T-cell lymphoblastic lymphoma

- Cord compression due to atypical T-cell lymphoma from paraspinal soft tissue: Report of a case
- Intracranial residual lesions following early intensification in a patient with T-cell acute lymphoblastic leukemia: a case report
- An Anti-CD7 Antibody-Drug Conjugate Target Showing Potent Antitumor Activity for T-Lymphoblastic Leukemia (T-ALL)
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T-cell Lymphoma

Epidemiology

Represents 1%-2% of all Non-Hodgkin's lymphomas.

More common in adolescents and young adults, predominantly males.

Rare in older adults, making this case notable.

Pathogenesis

Originates from immature T-cell precursors. Often associated with genetic mutations involving NOTCH1, CDKN2A, and other genes regulating T-cell development. Aberrant T-cell signaling pathways contribute to uncontrolled proliferation.

Clinical Presentation

Commonly involves the anterior mediastinum, causing mass effect symptoms like dyspnea, chest pain, or superior vena cava syndrome. May also affect lymph nodes, skin, or extranodal sites like the CNS, bones, and paraspinal soft tissue (as in the case described). Neurological symptoms such as paraparesis, sensory loss, or sphincter dysfunction can occur in cases with spinal cord compression.

Diagnosis

Histopathology: Sheets of small to medium-sized lymphoblasts with high mitotic activity.

Immunohistochemistry: Positive for T-cell markers (CD3, CD7) and markers of immaturity (TdT, CD1a, or CD99). Negative for B-cell markers (CD20, CD79a). Molecular Studies: May reveal genetic abnormalities like translocations or mutations. Imaging: MRI or CT scans often show bulky masses in typical locations (e.g., mediastinum or spine).

Prognosis and Recurrence

Aggressive behavior with a high likelihood of relapse, particularly in the CNS or epidural space. Early relapse, as described in the case report, is common due to the disease's infiltrative nature and resistance to therapy.

Treatment

Multimodal Approach: Combines intensive chemotherapy (regimens similar to those used in T-ALL), CNS prophylaxis (intrathecal chemotherapy or radiotherapy), and sometimes hematopoietic stem cell transplantation in refractory or relapsed cases. Surgical Intervention: Reserved for cases with acute complications like spinal cord compression or when a biopsy is needed for diagnosis. Outcomes depend on the stage at diagnosis, CNS involvement, and response to therapy.

Case reports

A 57-years-old man with less than 24 hours of onset symptoms of paraparesis, lower limb hypoesthesia and sphincter dysfunction who was operated due to dorsal tumor with epidural component which caused severe cord compression. Pathological analysis concluded atypical T-cell lymphoblastic lymphoma, Ther case was particularly aggressive and atypical due to its origin in paraspinal soft tissue. Despite specific treatment, the patient presented an early epidural relapse, frequent in this lymphoma subtype [1)].

1)
Ortega Rodriguez AA, Valbuena Dussan SN, Caro Cardera JL, de Manuel-Rimbau Muñoz J. Cord compression due to atypical T-cell lymphoma from paraspinal soft tissue: report of a case. Neurocirugia (Astur : Engl Ed). 2024 Nov 16:S2529-8496(24)00070-4. doi: 10.1016/j.neucie.2024.11.004. Epub ahead of print. PMID: 39557339.