EDITORIAL

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Angioimmunoblastic T-cell lymphoma: Case report of a diagnostic challenge presented as a lymphoproliferative syndrome

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Abstract:

Brazilian woman patient, 51-year-old, born within the State of Bahia, rural worker, married, Catholic, turned into living in São Paulo for 2 months. She become admitted to the Emergency Department at Santa Casa of São Paulo in October/2014 complaining of stomach pain, nausea, vomiting, lymphadenomegaly, fever, night time sweats and weight loss (10kg) that had begun about three months ago. She smoked 1 p.c. of cigarettes according to day for 36 years; however, she denied any beyond medical records or agrotoxic exposure. The entire blood count (CBC) showed anemia, eosinophilia, and thrombocytopenia. All the serologies for infectious diseases have been negative, besides for IgM EBV, that changed into fine. Abdominal ultrasound showed homogeneous hepatosplenomegaly, periportal lymphadenomegaly, a easy cyst in the proper kidney and small quantity of ascites. CT experiment of the chest showed small nodules inside the lungs, a small quantity of pericardial effusion, increased range of lymph nodes in mediastinal, tracheal and intracranial regions, multiplied size of lymph nodes in hilar region bilaterally as nicely as within the chains of the diaphragm, clavicles, and inside the axillaries chains. Myelogram ruled out Leishmaniasis. The bone marrow biopsy become most effective hypercellular, showing hyperplasia of the three myeloid types. Lastly, the cervical lymph node biopsy changed into done with immunophenotyping: CD45 diffusely advantageous; CD3 fine inside the small and medium cells; CD20 nice in immunoblasts; CD4 wonderful in maximum of the lymphocytes; T-cellular lymphoma with angioimmunoblastic features.

INTRODUCTION:

Angioimmunoblastic T-cell Lymphoma (AITL) is a unprecedented malignancy that simplest represents 2% of all non-Hodgkin lymphomas, but this is the maximum not unusual subtype of all the peripheral T-cell Lymphomas (15-20%). Most sufferers are elderly, and the median patients' age is round 60-years-old. The maximum prominent signs and symptoms at the time of presentation are generalized lymphadenopathy, hepatosplenomegaly, fever and weight loss. The scientific presentation may additionally mimic inflammatory, autoimmune and infectious diseases, or even other lymphoid neoplasms. Most of the sufferers usually have simultaneous extranodal disorder in spleen, liver, skin, lungs and bone marrow. The definite diagnosis is generally tricky, and can handiest be done with the aid of lymph node biopsy.

CASE REPORT:

Brazilian affected person PPMS, female, 51-year-old, born within the State of Bahia, rural worker, married, catholic, was dwelling in São Paulo for 2 months. She became admitted to the Emergency Department at Santa Casa of São Paulo in October/2014 complaining of belly pain, nausea, vomiting, lymphadenomegaly, fever, night sweats and weight loss (10kg) that had started about 3 months before. She smoked 1 pack-per-day for 36 years, but she denied any beyond medical history or agrotoxic exposure. The complete blood count (CBC) showed anemia, eosinophilia and thrombocytopenia. All the serologies for infectious diseases have been negative, besides for IgM EBV, that became advantageous. Abdominal ultrasound confirmed homogeneous hepatosplenomegaly, periportal lymphadenomegaly, easy cyst in the proper kidney and small amount of ascites. CT experiment of the chest showed small nodules inside the lungs, small quantity of pericardial effusion, extended wide variety of lymph nodes in mediastinal, tracheal and infracarinal regions, extended length of lymph nodes in hilar region bilaterally as nicely as in the chains of diaphragm, clavicles, and within the axillaries chains. Myelogram dominated out Leishmaniasis. The bone marrow biopsy turned into simplest hypercellular, displaying hyperplasia of the 3 myeloid types. Lastly, the cervical lymph node biopsy changed into done with immunophenotyping: CD45 diffusely tremendous; CD3 superb inside the small and medium cells; CD20 nice in immunoblasts; CD4 fine in most of the lymphocytes – T-cell lymphoma with angioimmunoblastic features.

DISCUSSION:

The case suggested above was excellent because it did now not have bone marrow involvement, what to begin with drove us away from the ideal prognosis. The AITL is thought to regularly involve bone marrow, however the histologic and immunophenotypic features at this web site are poorly defined. Therefore, we conclude that the lymph node biopsy is constantly indispensable for elucidating cases like this, which might be marked with the aid of a lymphadenopathy that persists for extra than four weeks or that still have signs suggesting malignancy (eg, fast increase in lymph node size, fever, night time sweats or weight loss).

The pathogenesis of AITL stays unclear. In some cases the disease is preceded with the aid of an allergic reaction, contamination or drug exposure. A number one monoclonal and polyclonal T-mobile proliferation secondary to Epstein-Barr virus infection additionally been suggested.

At the time of diagnosis, nearly all patients gift with advanced-stage disease (levels III-IV). In 70% of sufferers manifestations encompass B signs and symptoms, and 79% have splenomegaly. Cutaneous lesions vary broadly and can be encountered in approximately half of of cases, supplying as a nonspecific rash, generally macules and papules, and less generally purpura, urticaria, nodules or petechiae.

The differential diagnoses consist of infections, inflammatory or autoimmune illnesses and other lymphoid neoplasms. A diagnosis can most effective be executed by lymph node biopsy.

The molecular profile of the neoplastic T cells in AITL includes CXCL13, bcl-6 and CD40L, proven in gene expression studies [6]. 95% of AITL instances had accelerated expression of at least one of the following markers: SLAM-associated protein, programmed death-1 and c-Maf

The analysis of AITL could be very challenging, given the lack of medical and histological diagnostic criteria. In one of the biggest collection of instances of peripheral T-cellular lymphomas, agreement on prognosis by expert hematopathologists was simplest 81%. Cutaneous and histological findings may be nonspecific; therefore, scientific history, signs and complementary research are very important. Our patients have been recently identified with autoimmune hemolytic anemia, hypereosinophilic syndrome and arthritis; these, in conjunction with her signs and symptoms and except skin and lymph node biopsies, led us to the best analysis.

Keywords: Autoimmune Hemolytic Anemia, Biopsies, Arthritis, Hypereosinophilic Syndrome, T-Cell Lymphoma, Thrombocytopenia

Biography:

Leticia Alves Antunes has completed her Medical Degree from Federal University of Sao Carlos and is a former Resident in Internal Medicine from Santa Casa de Sao Paulo, Brazil. She is now applying for Medical Residency in the United States.