Clinical vignette: A case of peculiar midline necrotizing rhinosinusitis

Nathaniel DeFelice
Sara Marian S. Lucking
Peggy Beeley
A Case of Peculiar Midline Necrotizing Rhinosinusitis

Nathaniel DeFelice, MD, Sara Marian S. Lucking, MD, and Peggy Beeley, MD
Department of Internal Medicine, University of New Mexico School of Medicine, Albuquerque, NM.

INTRODUCTION:

◆ Granulomatosis with polyangiitis (GPA), ‘formerly known as Wegener’s Granulomatosis,’ is a potentially lethal necrotizing granulomatosus 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 organ distribution 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).

Case Report:

◆ A 46 yo female PMH of RA, pemphigus vulgaris on chronic steroids, persistent rhinosinusitis, and remote 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 Rt 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, R1-L1 with a telluric 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 (1:10, anti-DNA negative. ANCA negative. MPO and PR3 were mildly elevated at 117 and 144 (both normal ranges of 0-99). UA and BUN/VCP were unremarkable. Sinus 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 RLQ hemorraghic, 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 improved 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.

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 mesna 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.

References:


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