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Pneumocystis pneumonia in a patient with lupus nephritis

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ABSTRACT

Patients with lupus nephritis on immunosuppressive therapy are at an increased risk of opportunistic infections. Pneumocystis pneumonia infection though rare, does occur in such patients and is usually missed on commonly conducted investigations. Hence physicians must suspect it when clinical signs and symptoms do not improve in these patients despite being on treatment. being a diagnosed case of lupus nephritis (renal biopsy proven; immunofluorescence microscopy (IMF) showed full house positivity while serum serology revealing decreased levels of both serum C3 and C4, a strong positive ANA and rheumatoid factor). She had previously received three doses of cyclophosphamide. She was referred to us with the complains of pedal edema, fever and cough. Her examination revealed bilateral lung crepitations. Chest X-ray showed well defined shadows adjacent to the lung hilum with air bronchogram. In her bronchoalveolar lavage, a few pseudohyphae along with a positive test for Pneumocystis jiroveci pneumonia (PCP) was detected. Hence, the patient was treated for PCP pneumonia with sulfamethoxazole and trimethoprim. After completion of the course her condition had improved significantly and she was later on discharged.

Case Report

Implication for health policy/practice/research/medical education:
Patients with lupus nephritis on immunosuppressive therapy are at an increased risk of opportunistic infections. Pneumocystis pneumonia infection though rare, does occur in such patients and is usually missed on commonly conducted investigations. Hence physicians must suspect it when clinical signs and symptoms do not improve in these patients despite being on treatment.
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Introduction

Patients with lupus nephritis on immunosuppressive therapy are at an increased risk of opportunistic infections. Pneumocystis pneumonia infection though rare, does occur in such patients and is usually missed on commonly conducted investigations. Hence physicians must suspect it when clinical signs and symptoms do not improve in these patients despite being on treatment.

Case Presentation

A 20 year old female diagnosed case of lupus nephritis class IV and V, based upon her renal biopsy. Renal biopsy proven. Her immunofluorescence microscopy (IMF) showed full-house positivity (Figure 1) while serum serology revealed decreased levels of both serum C3 and C4, a strong positive ANA and a positive rheumatoid factor. She had previously received three doses of immunosuppressive therapy with cyclophosphamide. She presented this time with pedal edema along with undocumented fever on and off, cough with yellowish sputum since four days. She was currently on immunosuppressive therapy (prednisolone 15 mg/d) and for her current complaints she was admitted and started on IV broad spectrum antibiotics (Tazobactam).

On examination she was vitally stable, having bilateral crepitation on chest auscultation, while rest of her examination was unremarkable. Her initial laboratory reports showed a low hemoglobin of 9.2 g/dL along with a raised serum creatinine 2.3 mg/dL (Her previous serum creatinine was 0.7 mg/dL 2 months ago). Rest of her laboratory results were normal.

For these complains her chest X-ray showed well defined shadows adjacent to her right and left lung hilum with air bronchogram, more developed on the right side (Figure 2). Since she was producing sputum, her sputum sample was

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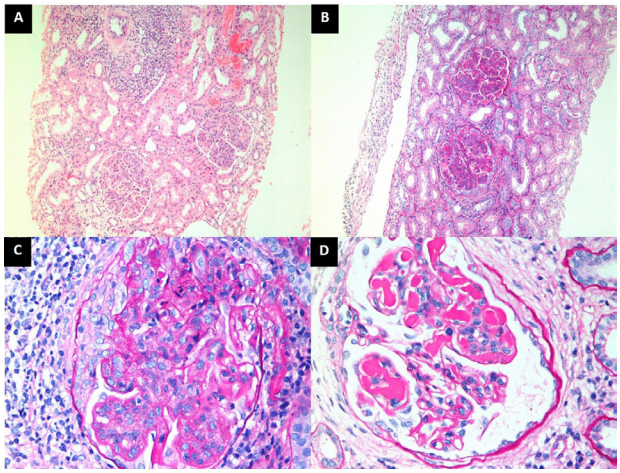

Figure 1. Histopathological features on renal biopsy. A. Low-power view showing grossly abnormal glomeruli and patchy dense interstitial inflammation. (H&E, ×100). B. Another area of biopsy showing markedly hypercellular glomeruli with prominent lobulation (PAS, ×100). C. High-power view showing mesangiocapillary pattern of morphology with a small crescent at 9'O clock position. (PAS, ×400). D. This glomerulus shows numerous hyaline thrombi and wire loop lesions, characteristically found in lupus nephritis (PAS, ×400).

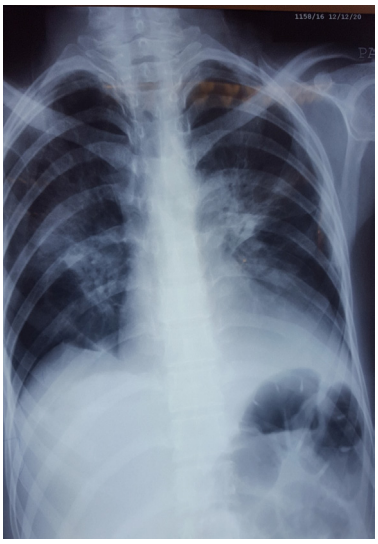

Figure 2. Chest X-ray, PA view showing well defined shadows adjacent to her right and left lung hilum with air bronchogram, more developed on the right side

sent for detailed report, culture and sensitivity and acid-fast bacilli (AFB) smear but all tests were negative.

Her viral markers were also negative. Her blood and urine cultures showed no growth while her Urine D/R was bland.

Since her chest symptoms did not improve and she was not maintaining her saturation, she was kept on intermittent oxygen support of 2 L. Patient's antibiotics were escalated to imipenem. Later on her bronchoalveolar lavage (BAL) pseudohyphae along with testing positive for pneumocystis pneumonia (PCP). Then, the patient was managed for PCP pneumonia and started on sulfamethoxazole and trimethoprim. After completion of the course her condition had improved significantly and she was later on discharged.

Discussion

Systemic lupus erythematosus (SLE) is an autoimmune disease characterized by multisystem involvement and the production of various autoantibodies (1). SLE may involve any organ, the most common ones being the skin, joints, kidneys, heart, lungs and CNS (2). Involvement of kidneys named lupus nephritis (LN) affects 20% to 75% of patients in the first 10 years of illness (3).

Development and progression of LN is regarded as a multistep inflammatory process which is incited by anti-DNA and anti-nucleosome antibodies, resulting in a self-maintaining inflammatory loop with spreading of glomerular inflammation. Pro-inflammatory antibodies such as C1q are involved (4). Early manifestations of LN include hematuria and proteinuria revealed by urinalysis (5). The diagnosis is however confirmed by biopsy results where the disease is staged as per different histopathological presentations (5). Intravenous cyclophosphamide combined with corticosteroids is the first line therapy for lupus nephritis (6). The resultant immunosuppression increases the risk of opportunistic infections.

However unlike most opportunistic infections, pneumocystis pneumonia (PCP) has a low prevalence of 0.45% in immunocompromised SLE patients (7). Lower lymphocyte count right before the onset of PCP and the higher dose of steroid therapy along with depressed CD4+ counts are considered possible risk factors for the development of PCP infection in SLE patients (8). This elevated risk is attributed to an altered immune response in the patients and adverse reactions to immunosuppressive therapy.

PCP typically presents with fever, non-productive cough, shortness of breath, weight loss and night sweats. Clinical diagnosis of PCP is made by using PCR of sputum, serum beta-D glucan test, lung biopsy and bronchoalveolar lavage (9,10). Non-invasive and reliable serum indicator of PCP and serum beta-D-glucan hold significant importance in the definitive diagnosis, especially when severe respiratory distress impedes bronchoalveolar lavage (BAL) (11).

PCP infection rarely contributes to a high mortality rate (60%) and can be a life threatening condition (12). According to two studies, all SLE patients who developed PCP, correlated with poor clinical outcome and massive kidney involvement (12,13). Severity and treatment of PCP may be different in different ethnicities. In the Asian population it may be more progressive and requires more effective treatment (14). Trimethoprim-sulfamethoxazole is reported to be an important constituent in the treatment regimen of PCP where the antibiotic shows a favorable

response (9).

Conclusion

Our study highlights the importance of diagnosis and treatment of all infections, in particular, in patients with lupus nephritis who are currently on immunosuppressive therapy. Physicians catering to such patients need to keep in mind common infections as well as rare entities like PCP, since the spectrum of disease infecting these patients can be very vast.

Conflicts of interest

There were no points of conflicts.

Authors' contribution

ZM managed the patient and wrote the manuscript. ShA managed the patient. SaA managed the patient. AH and HJ wrote the manuscript. MM was responsible for the histopathological diagnosis and images. All authors read and signed the final paper.

Ethical considerations

Ethical issues (including plagiarism, data fabrication, double publication) have been completely observed by the authors. Written informed consent was obtained from the patient for publication this case report.

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