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Thrombotic microangiopathy in juvenile dermatomyositis

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Objectives and study

A 4- year-old boy was admitted to our hospital because of progressive proximal muscular weakness of 4 weeks duration, heliotrope rash, erythematous skin lesions in the trunk and limbs and febrile fever. The muscle enzymes were elevated: creatine kinase 7,056 IU/L (0–170), aldolase 40.3 U/L (0–8), aspartate transaminase 601 U/L (10–37), alanine transaminase 289 U/L (10–37). The laboratory test revealed a hemoglobin level of 86 g/dL, a platelet count of 100,000/mm3, an erythrocyte sedimentation rate of 30 mm/h . A peripheral blood smear revealed schistocytes (2%). Urine analysis revealed large number of red blood cells and proteinuria 1.06 grams / liter. The level of total serum bilirubin was 37 μmol / l (normal up to 17 μmol / l). Serum creatinine was 200 μmol/l. Direct and indirect Coombs' reaction was negative. Prothrombin and thrombin time and D-dimers were within normal limits.The serologic tests for the antinuclear antibody, antineutrophil cytoplasmic antibody, antibodies to DNA, cardiolipin antibodies, antibodies to β2-glycoproteins were negative. Lupus anticoagulant was not detected. The electromyographic findings were consistent with the inflammatory myopathy. A renal biopsy contained 43 glomeruli and was evaluated by light and immunofluorescence microscopies. Diffuse fibrin-platelet thrombi were revealed in glomerular capillaries. Small arteries was narrowed by the swelling of the endothelium. The expression of the fibrin + +. There were no immune deposits at any location.

Methods

The patient received three pulses of methylprednisolone 30mg /kg daily, followed by oral prednisolone 60 mg/day. Alternate-day plasmapheresis with fresh frozen plasma for 1 week and intravenous cyclophosphamide treatment (750 mg/m^2/month) were started.

Results

The treatment resulted in slow improvement in his muscle weakness within 4 weeks along with markedly decreased muscle enzymes and normalization of renal function. Prednisolone was changed to 1 mg/day of oral prednisolone, which was later decreased to 0.5 mg/day followed by transition to long-term oral methotrexate 10 mg / m2/week (after 6 monthly pulses of cyclophosphamide).

Conclusions

The described clinical case demonstrates the varied nature of renal pathology at juvenile dermatomyositis. The kidney damage appeared to have been very severe. But under the well-timed and adequate therapy it underwent to a back development in a great degree.

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