scanned and no reduction in the mass was observed. Glofitamab therapy was initiated with off-label consent. The first and second cycle was completed. The patient did not develop cytokine release syndrome or neuropathy. A PET-CT scan was scheduled for 3 weeks later for response evaluation. The PET-CT scan showed that the intra-abdominal mass had regressed to 8 mm and the SUV(max) value to 3.44. The patient, who responded to glofitamab treatment, was offered autologous stem cell transplantation or CAR-T (Chimeric Antigen Receptor T-cell) therapy options. The patient requested to be referred to a center where CAR-T therapy could be performed. The patient is currently awaiting CAR-T therapy. **CONCLUSION:** In eligible DLBCL patients, salvage chemoimmunotherapy and/or monoclonal antibodies can be used as bridge therapies to OKIT or CAR-T therapies. Glofitamab is a bispecific antibody targeting CD20 and CD3, approved for r/r DLBCL patients after at least two prior lines of therapy. In a study, Glofitamab demonstrated a 46% ORR (27% CR; 19% PR) and manageable safety in heavily pretreated r/r DLBCL patients.

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A CASE OF REFRACTORY MULTIPLE MYELOMA WITH HYPOGLOSSAL NERVE INVOLVEMENT

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Multiple myeloma (MM) is a clonal stem cell disease originating from plasma cells. The development of MM neurological findings is mostly caused by hyperviscosity, hypercalcemia, amyloidosis, vertebral bone involvement and spinal cord compression due to fractures or nerve compression, neuropathy due to paraproteinemia. Brain involvement is very rare. It may present as cerebral lesion, parenchymal disease or leptomeningeal involvement. A case of isolated hypoglossal nerve involvement under MM treatment at the age of 52 will be presented. Cranial nerve involvement in multiple myeloma patients can occur without the development of plasmacytoma at the skull base, but rather through infiltration of the nerve sheath with plasma cells. Combination therapies must be administered to the patient. Isolated cranial nerve involvement is a very rare complication of multiple myeloma. INTRODUCTION: Multiple myelom is clonal stem cell disease originating from plasma cells. These cells produce monoclonal immune globulins, most commonly Immunoglobulin G (IgG) or Immunoglobulin A (IgA). The disease often leads to a variety of symptoms, including anemia, bone pain, increased incidence of fracture, hypercalcemia, renal failure and increased susceptibility to infections(1).Neurological complications in MM most commonly occur due to spinal cord compression from bone lesions, paraprotein associated neuropathy, hypercalcemia, hyperviscosity or amyloidosis(2). Central nervosus system (CNS) involvement may manifest as either a solitary cerebral lesion, intra-parenchymal infiltration, or diffuse leptomeningeal disease such as CNS myelomatosis(3). The average survival after CNS involvement is 3 months (1,3). **Clinical Presentation:**A 46-year-old male patient presented with complaints of pain in the left shoulder and chest that started on January 0, 2019, in addition to weight loss, weakness in the legs, and difficulty in walking. Laboratory Findings:In the examinations performed, wbc: 8.24 10e3/uL hbg: 9.23 gr/dl, plt: 118 10e3/uL, pnl: 5.65 10e3/uL, urea: 117 mg/dl, creatinine: 3.85 mg/dl, albumin: 2.79 gr/dl, globulin: 3.81 gr/dl, corrected calcium: 9.16 mg/dl, beta2 microglobulin: 0.44 mg/dl, serum free lambda light chain: 1190 mg/l increased and serum free kappa light chain: 5.25 mg/l, 24-hour urine immunofixation electrophoresis revealed free lambda light chain: 5.65 mg/l, Chain band was detected. In the MR imaging, there were nodular lesions in the iliac bone and sacrum, immunoglobulin values were IgA: 25 mg/dl, IgG: 293 mg/dl, IgM: 68 mg/dl, 80% plasma cells in the bone marrow. Treatment: The patient was diagnosed with multiple myeloma and started on bortezomib, cyclophosphamide, dexamethasone (VCD) chemotherapy. After 4 cycles of VCD and radiotherapy (RT), an increase in light chains was observed in the control evaluation, and a bortezomib, lenalidomide and dexamethasone (VRD) course was started. The patient, who underwent autologous BMT in December 2019, was followed up under lenalidomide cordexa maintenance treatment, and a relapse was detected in the control evaluations in June 2022. Ixazomib, lenalidomide and dexamethasone (IRD) treatment was started, and daratumumab VCD treatment was switched to due to lack of response. Outcome: The patient, who was followed up under daratumumab VCD treatment, developed complaints of decreased hearing and numbness in the jaw. In addition to the atrophy in the left half of the tongue and the complaint of shifting to the left when the tongue was taken out of the mouth, speech and swallowing were impaired. (Figure 1)Neurology consultation and detailed brain imaging showed increased thickness in the right maxillary sinus. In the PET-CT imaging, although there was no pathological involvement in the head, neck and mediastinal structures, widespread lytic lesions were seen especially in various vertebrae, femur and tibia, and it was evaluated as progressive disease. No plasma cells or other pathology was detected in intrathecal sampling. Peripheral smears were examined daily with suspicion of plasma cell leukemia, but plasma cells were not detected. No signs of neurological diseases such as cranial hemorrhage or embolism were found that would cause this clinic. Conclucion and Results: The patient was evaluated in the council with neurology and it was evaluated that there was isolated myeloma involvement of nervus hypoglossus and the treatment was arranged as dartumumab, pomalidomide and dexamethasone. Brain RT was performed. After 2 cycles of chemotherapy and radiotherapy, the patient's tongue numbness, speech and swallowing disorders improved. The complaint of left shift when the tongue came out of the mouth regressed(Figure 2). The patient's isolated nervus hypoglossus involvement improved with treatment, and his follow-ups are continuing. In conclusion, cranial nerve involvement in multiple myeloma patients can occur without the development of plasmacytoma at the skull base, but rather through infiltration of the nerve sheath with plasma cells. Combination therapies must be administered to the patient. Conclusion: Brain involvement may very rarely develop in 1% of patients with multiple

myeloma (4). Parenchymal involvement may be in the form of mass compression or leptomeningeal involvement due to plasmacytomas (5). The most common cranial nerve involvements in the literature are the oculomotor nerve, abducens nerve and hypoglossal nerve (2). Involvement of these cranial nerves is most commonly due to plasmacytomas originating from the skull base and sinuses (6). When other cases of multiple myeloma with hypoglossal nerve involvement were scanned in the literature, it was seen that there was plasmacytoma or leptomeningeal involvement (7,8). In our case, it is a very rare condition in terms of the absence of a mass lesion, plasmacytoma and leptomeningeal involvement in the brain. When the case was re-examined after the literature scan, it was seen that there was a prominent paranasal sinus wall thickening on the right side that developed during the period when the patient's complaints began, without any mass lesion or leptomeningeal involvement. However, in a case report, a patient with a soft tissue mass in the right paranasal sinus had significant plasmacytomas at the skull base, and clinically, there was oculomotor, facial and hypoglossus nerve involvement (9). This situation suggests that involvement in the paranasal region may require differentiation from classical infection conditions in patients at risk and close monitoring in terms of intracranial events. Plasmacytomas of the skull base occur as an extension of plasmacytoma originating from the clivus, petrous part of the temporal bone or from the submucosa of the sinonasal and nasopharyngeal (extramedullary plasmacytoma) region. Extramedullary plasmacytomas are most commonly seen in the nasal and paranasal sinuses, nasopharynx, tonsils and larynx (10). Due to the rarity of extramedullary plasmacytomas, data on treatment and prognosis are limited. However, studies have shown the effectiveness of radiotherapy. There are publications showing that 30-50 gy radiation most effectively reduces tumor sizes caused by multiple myeloma (11)..In our case, radiotherapy and appropriate chemotherapy were initiated. Cranial nerve involvement is very rare in cases of relapsed refractory disease, and radiotherapy and combined chemotherapy are among the treatment options.

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Adult Hematology Abstract Categories

Platelet Diseases

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A RARE DIAGNOSIS IN ADULTS: HEREDITARY THROMBOTIC THROMBOCYTOPENIC PURPURA

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Introduction: Thrombotic thrombocytopenic purpura (TTP) is a thrombotic microangiopathy (TMA) resulting from severely reduced activity of ADAMTS-13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13), a metalloproteinase responsible for cleaving von Willebrand factor (vWF). It is characterized by disseminated platelet-rich microvascular thrombi leading to organ ischemia, neurological abnormalities, renal dysfunction, thrombocytopenia, and microangiopathic hemolytic anemia (MAHA). Hereditary TTP (hTTP; also referred to as congenital TTP [cTTP] or Upshaw–Schulman syndrome) arises from pathogenic variants in the *ADAMTS-13* gene and follows an autosomal recessive inheritance pattern. Although extremely rare, it can be life-threatening. Patients with hTTP require special attention during certain life stages such as the neonatal period and pregnancy . In contrast to immune-mediated TTP (iTTP), which typically presents with a dramatic and acute onset, hTTP may manifest with a more insidious clinical picture including lethargy, impaired concentration, abdominal pain, and headache . Severe renal failure, although uncommon in iTTP, may occur in hTTP patients due to lifelong ADAMTS-13 deficiency, which can cause progressive accumulation of thrombi within the renal vasculature Hematologic examination may reveal pallor, purpura, and jaundice as signs of hemolysis, while laboratory findings typically show thrombocytopenia, unconjugated hyperbilirubinemia, elevated LDH levels, and decreased haptoglobin . Peripheral blood smear is often diagnostic, demonstrating schistocytes, nucleated red blood cells, and polychromatic red cells, consistent with intravascular hemolysis . Here, we describe the case of an 18-year-old patient with congenital TTP who was initially misdiagnosed with myelodysplastic syndrome (MDS) at an early age and received intermittent transfusions due to cytopenias. The aim of this case report is to raise clinical awareness regarding this rare and potentially fatal subtype of TTP, which can be rapidly and effectively treated if recognized early. In addition, it underscores the importance of reassessing patients at each presentation, even when a pre-existing diagnosis is available, and highlights the critical diagnostic value of peripheral blood smear, as the presence of schistocytes is pathognomonic for this condition. **Case report:** We present the case of an 18-year-old female patient, with no family history of hematologic disorders, who has been followed since childhood for anemia and thrombocytopenia. At the age of 18, she was diagnosed with congenital thrombotic thrombocytopenic purpura (TTP) and treatment was initiated. Her hematologic evaluation began in 2009, at the age of three, due to anemia and thrombocytopenia, during which she received platelet and red blood cell transfusions. Following a bone marrow examination, she was diagnosed with myelodysplastic syndrome (MDS). In 2018, she was also diagnosed with chronic kidney disease secondary to vesicoureteral reflux by the pediatric nephrology department, Türkiye. In 2024, at the age of 18, she presented to the emergency department with complaints of fatigue and dizziness. Laboratory tests revealed: WBC 5.36×10^9/L, Hgb 8 g/dL, Plt 8×10^9/L, INR 0.98, fibrinogen 211 mg/dL, creatinine 5.8 mg/dL, AST 21 U/L, ALT 15 U/L, uric acid 8.6 mg/dL, LDH 622 U/L, total bilirubin 2.5 mg/dL, direct bilirubin 0.35 mg/dL, and both direct and indirect Coombs tests were negative. The patient was admitted to the hematology clinic for further evaluation, Türkiye. Physical examination was notable only for mild pallor; no lymphadenopathy or organomegaly was detected. Peripheral