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Case Report: Vasculitis with Multiorgan Involvement-ANCA Associated Vasculitis (PR3+) Lung / Renal / Eye / Cutaneous Vasculitis

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- ORGAN THREATENING DISEASE{ACTIVE Urinary Sediment}
- Activity assessment: BVASV3—6 Pts
- Damage assessment-Vasculitic damage index-2

Highlights:

1) This article is an introduction and overview of the clinical presentation, lab evaluation and treatment of ANCA vasculitis.
2) Renal vasculitis is one of the most common of ANCA associated vasculitis and renal histology is the key predictor of the outcome [(1).]
3) Early diagnosis and intervention is very useful in these young patients to prevent mulyiorgan damage.

Abstract: ***Systemic vasculitides are heterogenous clinic pathologic disorder that share a common feature of vascular inflammation. The resulting disorder can vary depending on involvement of specific organs, calibre of vessels, underlying inflammatory process and individual host factor. The clinical description of vasculitis in a young patient with ANCA associated vasculitis is presented here.***

1. Introduction

Anti-neutrophil cytoplasmic autoantibody associated vasculitis is a group of autoimmune diseases characterized by inflammation and damage to small blood vessels

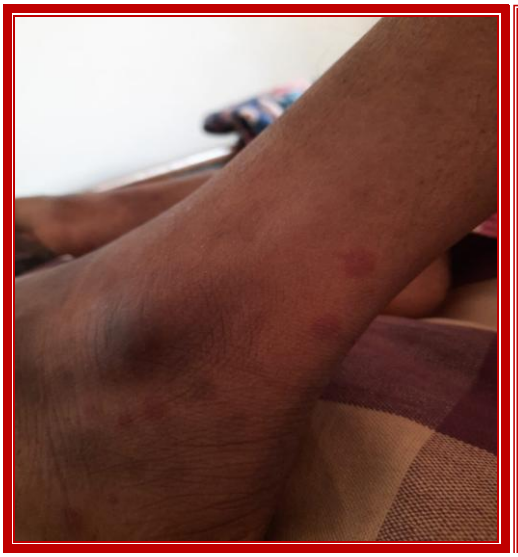
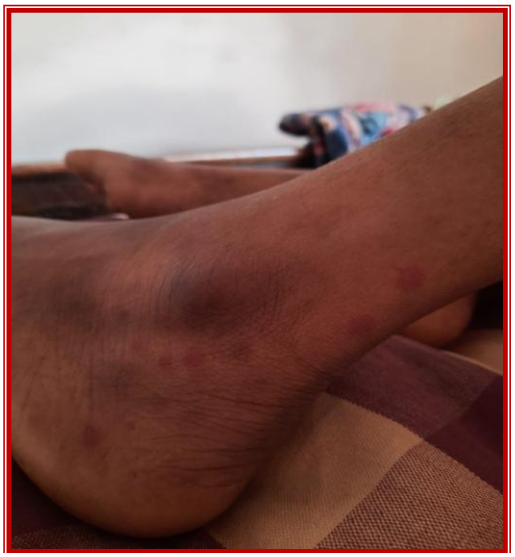
ANCAs are self reactive antibodies that bind to neutrophils and overly activate them. ANCA bind to white blood cells and activate the neutrophils which then bind to the cells lining the blood vessels, release toxic granules and further activate the immune system leading to vascular damage. These autoantibodies are present in high percentage of patients with Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome. They share the presence of ANCA and small vessel vasculitis, so collectively known as ANCA associated vasculitis.

Antibodies targeting the proteinase 3 (PR3} or myeloperoxidase MPO proteins on neutrophils are the most common ANCAs. Genetic and environmental factors cause this immune system malfunction. The strongest association of MHC a family of genesare associated with AAV.

Inhalation of silica or pesticides or alcohol and glues, prolonged use of cocaine, infection with certain virus and bacteria. Since small blood vessels are present throughout the body AAV causes many different symptoms.

2. Case Details

This is a 21 year old female, a college student came with complaints of recurrent attacks of fever, joint pain, abdominal distress, headache, blurring of vision, dyspnoea, orthopnoea, haematuria, photophobia, anorexia, weight loss, irregularity of mensural cycles, redness of eyes, arthritis involving the small and large joints for the past one year. she was evaluated and diagnosed to have ANCA [anti PR3]associated vasculitis involving eye, musculoskeletal system and lung. Investigations were suggestive of active urinary sediments. subnephrotic proteinuria and normal creatinine. secondary GN workup suggestive of Anti PR3> 200. She was referred for renal biopsy. She was started on Deflazocort 36 mg once a day.

On Examination
Patient conscious
Oriented
Answering questions relevantly
Co operative
Rt handed
Pallor+
Afebrile
No koilonychia
No lymphadenopathy
No thyroid swelling
No onychyolysisor pitting of nails
No red eye
CVS-S1normal, S2 normal, no additional sounds, no murmur
RS: Normal vesicular breath sounds, no added sounds
Abdomen-soft, non tender, no organomegaly
CNS-No focal neurologic deficit.
Not icteric
Not cyanosed
No clubbing
JVP normal
Pulse 90/min felt equally in all peripheral vessels
BP-120/80in the right upper limb supine
B/L Pitting pedal edema +
Palpable purpura++
Investigations
Haemoglobin-9. 8 gms
WBC count-9800
DC-71/22/14/3/0
MCV-91. 0fl
MCHC-53. 3 pg
MCH-24.5 pg
INR-0. 92
Platelet count-3, 15000
HCT-32.5
RBC-3.9
APTT-32.9 sec

Urinanalysis Routine
RBC-42/hpf WBC: 4-5/hpf EC: 4-5/hpf CRYSTAL-N nil, Yeast cell-nil PATH CAST: nil Mucus: nil Hyaline cast - present
Glucose: negative
Bilurubin: negative
Ketone: negative
Specific gravity: 1. 004
BLOOD: ++
PH: 7
PROTEIN: ++
UROBILINOGEN: NORMAL
NITRITE: negative
LEUCOCYTE: NIL
RBC: +
LFT
Bilurubin total: 0. 26 mg
Direct: 0. 10 mg
Protein total: 7. 8 gm
Albumin: 4. 0
AST: 13
ALT: 9
Alkaline phosphatase: 54
Creatinine0.8
ANA-Negative
CRP-15.1 mg
Urine 24 hrs protein: 1122mg
Urine volume 24 hrs: 1420 ml
Immunoglobulin G: 2008 mg/dl
Iron: 29 micro gm
TIBC: 233 micro gm
Serum electrolytes
Sodium -141
Potassium: 4. 1 mmol/l
Bicarbonate: 23 mmol/l
Anti PR3 &Anti MPO
Anti PR3: >200RU/ml
Anti MPO<2 RU/ml
Vitamin B12: 629pgm/ml
Folic acid: 69 ngm/ml
Hbs Ag: negative
ECHO normal
HIV: negative
HbsAg: negative

3. Discussion

Renal biopsy: Pauci immune PR3 ANCA associated focal necrotizing and diffuse sclerosing glomerulonephritis with 3/22glomeruli} circumferential cellular to fibrocellular crescentformation{2-cellular, 1-fibrocellular}

Diffuse acute tubular injurywith moderate tubulointerstitial scarring.

Comment-about 11/22 glomeruli appear globally sclerosed /scarred. Viable glomeruli are approximately.

Activity assessment: BVAS V3-6 points.

Damage assessment: Vasculitis damage index-2.

No hypocomplimentinemia.

Treatment: RITUXIMAB Infusion.

Tab. defcort 42 mg once daily for 2 weeks
30 mg once daily for 2 weeks
18 mg once daily for 2 weeks
15mg once daily for 2 weeks
12mg once daily for 2 weeks
9mg once daily for 2 weeks
Tab livogen once daily
Tab, sandocal two tablets in the night

Calcitriol granules60, 000 units once weekly for 6 weeks followed by once monthly for 6 months

Tab. septranDSonce daily
Tab. Losartan 25 mg once daily
Tab. pantop 40 mg once daily.

4. Conclusion

The diagnosis of ANCA associated vasculitis is made on the basis of clinical findings, biopsy of involved organs, and presence of ANCA. Testing for ANCA includes indirect immunofluorescence and antigen specific enzyme linked immunosorbent assay, which provides 99% sensitivity and Two major patterns of staining include cytoplasmic ANCA (C-ANCA), perinuclear ANCA (P-ANCA). C-ANCA is antibody to proteinase 3; P ANCA is antibody to myeloperoxidase. So, a more specific term proteinase 3 ANCA and myeloperoxidase ANCA are now in use. [4]this young patient who had been treated for a long time as a case of nonspecific arthritis is presented here to highlight the significance of this ANCA vasculitis and the timely management.

References

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