Dear Editor,

Idiopathic pulmonary hemosiderosis is characterized with episodes of hemoptysis, diffuse reticuloendothelial infiltrations in chest X-rays and iron deficiency anemia secondary to hemoptysis [1,2]. Although it is generally seen in childhood and adolescence, it can rarely be seen in adults as well [3]. Recurrent intraalveolar bleeding comprises the main pathophysiology of the disease. Reticulonodular infiltrations are seen in the chest X-rays due to these bleedings, and iron deficiency anemia ensues secondary to massive hemoptysis. The etiopathogenesis of the disease is not completely understood [4]. We report here a patient presenting with complaints of fever and cough who had diffuse infiltrations in his lungs whose symptoms and findings regressed with high dose methylprednisolone treatment.

A three-year-old male patient was brought to the hospital with complaints of fever and weakness for two days. Medical history of the patient revealed that he was followed up for a diagnosis of pulmonary hemosiderosis, he had received prednisolone treatment for two years in a dose of 2 mg/kg/day, and that he was taking azathioprine 1 mg/kg/day for the last eight months. Celiac antibodies were negative and had erythrocyte suspensions many times. The patient had a healthy female sibling and his parents were not relatives. Physical examination revealed paleness, hyperemia and hypertrophia in the tonsils; other system examinations were normal.

His hemoglobin and erythrocyte levels were 6.0 gr/dl and 3.13x106 /ul, white blood cell count and thrombocyte count were 7.4x109/l and 341x109/l, and mean corpuscular volume (MCV) and red cell distribution width (RDW) were 65.9 (fl) and 27.4 gr/dl, respectively. There were 45% neutrophils, 45% lymphocytes, and 10% monocytes in the peripheral blood smear. Hypochromia, microcytosis and anisocytosis were present. Total and direct bilirubin levels were 0.81 mg/dl and 0.33 mg/dl, respectively. Direct and indirect Coombs tests as well as cold Coombs test were negative. His haptoglobin level was <26 pg/dl, iron level 16 mg/dl, iron binding capacity 322 g/dl, ferritin 52 ng/ml and his corrected reticulocytes were 6%. Marked reticuloendothelial infiltrations were present in the chest X-ray of the patient (Figure 1a). An allergy skin test was performed with negative results for milk allergy. Ampicillin-sulbactam treatment was started, erythrocyte suspension in a dose of 10 cc/kg was transfused and azathioprine treatment was stopped. His control hemoglobin value was 8.6 g/dl. He was administered methylprednisolone for pulmonary hemosiderosis in a dose of 30 mg/kg for three days, following with 20 mg/kg for four days, and 10 mg/kg for one week. Control hemoglobin level was identified to be 9.7 g/dl. No decrease was seen in the control hemoglobin levels of the patient who was continued to receive iron treatment (6 mg/kg/day). The reticuloendothelial infiltration on the chest X-ray resolved completely (Figure 1b). The patient was followed up on methylprednisolone in a dose of 5 mg/kg/day for one week and then in a dose of 1 mg/kg daily. The patient was not brought to follow-up visits and then was brought to the outpatient clinic after two months with the complaint of cough and weakness. Apparently, he was not taking the steroid medications and the lesions appeared in his chest X-ray again. His hemoglobin level was found to be 6.7 g/dl and he was started high dose methylprednisolone treatment again.

Hemoptysis attacks and reticuloendothelial infiltrations due to recurrent alveolar hemorrhage are seen in idiopathic pulmonary hemosiderosis. Iron deficiency is typical. Clinical picture may be silent; however, it may be seen with signs of deep anemia and hypoxemia developing in days [1,2].

Systemic glucocorticosteroids are generally used for treatment. Other immunosuppressive agents such as azathioprine, cyclophosphamide, and hydroxychloroquine have also been used alone or in combination with oral corticosteroids [1]. Oral
prednisolone in a dose of 2 mg/kg daily could be started if the amount of hemoptysis is not much and if the patient tolerates oral glucocorticoid treatment [5]. Cases with marked improvement in lung symptoms and anemia with prednisolone treatment have been reported [6]. However, high dose methylprednisolone treatment was not administered in these patients up to now. We administered methylprednisolone in our patient in a dose of 30 mg/kg/day for 3 days, followed with 20 mg/kg/day for 4 days and 10 mg/kg/day for 1 week. The findings in the chest X-ray of the patient were markedly improved after the steroid treatment. No decrease was seen in the hemoglobin levels at the same time.

Before the high dose treatment with methylprednisolone azathioprine and prednisolone treatment in a dose of 2 mg/kg/day were applied in our patient with no marked improvement in the symptoms and signs. We suggest that high dose methylprednisolone treatment creates an efficacious response in a short time period of treatment.

References


