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Poster presentation

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Efficay of plasmapheresis in a case of severe pulmonary hemorrhage in pediatric Systemic Lupus Erythematosus

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We report the case of a 13 year old girl with SLE initially treated with hydroxycloroquine for a mainly mucocutaneous involvement.

Four months from onset, she presented renal involvement and an acute hemoptysis and she was admitted to our pediatric department. She also presented fever and abnormal laboratory findings (high serum creatinine level, severe proteinuria, Hb of 6.4 g/dl, high titre specific autoantibodies, low C3 and C4 levels).

Kidney biopsy underlined stage 4a lupic nephropathy; chest X-ray and CT scan showed diffused alveolar involvement, as for pulmonary hemorrhage. Cerebral SPECT showed abnormalities consistent with neurolupus. The patient underwent blood transfusion, and was treated with high doses of metilprednidsolone plus cyclophosphamide. Due to refractory anemia and worsening of respiratory symptoms (dispnoea with low peripheral oxygen saturation), she was additionally treated with 3 courses of Rituximab, and two further blood transfusions, with clinical stabilization but no major amelioration. After one week, severe clinical worsening with important dyspnoea, cutaneous pallor, poor general condition, low peripheral oxygen saturation (78%) was present: Hb level was 4 g/dl, chest X-ray showed worsened alveolar involvement. The patient was treated with 3 courses of plasmapheresis and two blood transfusions, with immediate response of the respiratory symptoms, recovery from anemia and marked improvement of the chest X-ray.

Pulmonary hemorrhage is a severe complication in SLE associated with high mortality. It is described in 5–6% of patients, presenting with anemia, hemoptysis, respiratory symptoms and characteristic changes in chest imaging.

We suggest plasmapheresis as an effective treatment to be considered promptly when suspecting this life-threatening complication.