and anti-LG1. Antibody-positive AE represents a distinct subgroup of encephalopathies characterized by autoimmune responses against various antigens in the brain parenchyma[1,2]. Due to clinical, imaging, and laboratory similarities with infectious and other autoimmune encephalitides, AE remains a diagnostic challenge. Patients typically present with subacute memory and cognitive decline over days to weeks. Encephalopathic syndromes may include behavioral changes, psychosis, seizures, and coma, reflecting a broad neuropsychiatric spectrum[3]. In addition to supportive and antiepileptic therapies, early initiation of immunosuppressive treatment is essential. First-line immunosuppressive therapies in AE include corticosteroids, intravenous immunoglobulin, and plasma exchange[4,5]. For patients unresponsive to first-line treatments, second-line therapies include anti-CD20 monoclonal antibodies (rituximab and ofatumumab), mycophenolate mofetil, cyclophosphamide, and azathioprine. In refractory cases, third-line therapies such as daratumumab, bortezomib, obinutuzumab, tocilizumab, anakinra, tofacitinib, and intrathecal methotrexate may be considered[5]. Several studies have demonstrated that the use of ofatumumab, a second-generation CD20 monoclonal antibody, results in reduced antibody titers and significant clinical improvement in AE[6,7]. **Case report:** A 22-year-old female patient was admitted to an ICU in Kosovo in June 2025 with impaired consciousness and was intubated. Her Glasgow Coma Scale (GCS) score was 3. She was transferred to our hospital's intensive care unit on June 19, 2025, with a presumptive diagnosis of autoimmune encephalitis. A brain MRI performed externally on June 16 showed symmetric signal abnormalities in the bilateral basal ganglia. A lumbar puncture was performed upon admission, and CSF was sent for paraneoplastic and autoimmune antibody panels. Immunosuppressive therapy with 1000 mg methylprednisolone was initiated. For focal motor seizure control, levetiracetam (3000 mg/day) and valproic acid (2000 mg/day) were added. Plasma exchange (1/1) was started every other day beginning on June 19. Tracheal aspirate cultures grew carbapenem-resistant Acinetobacter baumannii. The patient was started on colistin, to which the isolate was sensitive. Despite therapy, focal motor seizures persisted. A brain MRI performed on June 26 showed widespread encephalitic signal changes in the left fronto-temporal and parieto-occipital regions, as well as in the right temporal region. CSF analysis revealed negative results for both paraneoplastic and autoimmune antibody panels. Given the lack of response to 1000 mg methylprednisolone and seven sessions of plasma exchange, the patient was considered to have autoimmune encephalitis refractory to first-line therapies. Daily IVIG (20 g/day for 5 days, total 100 g) was initiated, followed by second-line therapy with ofatumumab on June 27, 2025. The treatment regimen consisted of 20 mg subcutaneous injections at weeks 0, 1, and 2, followed by monthly subcutaneous injections starting from week 4. Mycophenolate mofetil (2000 mg/day) was added to the regimen. For refractory seizures, lacosamide (2 × 200 mg) was started. Clobazam was added but proved ineffective. High-dose topiramate (800 mg/day) was initiated, which achieved substantial seizure control. Antiepileptic dosages were subsequently optimized. Weekly brain MRI scans demonstrated partial regression of prior lesions. The patient developed anemia following plasma exchange, for which erythrocyte transfusions were administered. The patient was extubated on July 14, 2025, and was able to follow simple motor commands such as eye opening and closing. On July 18, she was transferred from the ICU to a general ward. Due to weight loss, a high-protein diet was initiated. A follow-up brain MRI after extubation showed partial regression and signal changes in prior lesions. Immunosuppressive therapies (ofatumumab and mycophenolate mofetil 2000 mg/day) were continued. PET-CT revealed no evidence of malignancy. Viral serologies were unremarkable. At follow-up on August 11, 2025, neurological examination revealed normal conjugate gaze, full muscle strength (5/5), normal tandem gait, mild action tremor in both hands, negative parkinsonian signs, normal deep tendon reflexes, and absence of ataxia or pathological reflexes. Speech was mildly dysphonic with occasional brief hesitations during reading. Overall, her condition was stable, and she was discharged, Türkiye. **Conclusion:** DISCUSSIONAutoimmune encephalitis is an inflammatory brain disorder that may mimic infectious and other etiologies in terms of clinical, radiological, and serological findings, making diagnosis challenging. Since diagnosis is based on exclusion, immunosuppressive therapy must be initiated promptly. First-line therapies, including corticosteroids, intravenous immunoglobulin, and plasma exchange, are generally effective. In resistant cases, second-line therapy, especially anti-CD20 monoclonal antibodies such as ofatumumab, should be considered. Studies have demonstrated favorable responses with ofatumumab in patients with AE refractory to first-line therapies[8,9]. In our case, ofatumumab led to significant clinical improvement in a patient with severe, treatment-resistant AE. In conclusion, ofatumumab represents a promising treatment option for AE patients who fail to respond to first-line therapies such as corticosteroids, intravenous immunoglobulin, and plasma exchange.

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Adult Hematology Abstract Categories

Akut Lösemiler

PP 17

Long-term Success of Prophylactic Intrathecal Therapy in AML Patients with CNS Involvement: Two Cases with Extended Remission Following Stem Cell Transplantation

Birol Güvenç [1,*]

[1] *Çukurova University, Dept.of Hematology, Balcali_Adana,Turkiye*

Case 1: A 25-year-old female (born 1993) presented in 2018 with fatigue, anemia, and pancytopenia. Bone marrow biopsy revealed high blast count with flow cytometry showing CD33 (94%), CD117 (73%), MPO (98%) positivity, and limited CD34 (3-4%) expression. Cytogenetic analysis was negative for t(8;21), inv(16), t(15;17), and BCR-ABL, classifying the case as cytogenetically normal intermediate-risk AML. Brain MRI revealed

meningeal involvement at diagnosis. The patient received standard 7+3 induction (cytarabine + idarubicin) achieving complete hematologic remission. Due to absence of suitable donor, autologous stem cell transplantation was performed. Despite achieving complete remission and remaining asymptomatic, prophylactic intrathecal methotrexate and cytarabine was initiated every 6 months for CNS protection. Serial CSF examinations from 2019-2024 showed no blast cells. Bone marrow biopsies consistently demonstrated hypocellular marrow without blasts, with negative CD34 and CD117. The patient has maintained complete remission for 7 years without neurological symptoms or complications. Case 2: A 46-year-old male (born 1973) presented in 2019 with anemia, thrombocytopenia, and fatigue. Bone marrow analysis confirmed AML with flow cytometry showing CD33 (99%), MPO (98%), high HLA-DR expression, but negative CD34 and CD117, consistent with aggressive AML phenotype. CSF examination at diagnosis confirmed CNS involvement. After achieving complete remission with standard 7+3 induction (cytarabine + daunorubicin), the patient underwent allogeneic stem cell transplantation. Similar to Case 1, prophylactic intrathecal methotrexate and cytarabine was administered every 6 months despite clinical remission. Follow-up from 2020-2023 showed consistently negative CSF examinations and stable bone marrow remission with <5% blasts. The patient has maintained complete remission for 6 years without transplant complications or neurological sequelae. Discussion: These cases demonstrate several important clinical principles in managing AML with CNS involvement. First, both patients achieved sustained remission despite CNS involvement at diagnosis, traditionally associated with poor prognosis. The combination of intensive systemic therapy, stem cell transplantation, and prolonged prophylactic intrathecal therapy appears crucial for success. The extended prophylactic intrathecal therapy regimen (6-7 years) far exceeds standard recommendations but proved remarkably safe and effective. The 6-monthly interval appears optimal, providing adequate CNS protection while minimizing procedure-related risks and patient burden compared to more frequent administration. The contrasting transplant approaches (autologous vs. allogeneic) achieved similar outcomes, suggesting that the prophylactic intrathecal strategy may be more important than transplant type for CNS disease control. Both patients demonstrated excellent tolerance to repeated lumbar punctures without cumulative neurotoxicity. The absence of CNS relapse in both cases over 6-7 years strongly supports the efficacy of this prophylactic approach. Traditional concerns about prolonged intrathecal therapy causing neurotoxicity were not observed, possibly due to the extended interval between treatments. Conclusion: Extended prophylactic intrathecal therapy administered every 6 months following stem cell transplantation represents a safe and highly effective strategy for preventing CNS relapse in AML patients with initial CNS involvement. These cases challenge conventional limitations on prophylactic therapy duration and support consideration of extended prophylaxis in high-risk patients. The excellent long-term outcomes without significant complications suggest this approach should be considered for similar cases, potentially improving survival in this traditionally poor-prognosis population.

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PP 18

MYELOID SARCOMA PRESENTING IN THE RETROMOLAR TRIGONE WITHOUT MARROW INVOLVEMENT

Berrak Çağla ŞENOL ARSLAN*, Ali TURUNÇ, Ayşegül Ezgi ÇETİN, Birol GÜVENÇ

Çukurova University, Department of Internal Medicine, Division of Hematology, Türkiye

Introduction: Myeloid sarcoma (MS), also known as granulocytic sarcoma or chloroma, is a rare extramedullary tumor composed of myeloblasts. It may occur de novo, concurrently with acute myeloid leukemia (AML), or as a relapse of previously treated AML. Oral cavity involvement is rare, and isolated presentations without bone marrow disease pose significant diagnostic challenges . MS is biologically considered equivalent to AML and should be treated accordingly, even in the absence of systemic disease . **Case Presentation:** A 51-year-old woman presented with left facial swelling and dysphagia. MRI revealed a large mass in the left retromolar trigone extending to the skull base and infratemporal region with associated mandibular bone destruction. Incisional biopsy showed sheets of immature myeloid cells. Immunohistochemistry was positive for CD117, CD34, myeloperoxidase (MPO), and CD99, with a Ki-67 proliferation index of ~40%, confirming myeloid sarcoma. PET-CT revealed a hypermetabolic mass (SUVmax 7.27) and ipsilateral cervical lymphadenopathy but no systemic FDG-avid disease. Bone marrow biopsy showed no leukemic infiltration The patient was treated for acute myeloid leukemia and was started on a 7+3 chemotherapy protocol. The patient is being monitored during the post-treatment cytopenic period. **Conclusion:** ConclusionThis case highlights the diagnostic complexity of isolated myeloid sarcoma in an unusual location. Comprehensive immunophenotypic analysis is essential for diagnosis. Although marrow was uninvolved, the patient was initiated on AML-type induction chemotherapy due to the high risk of progression . Early systemic treatment, rather than localized therapy alone, is critical to avoid transformation into overt leukemia . Systemic chemotherapy using AML-like regimens should be commenced early, even in nonleukemic disease. Surgery and/or radiotherapy may be indicated for symptomatic lesions or tumors causing local organ dysfunction or obstruction. Allogeneic hematopoietic stem cell transplantation has demonstrated promising results, particularly in patients who achieved complete remission with AML-induction protocols, and recent advances in genetic profiling may enable the development of novel targeted therapies . Clinicians should maintain a high index of suspicion for MS in atypical head and neck masses.

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