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Granulomatosis with Polyangiitis (Wegner's) : Often Mistaken for Tuberculosis

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Introduction

Granulomatosis with Polyangiitis (GPA) was first described by Dr. Friedreric Wegener in 1936 as necrotizing granulomatous disease (Mahmood et al., 2013). Formally known as Wegner's Granulomatosis(WG), this is a rare, complex disease that affects about three in 100,000 people in the United States (Sriskandarajah, Bansal, Yeoh, Bansal, 2012). Hammoudeh et al. (2011) defines GPA/WG as a triad of grannolomatous inflammation of the respiratory tract, necrotizing glomerulonephritis, and vasculitis of small vessels. Since GPA/WG is so rare, and the symptoms are vague, treatment is often delayed in patients. This delay in treatment results in high morbidity and mortality. According to Hammoudeh et al. (2011), "early institution of treatment with prednisone and cyclophosphamide can significantly reduce morbidity and mortality"(p.18). Early diagnoses and initiation of treatment is key in the survival of a patient.

Pathological Process

The exact cause of GPA has yet to be identified. Dinić et al. (2013) defines GPA/WG as "anti-neutrophil cytoplasmic antibodies (ANCAs)-associated systemic vasculitis of unknown etiology" (p. 887). According to Alam, Dastider, Ahmed, and Rabbani (2012) both cellular and humoral immunity are thought to be involved in the pathogenesis of wegener's granulomatosis. " The initial pathologic lesion is granuloma believed to be caused by cellular immune process. The strong association on C-ANCA with this disease suggests the role of humoral immunity" (Alam et al., 2014, p. 98). Huang et al. (2013) suggest, "The main pathological features of limited Wegeners' in the lung including liquefaction and coagulation necrosis lesions, as well as large number of eosinophils, and a small amount of lymphoplasmacytic infiltration, the destructive vasculitis can be found in the small pulmonary arties and veins"(p. 5). Some believe that environmental factors, such as dust inhalation, is related to the cause, but this is only seen in about 10% of patients with GPA (Cornarmond & Cacoub, 2014). Also, it has been suggested that infectious agents, such as Staphylococcus aureus in the nasal carriage, may play a role in triggering the disease, particularly though a mechanism of molecular mimicry (Cornarmond & Cacoub, 2014).

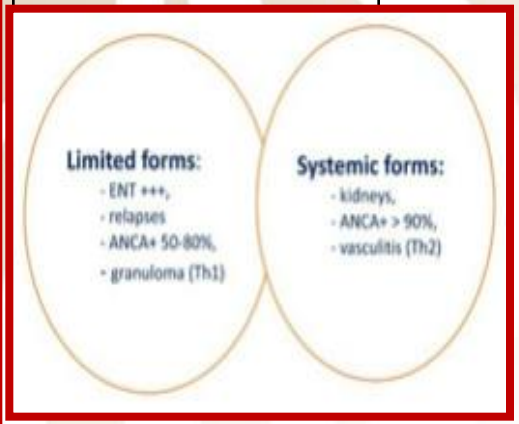


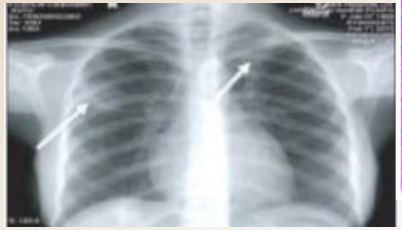
Diagram retrieved from Cornarmound & Cacoub, 2014, p. 1123

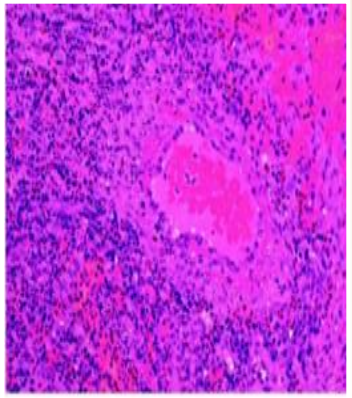
There are at least 2 different phenotypes that can be distinguished in GPA, localized, or diffuse. The less severe of the two is the localized form, which manifest through the ENT, and is limited to the upper respiratory tract (Cornarmond & Cacoub, 2014). This type is more recurrent and are more granulomatous with greater Th1 lympocyte polarization (Cornarmound & Cacoub, 2014).The more serious form, diffuse, manifests through the kidneys and/or lungs by alveolar hemorrhage (Cornarmound & Cacoub, 20141). With the diffuse from, relapse is less common, and there is greater Th2 lympocyte polarization (Cornarmound & Cacoub, 2014).

Case Study

A 37 year old women presents to the hospital with complaints of dyspnea, hemoptysis, joint pain, and sinusitis- type symptoms. She has not been eating very well due to a loss of appetite. Initial vital signs were:

- Heart rate:105 beats per minute
- Blood pressure: 102/68 mmhg
- Respiratory rate: 32 breathes per minute
- Oxygen saturation: 87% on room air
- Temperature: 102 degrees Fahrenheit
- Chest X-Ray showed pulmonary consolidation and infiltrates


Chest x-ray showing pulmonary consolidation and infiltrates (Alam et al., 2012, p. 100)


Histology slide showing multi-nucleate giant cells taken from patients nasal biopsy (Mahmood et al., 2013, p.412)

Initial thought was tuberculosis, but lab results showed three consecutive negative sputum samples for acid-fast bacilli, and a negative Montoux test.

Abnormal lab worked showed : hemoglobin 7.2 grams/dL, white blood cells 12,000, ESR 125 mm/hr, CRP 79.8 mg/L, serum creatine 3.9 mg/dl, BUN 64, C-ANA 39.8 U/ml (normal <2). Urinalyasis showed red cast cells.

On physical exam she was markedly pale, emaciated with a weight of 90 pounds, and a depressed nasal bridge. There was a large painful ulcer on her tongue. Nasal mucosa was full of crust and recent evidence of bleeding. Examination by ENT revealed a nasal septal perforation. A nasendoscopy was performed, which demonstrated lesions typical for GPA. Biopsies were taken showing fibrinous areas with multi -nucleate giant cells, which are consistent with GPA. Her knee and ankle joints were swollen and tender. All other labs and exams were normal. According to Alam, Dastider, Ahmed, and Rabbani (2012), the patient was diagnosed with Granulomatosis with Polyangiitis/ Wegeners Granulomatosis by the criteria met by the American College of Rheumatology classification of Wegeners Granulomatosis. According to the American College of Rheumatology, GPA is defined by the presence of at least 2 of the 4 following criteria:

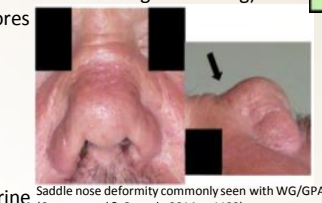
1) sinus involvement;
2) lung x-ray showing nodules, a fixed pulmonary infiltrate or cavities
3) urinary sediment with hematuria or red cast cells
4) histological granulomas within an artery or in the peripheral vascular artery or arteriole (Comarmond & Cacoub, 2014).

The patient meets three out of four classifications of this criterion. The patient was treated with Prednisolone and Cyclophosphamide and made a full recovery

Signs and Symptoms

- Sinonasl tract is most commonly affected region (Huchzermeyer et al., 2010)
- nasal membrane ulcerations and crusting
- saddle- nose deformity
- inflammation of the ear with hearing problems
- inflammation of the eye with sight problems
- cough (with or without the presence of blood)
- pleuritis (inflammation of the lining of the lung)
- rash and/or skin sores
- fever
- lethargy
- loss of appetite
- weight loss
- arthritic joint pain
- night sweats
- and blood in the urine

(Hammoudeh et al., 2011)


Saddle nose deformity commonly seen with WG/GPA. (Comarmond & Cacoub, 2014, p.1122)

Nursing Implications

Nursing professionals of all levels need to be aware of GPA/WG. Awareness of the disease is important because mortality rates go down with early treatment. With this disease being so rare and having only three in 100,000 people being affected, it is not on the top of the list for potential diagnoses. Also, having so many different signs and symptoms, and with those signs and symptoms mimicking other conditions, it is hard to diagnose. Educating staff members on the condition and teaching them the criteria made the American College of Rheumatology can lead to quicker diagnoses and treatment.

Conclusion

Granulomatosis with polyangiitis (GPA) / Wegener's Granulomatosis (WG) is a rare and often fatal form of systemic vasculitis but early diagnosis and treatment have a significant positive impact on outcome and prognosis. A delay in diagnosis of GPA increases the risk of living with dialysis or of death (Mahmood et al., 2013). The study of this case emphasizes the need for careful consideration and systemic analysis of patients presenting respiratory symptoms and signs suggestive of pulmonary TB, so that the diagnoses of GPA/WG is not delayed or missed.

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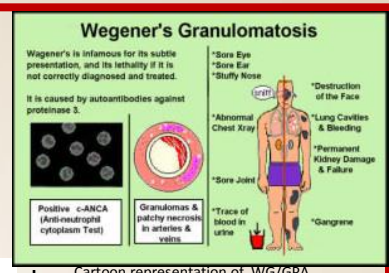

Cartoon representation of WG/GPA.
Picture retrieved from www.pedchrome.com

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