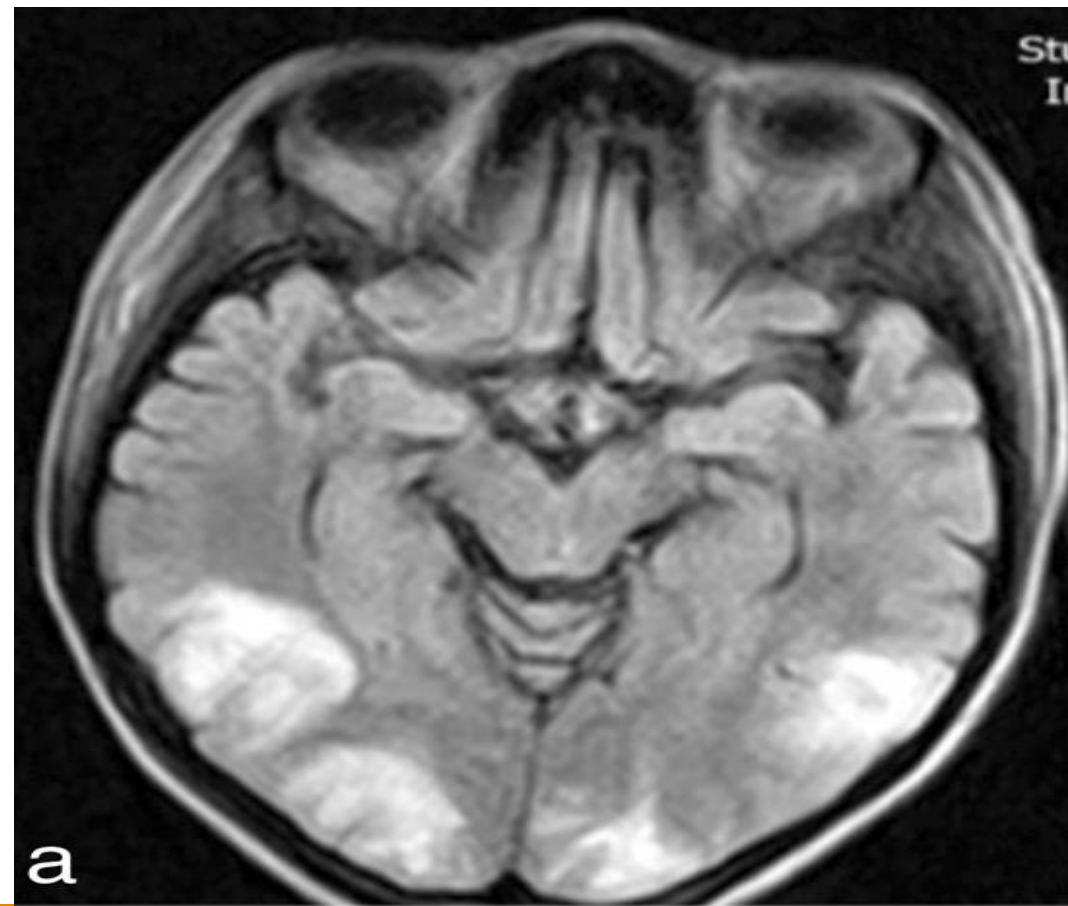
The specific characteristics of GPA, such as granulomas, giant multinucleated cells, are seldom seen.



Optimal therapy for CIMDL

It is challenging and immunosuppressive therapy including Corticosteroid, Cyclophosphamide, MMF and MTX could be considered in patients with extensive inflammatory disease or in patients who do not show an improvement following the cessation of cocaine use.

Posterior reversible encephalopathy is a syndrome highly associated with cytotoxic therapy. The syndrome typically presents with headache, visual abnormality, seizures and characteristic vasogenic edema on MRI.



Vasoconstrictor medications

Sustained use of vasoconstrictor medications, such as oxymetazoline can also result in similar injury to the nasal mucosa through ischemia and also mimic chronic sinusitis ,but will not typically be associated with positive ANCA testing.

Take home message

Awareness of ANCA positivity along with clinical manifestations similar to vasculitis in CIMDL could lead to a correct diagnosis of CIMDL, especially in regions with less popularity or availability of cocaine.