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Cutaneous infarcts and antineutrophil cytoplasm antibodies positive vasculitis in angioimmunoblastic T-cell lymphoma: A rare presentation

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Angioimmunoblastic T-cell lymphoma (AITL) is one of the rare sub-types of peripheral T-cell lymphoma, comprising 2-5% of all non-Hodgkin lymphomas. AITL is associated with many systemic features like fever, rash or arthritis. These systemic features may precede the appearance of other features of lymphoma. The disease may present with autoimmune phenomena, such as the presence of circulating immune complexes, cold agglutinins, hemolytic anemia and rheumatoid factor and anti-smooth muscle antibodies. In approximately half of the patients, polyclonal hypergammaglobulinemia is present. Vasculitis is not common but comprises antineutrophil cytoplasm antibodies (ANCA) negative vasculitis. A 65 year old male presented with complaints of a short febrile illness associated with weight loss, dry cough, generalized weakness, pruritus and paresthesias of bilateral lower limb. He was admitted to medical ward and after 12 hours patient developed dyspnea and blackish discoloration of digits. Clinical examination revealed scattered polyphonic wheeze over bilateral chest. A working diagnosis of secondary vasculitis was made and was started on methylprednisolone pulse in view of impending digital gangrene. He responded to the treatment with no further progression of digital ischemia. Autoimmune workup revealed pANCA strongly positive by ELISA; however ANA, DCT/ICT was negative. Chest and abdomen showed hepatosplenomegaly with hilar and pretracheal lymphadenopathy. Serum protein electrophoresis revealed monoclonal gammopathy, IgG lamda restricted. A lymph node biopsy of left axilla was done and HPE was suggestive of angioimmunoblastic T-cell lymphoma. He was started on chemotherapy CHOP and has shown response. AITL is one of the uncommon but aggressive neoplasms which have a varied clinical presentation including constitutional symptoms, lymphadenopathy, organomegaly, autoimmune phenomena especially hemolytic anemia and thrombocytopenia and polyclonal hypergammaglobulinemia. This case emphasizes the atypical presentation of a rather rare disease and the need for high index of suspicion, which will help in initiating chemotherapy well in time and to prolong median survival.

Biography

Abhish Mohan has his interest in hematolymphoid malignancies and has passion to improve the diagnostic capabilities of resource limited countries which will benefit a large number of people. His other areas of interest include Military Medicine with special emphasis in high altitude medicine.

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