

**Introduction:** T-cell prolymphocytic leukemia (T-PLL) is a rare T-cell leukemia, accounting for approximately 2% of lymphocytic leukemia cases in adults over 30 years of age. It has an aggressive clinical course and poor prognosis, in addition to a low response to conventional chemotherapy and a short survival time. This disease is characterized by post-thymic lymphocyte proliferation with small to regular size. The cells have weak CD3 expression and CD4 expression on the cell membrane occurs in over half of the cases. Co-expression of CD4 and CD8 occurs in about 25% of cases. A smaller proportion of cases have cells with only CD8 expression on the cell membrane. The main feature is severe leukosis in peripheral blood, but involvement of bone marrow, lymph nodes, liver, spleen and skin are not uncommon. **Case report:** Previously healthy 57-year-old woman presenting with abdominal pain and distension, weight loss, asthenia, and decreased general condition for 2 months. Physical examination revealed splenomegaly (19 cm RCE) and hepatomegaly (18 cm RCD), generalized lymphadenopathies of no more than 2 cm. CBC with Hb 7.5 g/dL, large leukocytosis  $472 \times 10^9/L$  with 92% prolymphocytic cells and platelet count was  $31 \times 10^9/L$ . Multi-parametric flow cytometric analysis (MFC) in peripheral blood showed a mature T-cell lineage strongly positive for CD3, CD8, CD7 and CD26, negative for CD4, CD45 RA and RO and NK marker isoforms (CD56, CD57, CD94) but positive for cytoplasmic TCL1. TCR rearrangement evaluation showed clonally CD8 T-cells positive for  $V\beta$  chain 8. Conventional cytogenetic examination showed complex karyotype with a rare derived chromosome: der(2)t(1;2)(q21;q37), plus iso chromosome 8q, deletion 11q22, structural changes involving chromosomes 14, 13, 17 and several markers. FISH confirmed additional signals in MYC (8q), IGH (14q) and TP53 (17p) genes. The patient treated with chemotherapy (Fluorouracil, Cyclophosphamide and Mitoxantrone), with no response. Since she maintained lymphocytosis, lymphadenomegaly and hepatosplenomegaly, she was treated with Alemtuzumab (anti-CD52) with a plan to do 12 cycles, but showed progression within 3 months, requiring discontinuation of the medication. The patient was referred for palliative care in May 2021. **Conclusion:** We report the case of a patient with T prolymphocytic leukemia presenting with an elevated peripheral blood white blood cell count, complex karyotype with a rare derived chromosome, der(2)t(1;2)(q21;q37), aberrant immunophenotype and progressive disease despite treatment with alemtuzumab.

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#### CEREBELLAR DIFFUSE LARGE B CELL LYMPHOMA IN IMMUNOCOMPETENT FEMALE PATIENT

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**Background:** Diffuse large B cell lymphoma (DLBL) is the most common type of lymphoma worldwide, representing about 25% of all non-Hodgkin lymphomas. Among these lymphomas, primary DLBL of the central nervous system (CNS) is a rare category listed at 2016 revision of the World Health Organization classification of lymphoid neoplasms. First line treatment is based on methotrexate (MTX), with studies showing complete remission rates of 30 to 60% in patients who receive high-dose MTX, alone or in combination with other agents. The rarity of primary CNS lymphomas makes randomized trials difficult to do, but regimens usually include MTX plus one of the following drugs: cytarabine, procarbazine, temozolomide or rituximab in CD20 positive tumors. **Aims:** to present a case of primary CNS lymphoma presenting as multiple lesions in the cerebellum. **Case presentation:** We present a case of a 57-year-old female patient, without comorbidities or HIV seropositivity, diagnosed with diffuse large B cell lymphoma with cerebellar origin. During a period of 4 months the patient presented progressive difficulties to accomplish everyday activities quickly evolved, and were accompanied by aphasia, dysarthria, lack of motor coordination, nausea and vomiting. Her handwriting was unintelligible. A brain MRI showed enhanced leptomeningeal area associated with middle cerebellar peduncle lesions on both sides. Biopsy confirmed the diagnosis of lymphoma with immunohistochemistry showing positivity for CD20 and 40% Ki67. Bone marrow biopsy was normal and the serology for HIV, Epstein-Barr virus and hepatitis B and C viruses were negative. She was treated high dose induction immunochemotherapy with methotrexate (four doses of 3.5 g/m<sup>2</sup>), cytarabine (two cycles of 2 g/m<sup>2</sup>, twice a day for 2 days) and rituximab (four doses of 375 mg/m<sup>2</sup>). After the first cycle, the patient developed foreign accent syndrome (FAS) changing from her Brazilian accent to a Portuguese accent (within the same language). Despite FAS, other neurological symptoms ameliorated and the patient can once again write with her normal handwriting, have conversations and read. She was referred to complementary whole-brain radiotherapy. **Discussion:** Literature on cerebellar primary central nervous system lymphoma is scarce.<sup>1</sup> A review of PCNSL reported only 9% of tumors with cerebellar involvement. The addition of high-dose cytarabine to high-dose methotrexate seems to be associated with improved response in patients with primary CNS lymphoma. However, cerebellar toxicity is a dose-dependent adverse effect of cytarabine and a concern in cases with previous cerebellar syndrome. FAS may be related to involvement of 'linguistic cerebellum'<sup>2</sup> but apparently not associated with cytarabine. We showed that this combination was possible, without major neurologic events. **Conclusion:** Early diagnosis and treatment is important for the successful management of this aggressive and uncommon cerebellar tumor. Symptomatic improvement was fast after MTX initiation and the addition of cytarabine is feasible.

#### REFERENCES

1. Ghannam M, Mansour S, Jumah F, Berry B, Beard A. Cerebellar large B-cell lymphoma: a case report. *J Med Case Rep.* 2018;12:341.