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Rare Disease - CANCA Associated Vasculitis (Wegener's Granulomatosis)

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Abstract: ***Wegener's granulomatosis (WG) is a rare systemic disease characterized by necrotizing granulomatous inflammation of the upper and lower respiratory tract, glomerulonephritis and vasculitis. It occurs as a limited form or generalized form and usually presents with nonspecific symptoms in its early stages, making the diagnosis of this disease more elusive. Strawberry gingivitis is the most common oral manifestation and is characteristic. Prompt recognition of this early manifestation is of utmost importance for the institution of early treatment, thereby avoiding serious complications.***

Keywords: Vasculitis, ANCA, Granulomatosis, Polyangiitis, Treatment

1. Introduction

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's Granulomatosis (WG) is a long- term systemic disorder that involves both granulomatosis and polyangiitis. It is a form of vasculitis (inflammation of blood vessels) that affects small- and medium-size vessels in many organs but most commonly affects the upper respiratory tract and the kidneys.

2. Clinical Examination

Temperature- 100.8'F
Pulse- 108/min
BP 102/74mmHg
SpO2-84% with room air.
Patient was tachypneic and restless

3. Case History

A 38yr old female presented with breathlessness (even on rest), continue high grade fever with chills. She had productive cough which was non-bloody. She had on and off episodes of similar history. No cardiac complains were present. She also complained of night sweats, Fatigue, lethargy, Loss of appetite, Weight loss.

4. Investigations

CBC: Total leucocyte count was gradually increasing from 11400 to 23000 to 27600.
ESR: was 67mm
CRP: was 79.8 mg/l
Electrolytes: normal.
Urine routine and blood cultures were inconclusive.
X-ray chest: showed subpleural inhomogeneous radiopacity in B/L Lung fields and also lung absces suggestive if infective Etiology.
CECT Chest: was suggestive of cryptogenic organizing pneumonia.
Sputum AFB was negative, but on culture, showed Acinetobacter haemolyticus growth.
2D Echo: Showed normal functioning heart.
C-ANCA: POSITIVE.

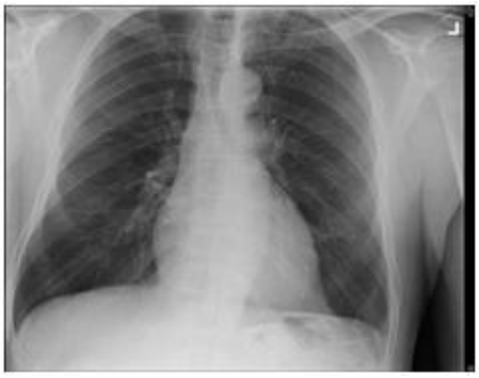

Image 1: X-RAY CHEST (PA)

Diagnosis
c-ANCA associated small blood Vasculitis (Wegener's granulomatosis) complicated with organizing pneumonia.

5. Treatment

Patient was given Oxygen inhalation and later on put on Bipap. She was given intravenous antibiotics. After the diagnosis of vasculitis, she was started with intravenous Cyclophosphamide and methylprednisolone. After that 3 infusions of Cyclophosphamide 15mg/kg every 2 weeks, later on every 3 weeks thereafter. Glucocorticoids were started as Prednisone 1mg/kg/day for 1st month and then tapered.

6. Discussion

The incidence is 10–20 cases per million per year.

Before modern treatments, the 2-year survival was under 10% and average survival five months. Death usually resulted from uremia or respiratory failure. The revised Five-factor score is associated with 5-year mortality from GPA and is based on the following criteria: age greater than 65 years, cardiac symptoms, gastrointestinal involvement, chronic kidney disease, and the absence of ears, nose, and throat symptoms.

With corticosteroids and cyclophosphamide, 5-year survival is over 80%. Long-term complications are common (86%), mainly chronic kidney failure, hearing loss, and deafness. The risk of relapse is increased in people with GPA who test positive for anti-PR3 ANCA antibodies and is higher than the relapse risk for microscopic polyangiitis.

7. Conclusion

GPA treatment depends on the severity of the disease. Severe disease is typically treated with a combination of immunosuppressive medications such as rituximab or cyclophosphamide and high- dose corticosteroids to induce remission and azathioprine, methotrexate, or rituximab to keep the disease in remission. Plasma exchange is also used in severe cases with damage to the lungs, kidneys, or intestines.

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