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CORRESPONDENCE

Tapering Doses of Methylprednisolone Pulse in the Treatment of Macrophage Activation Syndrome Associated with Systemic Juvenile Idiopathic Arthritis

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To the Editor: Macrophage activation syndrome (MAS), a complication of systemic juvenile idiopathic arthritis (sJIA), is characterized by cytokine storm and life-threatening multiorgan dysfunction [1, 2]. Although corticosteroids are the backbone of the therapy, anti-IL-1 agent (anakinra) is now being increasingly used. However, the cost and availability is a limiting factor in India. In such situations, tapering dose regimen of intravenous methylprednisolone (IVMP) may be considered. We report two such cases.

Case 1: A 6-y-old girl was diagnosed to have sJIA with MAS (fever, arthritis, lymphadenopathy, ascites, hepatosplenomegaly, pancytopenia, transaminitis, hypofibrinogenemia, elevated ferritin, and hemophagocytosis on bone marrow). She was initiated on IVMP pulse therapy (30 mg/kg/d). Cyclosporine A was initiated on day 4. Because of partial response, IVMP pulse therapy was continued for 10 d and etoposide initiated on day 8. She showed some improvement and IVMP pulse therapy was gradually tapered (20 mg/kg/d, 15 mg/kg/d, and 10 mg/kg/d for 3 d each). After 19 d, oral prednisolone was initiated (2 mg/kg/d). It took 2 wk for anakinra to be imported and the first dose (2.5 mg/kg subcutaneously) was administered on day 27.

Case 2: A 6-y-old boy was diagnosed to have sJIA with MAS (fever, evanescent rash, arthritis, anemia, thrombocytopenia, leukocytosis, and high serum ferritin). He was initiated on IVMP (30 mg/kg/d) for 5 d followed by oral prednisolone (2 mg/kg/d). After an initial recovery, his fever recurred and laboratory investigations showed relapse of MAS. He was reinitiated on IVMP (30 mg/kg/d) for 5 d followed by a slowly tapering regimen (20 mg/kg/d for 3 d, 15 mg/kg/d for 3 d, 10 mg/kg/d for 3 d). Oral cyclosporine A was initiated on day 3.

Even though the drug side-effect profile favors use of anakinra in MAS, it may be suggested to use tapering IVMP protocol in countries where anakinra is not being marketed or cost is a significant constraint.

Declarations

Conflict of Interest None.

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