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Autoimmune Lymphoproliferative Syndrome; A Case Report

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Abstract:

Autoimmune lymphoproliferative syndrome is a disorder of lymphoid system regulation characterized by chronic splenomegaly, lymphadenopathy and autoimmune phenomena especially immune-mediated cytopenias. The hallmark of the disease is the presence in peripheral blood and lymphoid tissue of increased numbers of a normally rare T lymphocyte subset, usually referred to as “double-negative” T cells. Here the authors report a 16-year-old boy when he was first hospitalized for diffuse petechiae, purpura and epistaxis at 9 years of age.One year later,he was readmitted for high fever and recurring cytopenia. On examination several enlarged, nontender lymph nodes involving cervical and submandibular areas and a huge spleen were detected.Lymph node biopsy was performed two times. According to flowcytometry of peripheral blood and immunophenotyping of lymph node tissues which revealed increased numbers of CD3+CD4-CD8-T lymphocytes, autoimmune lymphoproliferative syndrome was suggested for him. Autoimmune lymphoproliferative syndrome should be considered in differential diagnosis of any patient with unexplained Coomb’s positive cytopenias, hypergammaglobulinemia, generalized lymphadenopathy and splenomegaly. The confirmation of the diagnosis should be based upon genetic analysis and detection of the affected genes involved in fas pathway.

Keywords:

Autoimmunity ، Splenomegaly

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