Clinical vignette: A case of peculiar midline necrotizing rhinosinusitis

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A Case of Peculiar Midline Necrotizing Rhinosinusitis

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INTRODUCTION:

- Granulomatosis with polyangiitis (GPA), ‘formerly known as Wegener’s Granulomatosis,’ is a potentially lethal necrotizing granulomatous vasculitis disease that can affect the lungs, sinuses, kidneys, gastrointestinal tract, and skin that requires prompt diagnosis and treatment, yet presenting signs and symptoms are highly variable in severity and often difficult to distinguish from those of a patient with non-vasculitic processes.
- Limited disease GPA presents with findings in the upper and lower respiratory tract without other organ involvement. Limited form patients are more likely to be younger at presentation, female, a greater likelihood of exacerbation of previous disease after a phase of remission, and a high prevalence of upper respiratory tract tissue destruction such as saddle-nose deformity. They are also less likely to be ANCA positive or have antibodies to proteinase 3 (PR3) or myeloperoxidase (MPO).
- Severe GPA (also known as generalized or systemic) disease presents with multisystem manifestations.
- Diagnostically, ANCA testing has become important as GPA is associated with anti-neutrophil cytoplasm antibodies (ANCA). most commonly cytoplasmic or C-ANCA, and PR3. Indeed, 96% of patients with severe GPA and 81% of patients with limited GPA develop positive ANCA titers. The difficulty is identifying those patients with limited form who have not yet developed positive ANCA titers.

CASE REPORT:

- A 46 yo female PMH of RA, pemphigus vulgaris on chronic steroids, persistent rhinosinusitis, and recent cocaine use was transferred from an outside hospital with 20 days of progressively worsening flu-like symptoms, nasal discharge and watery eyes with facial swelling and pain. Her symptoms had evolved over the previous week to include worsening, muffled hearing in Lt-Rt ear, and yellow-green drainage from a Lt infraorbital ulcer. Patient was started 2 weeks prior on an empiric course of antibiotics without improvement in symptoms. Physical exam revealed a mildly ill appearing woman with normal vital signs and a saddle nose deformity (first noted by the patient 1 year ago). Patient had swelling under both eyes, Rt Lt with a fulminating tract 1 cm below the Rt inferior lid margin. There was minimal surrounding erythema, fluctuance, and significant tenderness. Purulent fluid was readily expressed. Lt tympanic membrane was extremely injected and thickened with Weber lateralization to the left and air greater than bone conduction was noted bilaterally. Labs were significant for ANA >1:40, anti-DNA negative. ANCA negative. MPO and PR3 were mildly elevated at 117 and 144 (both normal ranges of 0-99). UA and UA/wBC were unremarkable. Skin biopsy demonstrated marked ulceration, granulation tissues and acute and chronic inflammation without evidence of vasculitis. Patient was discharged but returned before follow up appointment due to worsening symptoms. On exam patient had a small RLE hemorhagic, crusted plaque. Plaque biopsy demonstrated suppurative granulomatous inflammation with background vasculitis. Patient was started on GPA dosing steroids and cyclophosphamide/metha. She was discharged home and at follow-up 3 weeks later she reported decreased energy, improved hearing and decreased pain and swelling around her eyes. She continued follow up with ENT and rheumatology and continues on monthly cyclophosphamide infusions.

LIMITED DISEASE GPA

- 1. No red blood casts in the urine.
- 2. If hematuria is present the serum creatinine is <1.4 mg/dl and there must be no evidence of a rise in creatinine >25% above the patient’s baseline.
- 3. Pulmonary involvement must be circumscript, such that the room air PO2 is >70 mmHg or the room air O2 saturation by pulse oximetry is >92%.
- 4. Pulmonary hemorrhage may be treated as limited disease provided there is no evidence of progression of the process.
- 4. No disease may exist within any other critical organ such as the gastrointestinal tract, eyes, central nervous system that without the immediate institution of maximal therapy, such as pulse methylprednisolone and daily oral cyclophosphamide threatens the function of the critical organ and/or the patient’s life.

SEVERE DISEASE GPA

Any patient with GPA or Wegener’s Granulomatosis whose disease is unable to be classified as limited but, by definition, severe disease.

TREATMENT AND PROGNOSIS:

- Cyclophosphamide: PO 2 mg/kg/day, IV 15 mg/kg every 2 weeks for 6 weeks then every 3 weeks. Administer with mycophenolate and high-dose glucocorticoids such as prednisone or its equivalent 1mg/kg for 2-4 weeks depending on response then taper.
- Rituximab has been shown to be effective in inducing remission of GPA as cyclophosphamide in two randomized control trials. Can be used as second line drug.
- Mycophenolate would only be used as third line drug.
- Methotrexate may be used in selected patients with mild disease.
- Plasma exchange may be used in selected patients with severe disease.

DISCUSSION

- Prior to treatment advances in the 1970s when patients first were given cyclophosphamide and corticosteroids, GPA was a fatal disease.
- Delayed diagnosis can mean a delay in treatment which can translate into a persistence in deafness, systemic manifestations of the disease, and more aggressive therapy.
- Limited form GPA can present, as with our patient, as a midline necrotizing rhinosinusitis with saddle nose deformity and hearing loss. This often presents in younger women. ANCA negative without pulmonary or renal involvement.
- Diagnosis requires high suspicion of disease and often requires a prolonged period of continued testing and observation. It can be assisted by serology including C-ANCA, imaging, and biopsies showing granulomatous inflammation and vasculitis.
- Early diagnosis can reverse hearing loss and reduce disease progression. Hence due diligence in evaluation is paramount in diagnosing GPA.

EPIEDEMOLOGY:

- GPA is a rare disorder with a prevalence of 23.7-156.5 per million and incidence of 3-14.4 per million annually.
- Typical age of presentation is 40-60s, with a slight female predominance in limited form GPA, but suggestive male dominance in severe form.
- Studies suggest there is potentially a greater incidence in the Northern Hemisphere

REFERENCES:


Disclosure: 
- Photos obtained and presented with consent of patient.
- Please e-mail for a complete bibliography and image sources: