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Case Report

Granulomatosis with Polyangiitis (Wegener's Granulomatosis), Diagnosis, Clinical Approach, Initial Treatment and 1 Year Follow Up (Case Report)

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Abstract

Introduction**:* Wegener's granulomatosis (WG) is a rare long-term systemic disorder that involves the formation of granulomas and inflammation of blood vessels. ***Aim: of this study was to present a case of (WG) that was diagnosed with kidney biopsy while the lung infiltrates were not reachable. ***Materials and methods:*** 57-year-old male patient admitted for invasive diagnostics due to lung infiltrates. ***Results**:* The patient had positive C-ANCA on bloodwork, CT scan showing multiple bilateral lung infiltrates and an inconclusive result from a nasal mucosa biopsy. The lung infiltrates being difficult to reach for biopsy and having confirmed kidney involvement with high creatinine and albuminuria enabled us to do kidney biopsy. The biopsy showed a vasculitis with positive C-ANCA confirming (WG). A high dose of Methylprednisolone (500mg for 3day, then 250 for 3 day and finally 125mg for 3 days) A cyclophosphamide and maintenance dose steroids therapy was initiated by a consulting rheumatologist after discharge. A complete remission of the lung involvement was seen on a CT 2 months after. ***Discussion:*** The least invasive site for biopsy being the nasal mucosa is always the best site for biopsy. Having received an inconclusive result leaves the kidney and the lung as secondary options. In our case the kidney was considered due to a better possible yield of viable material ***Conclusion:*** Present kidney involvement is always a site for biopsy with a high yield for confirming (WG) despite the risk of bleeding complications who were absent in our case.

Keywords: Wegener's granulomatosis, lung infiltrates, kidney vasculitis.

1. INTRODUCTION

Granulomatosis with polyangiitis (GPA) is a potentially lethal systemic disorder that is characterized by necrotizing vasculitis of small arteries and veins. The respiratory system is most commonly affected in limited forms of the disease, however upper and lower respiratory system, systemic vasculitis, and necrotizing glomerulonephritis are the characteristic components of the disease triad [1]. The classic diagnostic criteria for GPA were based on the initial detailed clinical and pathologic findings as described by Godman and Churg in 1954 [2, 3]. Epidemiologically it involves of 8–10 per million depending on geographic location. The peak incidence is observed at 64–75 years of age [4]. Being a systemic vasculitis, it involves multiple organic systems. The classic triad consists of necrotizing granuloma of upper and lower respiratory system, systemic vasculitis, and necrotizing glomerulonephritis. The kidneys are usually spared in the limited form of GPA. Classic GPA can sometimes begin with limited organ involvement and then convert to a more generalized form with nose, lung and kidney being affected [5]. Prognostically without treatment life expectancy is usually 5 months, with a 1-year survival rate of less than 20% [6-8]. With adding systemic corticosteroids survival double to around 12 months [9]. Adding Cyclophosphamide can achieve complete remission and life-long survival [10]. Second line treatment involves Rituximab, plasma exchange and Avicopan (complement 5a receptor antagonist) [11, 12].

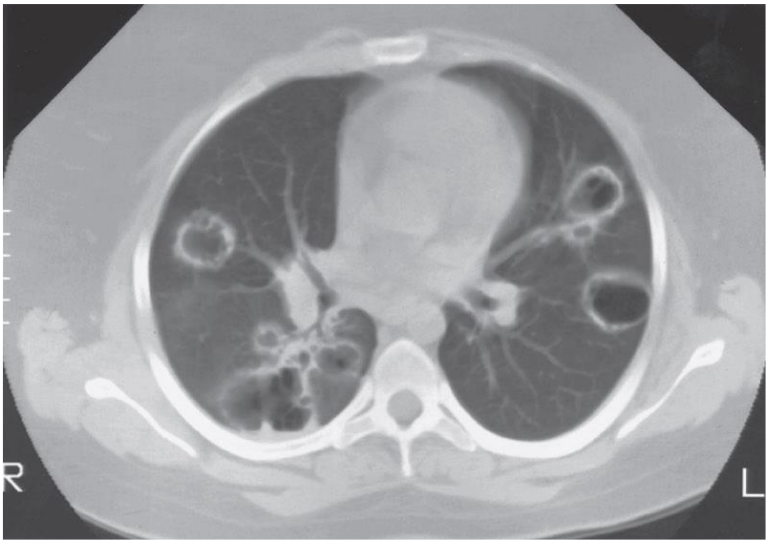

Fig. 1: CT scan of a patient with granulomatosis with polyangiitis

2. The aim of this study was to present a case of (WG) that was sent to our clinic for diagnostics due to lung involvement, but diagnosed with a kidney biopsy and later followed up and treated by a rheumatologist.

3. MATERIALS AND METHODS

A 57-year-old male patient, ad who underwent full clinical evaluation and invasive diagnostics in our clinic between 11.11.2024 and 26.11.2024 and the 1st year of militance therapy done by a rheumatologist. We will be performing a clinical analysis of the available data for both inpatient and outpatient setting done for diagnostics, initial treatment and maintenance treatment in the following year.

4. RESULTS

4.1 Background:

The patient had a history of fever reaching 40 degrees Celsius, nasal congestion, weight loss, elevated C-ANCA seen on blood work, and a CT scan showing multiple bilateral lung infiltrates with tendency for cavitating. In a local center he was treated with different antibiotics without improvement. Previous to the hospitalization the patient had a non-conclusive nasal mucosa biopsy and broncho biopsy. Even without a biopsy to confirm it, a suspicion for GPA was previously considered. From relative personal medical data, the patient was a non-frequent smoker, worked with automobile tires and was allergic to penicillin and metamizole. His father died from lung cancer. Before the hospitalization he did not report any previous chronic or acute illnesses and no chronic medication use.

4.2 Clinical presentation

The above-mentioned symptoms were present for 6 months. At presentation aside from an obstructive auscultatory finding on the chest no other visible objective abnormalities were noted.

4.3 Lab findings

During the hospitalization the patient had reduction in creatinine clearance, with serum creatinine reaching 174 umol/L at worst. Loss of albumin, confirmed with 24-hour proteinuria (albumin loss at 0.648 g/L on 1.8L of diuresis) suggestive and confirming kidney involvement. Inflammatory markers remained high with Leucocytes reaching 22,65 x10^9/L before the initial treatment with steroids. IgG was at 11.9 g/L; IgM at 0.51 g/L Complement C3 (S) 1.40 g/L Complement C4 (S) 0.36 g/L CRP at worst 75,5mg/L, C-ANCA again high at 84. Due to the potential need of invasive diagnostics, hemostatic findings were also examined and were in normal range.

4.4 Multidisciplinary approach:

Due to the multi system involvement a rheumatologist was consulted firstly, who asked above all for other lung conditions to be excluded, regardless of having the necessary criteria for the diagnosis. To biopsy the infiltrates in the lung was technically difficult due to their position and size and also with a high risk of getting a false negative biopsy or complications involving the biopsy process. Having kidney involvement, a nephrologist was consulted. He demanded the hemostatic blood workout and 24hour proteinuria. Having the required criteria kidney biopsy was scheduled, previously discussed with the nephrologist. The biopsy yielded 3 tissue samples and went without complications.

4.5 Histopathology:

confirmed an early kidney involving vasculitis (pauci-immune glomerulonephritis) with positive C-ANCA confirming (WG). Of the all visible 14 glomerulus 10 had thrombotic changes accompanied with leucocyte infiltrate. A collapse was noted on the basal membrane with neutrophil infiltrate. The tubules had ischemic changes, interstitial edema resulting in coagulating central necrosis. These changes as a whole

were most consistent with early kidney involvement due to vasculitis of the small capillary blood vessels and the preterminal arterioles confirming Wegener's granulomatosis and pauci immune complex glomerulonephritis (early stage).

4.6 Initial treatment

A high dose Methylprednisolone was given afterwards with decrease in the doses every 3 days as per protocol. A rheumatologist was consulted again, who initiated therapy with cyclophosphamide and maintenance dose steroids after discharge, with a consideration for a switch to rituximab in the future. Initially the doses of Methylprednisolone were 500mg for 3 days, then reducing to 250…120…60…40mg with a 3-day gap. After the initial treatment the patient was transferred to the rheumatology clinic for additional assessment and continuation of the treatment.

4.7 Aftermath

After 2 months of the initial treatment, with addition to cyclophosphamide on a monthly regimen together with methylprednisolone again at 500mg and continuation of the maintenance systemic steroids (firstly 10mg of prednisone orally), a control CT of the lungs was performed with no progression of the lung involvement. After the initial treatment the patient is now regularly followed up by a rheumatologist. The cyclophosphamide was firstly administered at 750mg once each month (with consideration to reduce it to once each 3 mounts), together with a 500mg of methylprednisolone. The maintenance therapy consists of prednisolone 10mg daily as when it was started Mycophenolate mofetil at 500mg 2x3 and pain medication when needed. Lastly reported from 08.07.2026 the patient is currently on 500mg of cyclophosphamide, Methylprednisolone at 80mg each month, a maintenance dose of prednisone 5mg daily. Kidney function is maintained above 59ml/min creatinine clearance and no lung complications are present. After the initiation of the treatment the patient developed oropharyngeal candidiasis which was treated with regular antimycotic regiment and 2 times an upper-mid airway infection that required short hospitalization. Both cases are considered complications of the initiated immunosuppressive therapy. Until now the only side effect reported regarding the steroid treatment is hypertension which can overlap with the kidney involvement and is being treated with angiotensin receptor blocker and beta blocker. No other complications involving the disease are until now noted.

5. DISCUSSION

This case is a presentation of a full diagnostic workout for (WG). It being a systematic disease requires a systematic and multidisciplinary approach both for diagnosis and the initial and maintenance treatment [13]. Having typical clinical presentation and increased c-ANCA with lung and kidney involvement is enough to apply a work diagnosis [14]. The diagnosis needs to be confirmed with a biopsy, however, in urgent situations like bleeding from the respiratory tract or increased risk of bleeding imaging, and positive PR3-ANCA blood tests strongly prove the disease [15, 16]. The safest sites are the skin, nasal mucosa and the upper respiratory tract due to the least risk of bleeding. The kidney and the lung are the other two sites with significantly increase bleeding risk and complications. However, the so called "safer" sites have a lower diagnostic yield (similar to our case) [14, 16, 17]. If the lung infiltrates or the mucosal edema is inconclusive or not available for viable biopsy due to any reason, a kidney biopsy can always help for diagnosing (WG), with C-ANCA being the second key criteria [17]. The initial treatment is done as soon as the biopsy is confirmed by the specialist who is doing it (in this case a pulmonologist) [18]. The maintenance treatment is done by a rheumatologist [17, 18].

6. CONCLUSION

Lung nodules with decrease in kidney function and positive C-ANCA complete the diagnostic criteria for (WG), biopsy of either the kidney or the lung is not regularly performed unless for exclusion of other conditions or to deal with inconclusiveness. The site for biopsy is usually the nasal or upper airway mucosa being the site of least complications which out weights the risk of false negative yield due to its repetitiveness and other potential sites.

7. Disclosure:

A preliminary version of this article was presented at the 2nd International Marcus Aurelius Scientific Research Congress April 2026 in Ankara Türkiye.

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