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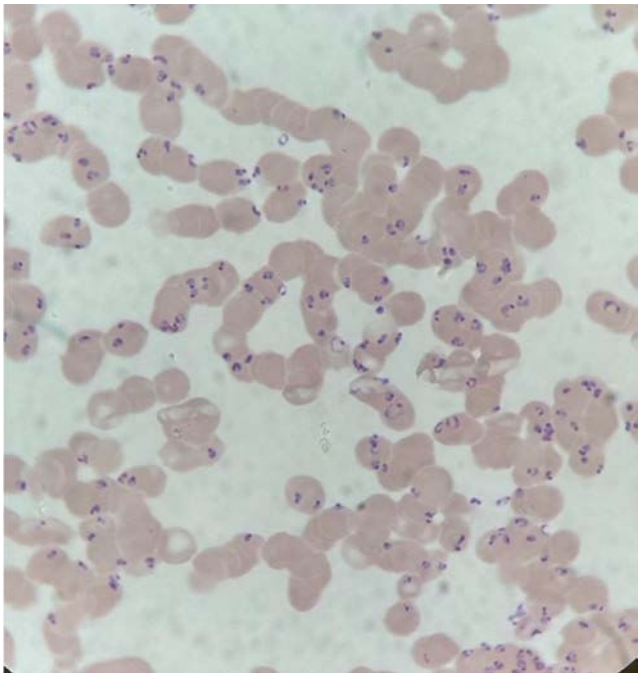

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PP 33

A CASE OF THROMBOTIC THROMBOCYTOPENIC PURPURA RELATED TO MALIGNITY AND CHEMOTHERAPY

Ebru Kavak Yavuz [1,*], Songül Beskisiz Dönen [1], Etem Özkaya [1], Esra Pirinççi [1], Abdullah Karakuş [1], Orhan Ayyıldız [1]

[1] *Dicle University*

Objective: Thrombotic thrombocytopenic purpura (TTP) is a life-threatening multisystem disease. TTP progresses with Microangiopathic hemolytic anemia (MAHA), fever, thrombocytopenia, neurological symptoms, and renal failure. Due to microangiopathic hemolytic anemia, schistocytes are seen in the peripheral blood smear, resulting in thrombocytopenia. Damage to the brain and kidneys occurs due to microvascular thrombosis, and this is how symptoms appear. In pathogenesis, it is caused by the deficiency of ADAMTS 13 (a disintegrin and metalloproteinase with thrombospondin type 1 motif, member 13), which breaks down the von Willebrand factor (VWF) found in the endothelium into multimers, or the development of antibodies against it. Due to ADAMTS 13 deficiency or decrease in its activity, VWF cannot be separated into small pieces and is arranged in large pieces in the endothelium, causing widespread intravascular thrombosis. Many factors can be considered as triggers for the development of TTP, such as pregnancy, malignancy, medications, and autoimmune diseases. **Case Report:** A 59-year-old female patient was admitted due to thrombocytopenia, epileptic seizure, hematemesis and decreased consciousness while being followed up due to cholangiocellular carcinoma. Due to malignancy, 6 cycles of gemcitabine and carboplatin treatment were applied. The last cure was 6 months ago. In followers, WBC 3.24 10^3/uL (3.7-10 10^3/uL), Hbg 8g/dL(12.9-14.2 g/dL), MCV 84 f/L(81-96fL), platelet 27 10^3/uL (155-356 10^3u/L), total bilirubin 13 mg/dL (0.3-1.2 mg/dL), indirect bilirubin 5.36 mg/dL (0-1.5 mg/dL), LDH 408 U/L (0-247u) /L), creatinine 1.59 mg/dL (0.51-0.95 mg/dl), protein 1+ in full criterion examination, INR 1.36, PT 15.4 sec (10-15 sec), APTT 22.2 sec (21-29 sec) fibrinogen was 1.46 g/L (1.8-3.5 g/L), 3-5 schistocytes were seen in each area in the peripheral smear. Plasmapheresis treatment was started with the preliminary diagnosis of TTP and steroid 80 mg was given. ADAMTS 13 tests were requested. ADAMTS 13 level is 3.78% (40%-130%) low and ADAMTS 13 inhibitor > 80 U/mL (<12U/mL negative, 12-15 U/mL borderline >15U/mL positive), ADAMTS 13 antigen<0.01lU/ mL (0.19-0.81 lU/mL) was seen. As the patient's thrombocytopenia continued, plasmapheresis was started to be performed twice a day after a week. With this treatment, weekly treatment of medicinal rituximab, which could not be treated with platelets, was arranged. However, the patient did not respond to treatment and died. **Conclusion:** In cancer assosiated TTP, endothelial cells are damaged due to abnormal angiogenesis and tumor cell invasion, and vWF multimers in the endothelial wall are exposed. In addition, ADAMTS 13 activity decreases due to antibodies formed against ADAMTS 13. Some chemotherapeutics such as mitomycin c, gemcitabine can cause TTP. When a diagnosis of TTP is considered, plasma exchange should be started immediately. In addition to plasma exchange, steroids are given in the treatment and if there is no response, other immunosuppressive treatments are added. Our patient with high ADAMTS 13 inhibitors is a condition that is thought to contribute to the mortality of TTP. In a study, it was observed that low ADAMTS 13 activity, as well as high ADAMTS 13 inhibitor and low ADAMTS antigen, caused an increase in mortality.

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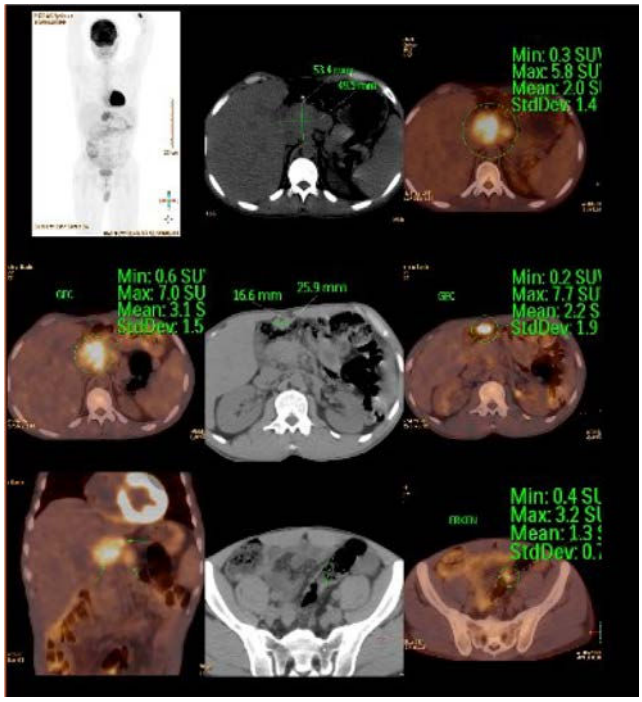
PP 34

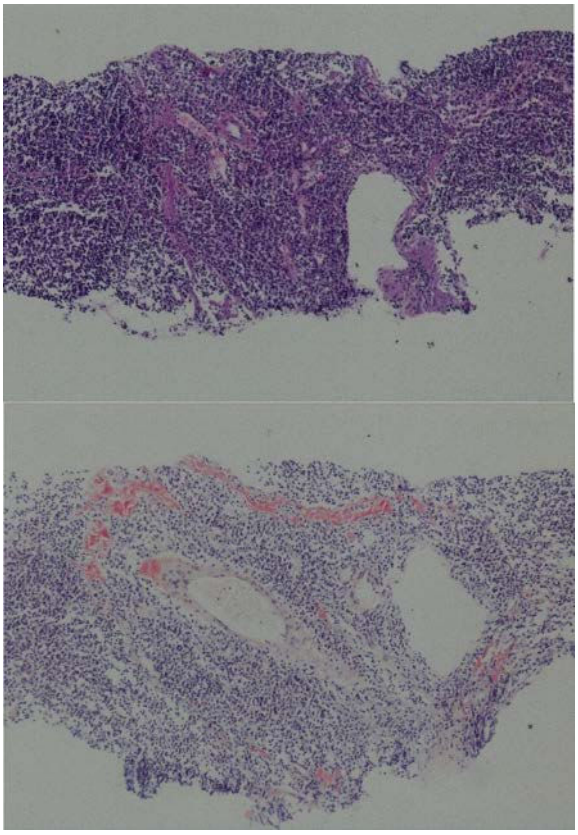
A RARE DISEASE ASSOCIATED WITH IG4, CHARACTERIZED BY SYSTEMIC AMYLOIDOSIS AND LYMPHOPLASMACYTIC CELL DOMINANCE: A CASE PRESENTATION

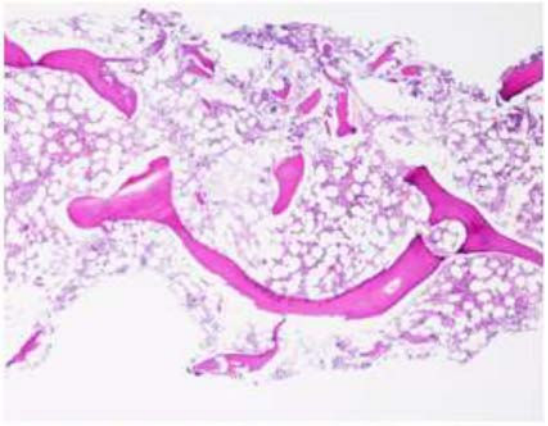
Şerife Emre Ünsal [1,*], Mihriban Yıldırım [1], Hacı Ahmet Aslaner [1], Neslihan Mandacı Şanlı [1], Gülşah Akyol [1], Muzaffer Keklik [1], Özlem Canöz [2], Olgun Kontaş [2], Ali Ünal [1]

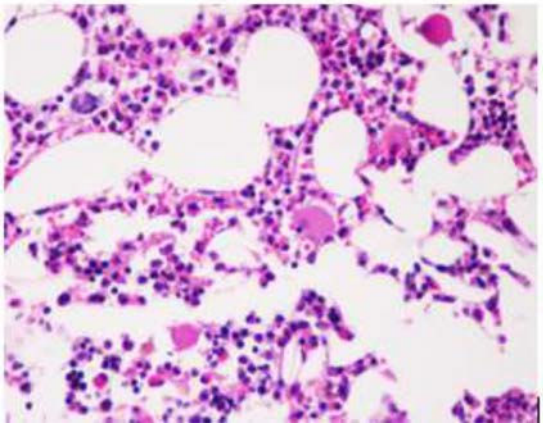
[1] *Erciyes University Faculty of Medicine, Department of Hematology*
[2] *Erciyes University Faculty of Medicine, Department of Pathology*

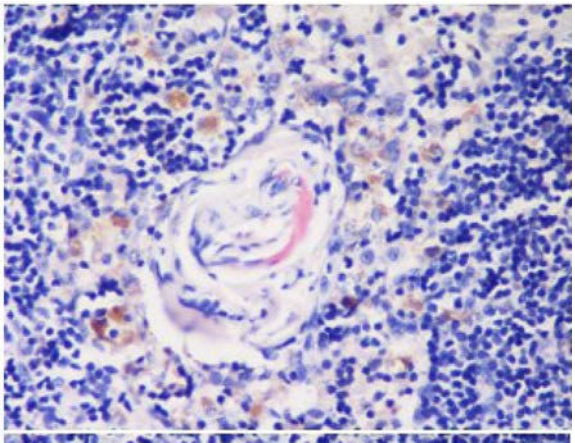
Objective: Immunoglobulin G4 (IgG4)-related disease has been identified in the last 10-15 years, though it was previously known in the literature under different names as an autoimmune disorder. The spectrum of the disease is quite broad. It can present with involvement of a single organ or multiple organs simultaneously, including autoimmune pancreatitis, Mikulicz syndrome, Küttner tumor (chronic sclerosing sialadenitis), sclerosing cholangitis, and retroperitoneal fibrosis. It most commonly occurs in males over the age of 50. In this case presentation, we will discuss a patient who presented with systemic amyloidosis and was diagnosed with IgG4-related disease. **Case Report:** A 36-year-old male patient presented to the hospital with complaints of abdominal pain and constipation. He was evaluated through detailed anamnesis and physical examination. The patient was found to have iron deficiency anemia and elevated acute phase reactants. An abdominal ultrasound revealed a mass in the epigastric region, leading to admission to the gastroenterology department. A CT scan of the abdomen showed a 62 × 55 mm lesion in the epigastric region. A trucut biopsy was performed, which was reported as amyloidosis. The biopsy revealed an increase in plasma cells. A PET-CT scan identified hypermetabolic lymph nodes in the celiac trunk region. A biopsy taken from these nodes was also reported as amyloidosis, with no evidence of monoclonality. Results showed positivity for CD138, Kappa, Lambda, Congo red, and IgG4, with negativity for HHV8. Serum IgG level was 3256 mg/dL, albumin was 3.59 g/dL, total protein was 8.59 g/dL, sedimentation rate was 65 mm/h, and elevated levels of free kappa and lambda light chains were detected. The patient developed renal failure and hyperkalemia. A renal biopsy showed positive staining for AA amyloid, and a bone marrow biopsy was subsequently performed. The PET-CT scan did not reveal plasmacytoma or osteolytic lesions. The bone marrow biopsy showed 7-8% staining with CD38 and CD138. Positive staining was noted for AA amyloid, IgG, and IgG4, particularly in plasma cells. An initial diagnosis of lymphoplasmacytic lymphoma was considered, and excisional biopsies of lymph nodes were planned. The excisional biopsy of the left axillary lymph node was reported as amyloidosis, leading to a referral to the rheumatology department to investigate secondary causes of amyloidosis. IgG subclasses were tested, revealing an IgG4 level of 700 mg/dL. The patient was started on corticosteroid therapy at a dose of 1 mg/kg. **Conclusion:** IgG4-related disease is a fibroinflammatory condition that can affect any organ simultaneously or at different times. It is a systemic disease that can involve all organs and often presents with organomegaly, mimicking malignancy. The immunopathogenesis of the disease is not yet fully understood. The most critical step in diagnosis is the histopathological evaluation of the affected organ. Histopathological features distinguishing the disease include dense lymphoplasmacytic infiltrates with predominance of IgG4-positive plasma cells, storiform fibrosis, and obliterative phlebitis. There are no specific diagnostic tests for IgG4-related disease, making differential diagnosis very important. The first comprehensive diagnostic criteria for IgG4-related disease were established in 2011, and new classification criteria were introduced in 2019. A serum IgG4 level of ≥ 135 mg/dL is significant for diagnosis. The primary treatment for IgG4-related disease is corticosteroids, which typically respond well to therapy. Most patients show a response to treatment within 4 weeks. With therapy, patients often experience a reduction in symptoms, a decrease in the size of masses in affected organs, improvement in organ function, and a general decline in serum IgG4 levels over several weeks. After the initial response, the dose should be gradually reduced by 5 mg every 2 weeks to maintain remission, ideally for a duration of 3-6 months at the lowest effective dose. However, relapses can occur, and in cases of resistant or recurrent disease, additional treatments such as rituximab and other immunosuppressive agents may be required. These include azathioprine (2 mg/kg/day), mycophenolate mofetil (1-1.5 g/day), and cyclophosphamide (50-100 mg/day). Biological agents such as infliximab, tocilizumab, calcineurin inhibitors, and bortezomib may be used for refractory cases. Studies evaluating the effectiveness of monoclonal agents like abatacept, inebilizumab, and elotuzumab in the treatment of IgG4-related disease are also available. Early diagnosis and appropriate treatment are crucial for controlling the disease and preventing complications.

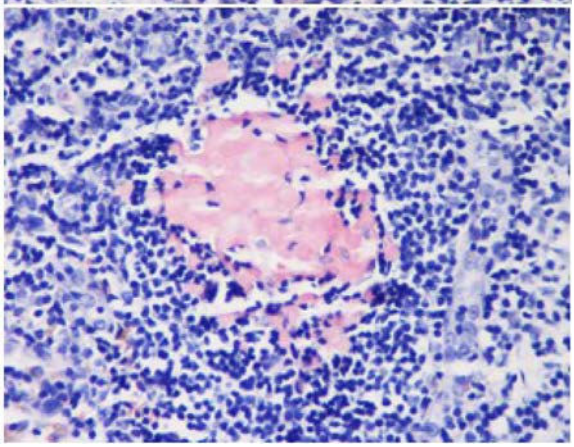


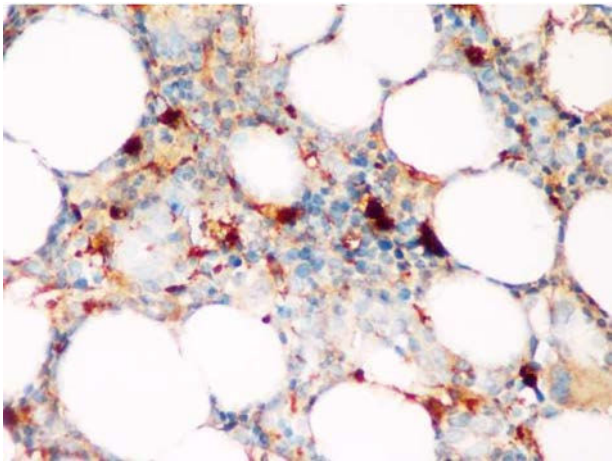


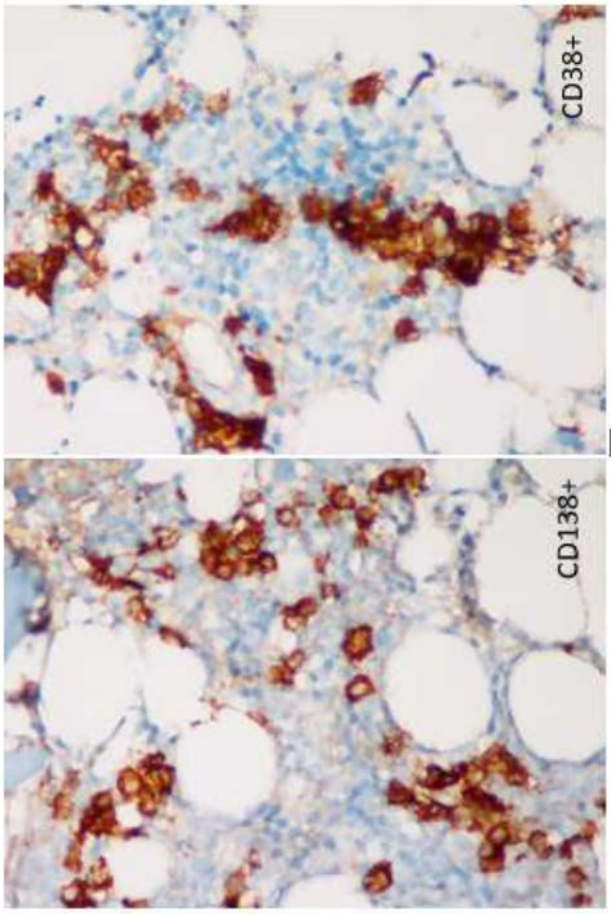












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PP-35

B-LINEAGE PROGENITORS AND CD38-POSİTİVE B CELLS ARE ASSOCIATED WITH SURVIVAL RATES IN BREAST CANCER PATIENTS

Svetlana Chulkova [1,2,*]

[1] *FSBU "N.N. Blokhin National medical research center of oncology" of the Russian Ministry of Health, Moscow; Kashyrskoe sh.24, Moscow, 115478, Russia*
[2] *Pirogov N.I. Russian National Research Medical University of the Russian Ministry of Health; 1, Ostrovitianova st., Moscow, 117997, Russia*

Objective: The immune system plays an increasingly important role in the development of targeted strategies for breast cancer. According to mRNA sequencing data from The Cancer Genome Atlas (TCGA) high expression B cell signatures has beneficial effects on survival rates in many tumors. Bone marrow (BM) is poorly understood from the point of view of the prognostic role of hematopoietic cells and subpopulations of lymphocytes in patients with breast cancer (BC). **Methodology:** . Study was carried out in 107 BC patients. The immunological and morphological methods were applied. Multiparameter flow cytometry with antibodies to B-cell populations was used (CD19, CD20, CD5, CD38, CD10, CD45, HLA-DR, CD27), FACSCANTO II. Studies of BM lymphocyte subpopulations were carried out in the gate of CD45++ cells. The duration of the follow-up period after surgery was 8 years. **Results:** The total percentage of B cells in BM was significantly associated with the prognosis of BC. B-1 cells were associated with progression-free and disease-free survival. Disease progression was observed at low levels of B1 cells. In cases more than 10% B-lymphocytes in the BM of BC patients overall survival (OS) rates were more favorable ($p = 0.01$). Especially for BC with a high Ki-67. Disease progression was observed in 1/3 of BC patients with low levels of B1 cells. CD38 expression on B cells was a prognostically favorable factor: the role is realized during 5–10 years of follow-up after surgery. Level CD38+ B cells more then 10% correlated with high OS, $p = 0.02$. The presence of CD10+CD19+ B-lineage precursors was associated with a more favorable prognosis (OS, the threshold level 12%, $p = 0{,}04$). The prognostic role of the CD10 antigen was realized when patients were observed for more than 5 years. **Conclusion:** . Total relative number of (more than 10 %) of BM CD19+ cells were significantly related to OS in BC. B-cell precursors and CD38+ B cells were associated with favorable prognosis. Prognositic role of B-lineage precursors and CD38-positive cells was in the periods of 5–10 years after surgery.

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PP 36

SUCCESSFUL CHEMOTHERAPY ADMINISTRATION DESPITE HYPERSPLENISM AND PANCYTOPENIA: A CASE OF METASTATIC RECTAL ADENOCARCINOMA

Adil Uğur Kaan Güngör [1,*], Abdurrahman Aykut [2], Berksoy Sahin [2], Hatice Asoğlu Rüzgar [2]

[1] *Çukurova University, Faculty of Medicine, Department of Internal Medicine*
[2] *Çukurova University, Faculty of Medicine, Department of Medical Oncology*

Introduction: Cytopenias in oncology patients present a significant barrier to the administration of chemotherapy. Hypersplenism is one of the leading causes of cytopenia. In this case report, we aim to present a patient diagnosed with metastatic rectal adenocarcinoma, who developed hypersplenism due to liver metastasis and was successfully treated with chemotherapy despite the cytopenias. **Case Report:** In September 2023, a 42-year-old female patient was diagnosed with rectal adenocarcinoma with liver metastasis. Genetic analysis revealed K-Ras, N-Ras, and BRAF mutant/wild type, MSI stable, and Her2 negative. The patient received 3 cycles of FOLFIRINOX chemotherapy. During follow-up, her hemogram results were as follows: hemoglobin: 8.6 g/dL, platelets: $26 \times 10^3/\mu$L, leukocytes: $0.81 \times 10^3/\mu$L, and neutrophils: $0.37 \times 10^3/\mu$L. PET-CT evaluation showed regression in the metastatic lesions and newly developed splenomegaly