Simultaneous Presentation of Disseminated Intravascular Coagulation and Hemophagocytic Syndrom in a Patient with Adult-Onset Still's Disease

Sir,

Adult onset Still's disease (AOSD) is a rare systemic inflammatory disease with a prevalence less than 1 case per 100,000 people. Generally, AOSD has a good prognosis, but some complications such as Disseminated Intravascular Coagulation (DIC) and Hemophagocytic Syndrome (HPS) become severe and life-threatening. Here we report a young woman who experienced DIC and HPS secondary to AOSD. Overlapping complications developed rapidly and the patient died of respiratory failure at last. We recommend that more attention should be paid to the complications of AOSD and its triggering factors.

A 33-year-old woman was admitted to our hospital with one week of high fever, skin rashes and arthralgia. Although she had taken antibiotics for one week (prescribed at a local clinic), there was no improvement. White blood cell count was 13 x 10^9/L. Serology for Anti-Nuclear Antibodies (ANA), Rheumatoid Factor (RF), Epstein-Barr Virus (EBV) and Cytomegalovirus (CMV) was negative. Blood, bone marrow and sputum cultures were all negative. No evidence of hematological malignancies was founded except for some activated phagocytic macrophages in bone marrow aspirate. Lymphoma was excluded by gene re-arrangements test of TCR and IgH. During hospitalization, the patient experienced intermittent high fever (> 39°C), arthralgia and skin rashes. Steroid therapy was effective in controlling temperature; the skin rashes faded followed by lowering of body temperature. According to the Yamaguchi criteria,1 the patient was diagnosed as AOSD (Figure 1). On the 12th day after admission, signs of infection such as high fever, cough, yellow phlegm and chest distress appeared. Laboratory data showed high white blood cell count, CD64 index and C-reactive protein (CRP). Chest CT suggested pulmonary inflammatory disease. Two days later, DIC was suspected and confirmed by the abnormal functions of coagulation and fibrinolysis. She was given appropriate antibiotics against infection and low-molecular-weight heparin, Argatroban along with transfusion for DIC (Figure 1). When the platelet count dropped, intravenous heparin injection was stopped and platelets administrated instead. Though the coagulation function improved, hemoptysis aggravated progressively which was caused by pulmonary embolism and DIC. On the 17th day, cytopenia developed rapidly accompanied by hyperferritinemia, hypertriglyceridemia and severe liver dysfunction. Together with increased activated macrophages previously observed, DIC secondary to AOSD was diagnosed. Meanwhile, immune dysfunction was noticed by the low ratio of CD4+/CD8+ T-cells (0.18 vs. normal range: 1.41 ± 0.31) associated with high level of CD4+CD25+CD127low regulatory T-cells (Tregs) (13.2% vs. normal range: 5.23 ± 0.94%). Though high dose immunosuppressive therapy was administrated, respiratory symptoms including dyspnea and hemoptysis aggravated and the patient died of respiratory failure.

Besides DIC, HPS is another potentially fatal complication. Excessive activation of macrophages and impaired immune response cause "cytokine storm" which aggravates inflammatory responses and results in multiorgan dysfunction.2,3 Though the cause of secondary HPS remains unclear, both abnormal by activated CD8+ T-cells and high level of Tregs strongly point towards this process.4-6 Unbalanced homeostasis caused by abnormal inflammatory response and autoimmune dysfunction are common features shared by AOSD and HPS.

We postulate that infection as a triggering factor initiated and aggregated this pathological process. The cascade amplification effect caused by excessive cytokine and impaired immune response make the disease more troublesome and difficult to control. We wish to emphasize that more attention should be paid to the triggering factors such as infection at any stage in this disease.
REFERENCES


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