Fulminant Presentation of Systemic Lupus Erythematous in A Four Year Old Girl

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ABSTRACT

Systemic Lupus Erythematous (SLE) is a chronic systemic disease which can involve multiple organs such as skin, kidney, and brain. Lung is another organ that can be affected. Pulmonary complications including pleuritis, pneumonitis, infectious pneumonia, pulmonary hemorrhage, pulmonary hypertension and pneumothorax have been reported in patients with SLE. Pulmonary involvement is relatively frequent in adult patients. It has infrequently been reported in children with SLE. However, pulmonary manifestations may be an initial and/or life threatening complication of SLE [1].

We report here a 4 year old Saudi girl who presented with prolonged fever and respiratory manifestations due to acute lupus pneumonitis associated with pulmonary Tuberculosis (TB) then developed recurrent pulmonary hemorrhage and acute respiratory failure.

Keywords: Plasmapheresis, Pulmonary hemorrhage, Acute lupus pneumonitis, Juvenile systemic lupus erythematous, Tuberculosis

1. INTRODUCTION

Acute lupus pneumonitis is an uncommon but life threatening complication of SLE with an estimated incidence of 10% in juvenile systemic lupus erythematous (JSLE) [2].

The diagnosis is made by exclusion of infection, acute pulmonary edema, hemorrhage and infarction. Patients are extremely ill, febrile, tachypnic and hypoxic.

The chest radiograph reveals ill-defined bilateral patchy air space consolidation in the periphery [3,4].

Pulmonary hemorrhage (PH) is a rare and life-threatening manifestation of SLE, occurring in 1–5% of adult cohorts with mortality rates ranging from 70%–90%. The majority of PH reported in SLE refers to the adult population, and there is a paucity of data in pediatrics [5]. It is an uncommon occurrence in children; although the exact prevalence is not known, it occurs in less than 5 percent of patients [6], and is extremely rare as an early manifestation of this condition [7]. Additionally, outcomes were favorable with initiation of aggressive, multi-modal Immune-suppressive therapy with no recurrences [5].

Moreover, Tuberculosis infection in the setting of SLE is one of the most difficult conditions to manage as clinical features and laboratory investigations can coexist in both diseases and the presentation may be variable. The prevalence of the disease in the community is also an important deciding factor [8].

2. CASE REPORT

4 year old Saudi girl presented with prolonged fever weight loss cough and skin rash. The rash involving malar area and aggravated by sun exposure. Physical examination revealed malar rash sparing the nasolabial folds and generalized unremarkable lymphadenopathy, heart, chest and abdomen within normal limits. Chest x ray showed bilateral infiltrates so, she was admitted for further workup and was started on IV antibiotics. She was then diagnosed as SLE based on clinical features and lab. Criteria (high anti dsDNA, ANA, anticardiolipin IgM, Pancytopenia, hemolytic anemia, low C3 & C4), and was started on hydroxychloroquine.

| Table1: Results of blood work at time of admission |
|---------------------------------|------------------|------------------|------------------|
| Value                          | Normal Range     | Value            |
| WBC                            | 5.0-15.0         | 3.42 x10³/µl     |
| Hb                             | 11.5-13.5        | 6.3 g/dl         |
| PLT                            | 200-450          | 326 x10³/µl      |
| ANC                            | 1.5-8.0          | 1.81             |
| Retics                         | 0.5-2.5          | 2.3 %            |
| ESR antidsDNA                  | 1.0-30           | 44 mm            |
| ANA                             | 0.9-1.8          | +ve              |
| C3                             | > 15 –ve         | 0.209 g/l        |
| C4                             | 0.1-0.4          | 0.056 g/l        |
| Anticardiolipin IgM            | < 15 –ve         | 25.5 unit        |
| DCT                            | -ve              | +ve              |
| Urine                          | -ve              | RBC +++           |
| Quantiferon TB gold            | Indeterminate    | -ve              |

WBC white blood cells, Hb hemoglobin, PLT platlets, Retic reticulocyte, ANC absolute neutrophil count, ANA antinuclear antibody, DCT direct coom’s test, RBC red blood cell, Prot. protein, ESR erythrocyte sedimentation rate, C3 complement component 3, C4 complement component 4.

One week after admission to hospital, the patient continued to have fever, and she developed tachypnea, respiratory distress and hypoxemia. Chest radiograph showed new infiltration despite she was on IV antibiotics. CT chest was done showed severe bilateral alveolar infiltrations with ground glass pattern together with bilateral hilar adenopathy (Images 1 and 2).

Acute lupus Pneumonitis was diagnosed based on the clinical picture and the radiologic findings. The possibility of pulmonary Tuberculosis (TB) could not be ruled out. Quantiferon TB gold test came indeterminate, thus, based on CT chest findings and being in an area of high prevalence of the disease she was started on antituberculous medications. Her general condition deteriorated, so patient shifted to pediatric intensive care unit where she showed fulminant course, she developed respiratory failure, so
she was connected to mechanical ventilator. She was mechanically ventilated with high settings for 4 weeks. Initially in PICU she received methylprednisolone 30mg/kg. Two days later, she developed severe pulmonary hemorrhage with significant drop in hemoglobin, managed with blood transfusion and methyl prednisolone (IV) was extended to 5 days, she showed partial response. Three days later she developed another attack of severe pulmonary hemorrhage with desaturations and hemodynamic instability. There was significant drop in hemoglobin and platelets count complicated by picture of autoimmune blood hemolysis, she received multiple PRBC and platelet transfusions and started on intravenous immune-globulins (IVIG). She showed partial response, but again few days later she developed recurrent pulmonary hemorrhage with persistent picture of autoimmune blood hemolysis, complicated by pneumothorax and candida sepsis treated with chest tube insertion & antifungals respectively. Plasmapheresis started, she needed 13 cycles, in addition to methyl prednisolone and IVIG. After completed 13 cycles of plasmapheresis she showed good response, pulmonary hemorrhage stopped, her vital signs became stable, and her fever subsided, repeated blood culture came negative. She received the first dose of Cyclophosphamide, then she was extubated gradually after four weeks of high settings of mechanical ventilation. After that patient shifted to pediatrics ward, her physical examination was normal, and she was referred to higher center, to be seen by pediatric rheumatologist. She was seen in pediatric clinic regularly, she finished course of Cyclophosphamide (six doses), she didn’t require any admission and her renal function remained normal (till last visit to pediatric clinic).

3. DISCUSSION

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease that can involve any organ system with a wide range of disease manifestations, and can lead to significant morbidity and even mortality. The disease may present with acute, severe, and life-threatening symptoms, or present as a fluctuating and chronic process that impacts several systems and increases the risk of malignancy [9]. Because of its protean manifestations and presentations, lupus must be considered in the differential diagnoses of many conditions, including fever of unknown origin, arthralgia, anemia, Idiopathic thrombocytopenic, new-onset kidney disease, psychosis, and fatigue. Excellent clinical evaluation from history, physical examination and laboratory finding is very crucial, because early diagnosis and appropriate early treatment have improved the prognosis of this fatal disease. In this case the patient referred to pediatric clinic because of fever of unknown origin. History alone gave us important information together with photosensitivity, weight loss and mild cough. Physical examination showed skin rash involving malar area, sparing the nasolabial folds. Laboratory results supported the diagnostic criteria of SLE. Hospital course of SLE is highly unpredictable. In this case, after few days of admission patient started to have respiratory distress, CXR showed bilateral infiltrations, so antibiotics started as bacterial pneumonia, the patient did not respond to antibiotics, CT chest done -to exclude other cause- which showed severe bilateral alveolar infiltrations with ground glass pattern together with bilateral hilar adenopathy. The impression of radiologist was tuberculosis (TB).

With all these findings we thought of two differential diagnoses:

- Acute lupus Pneumonitis based on the clinical picture and the radiologic findings.
- Pulmonary Tuberculosis (TB) which could not be ruled out as Quantiferon TB gold test came indeterminate.

Patient managed in both lines because of:

- Being in an area of high prevalence of TB
- Patient needed to start on methyl prednisolone, and
- High possibility of starting immunosuppressive medications.

Although the patient was managed in both lines the patient deteriorated rapidly, and developed severe recurrent pulmonary hemorrhage, then the patient developed autoimmune blood hemolysis, severe thrombocytopenia and sepsis. All modalities of treatment were tried to stop the pulmonary hemorrhage. We noticed that plasmapheresis had the main role in stopping pulmonary hemorrhage.

4. CONCLUSION

SLE has protean manifestations and unpredictable course. Aggressive appropriate management is very crucial. Pulmonary hemorrhage is rare but serious pulmonary complications of SLE. Because pulmonary
hemorrhage can be a life-threatening complication, any child with SLE who experiences acute shortness of breath should be evaluated promptly for pulmonary hemorrhage. If pulmonary hemorrhage is diagnosed, treatment should not be delayed. Moreover, Tuberculosis infection in the setting of SLE is one of the most difficult conditions to manage as clinical features and laboratory investigations can coexist in both diseases and the presentation may be variable, in addition it can affect the outcome of the disease.

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