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A rare subtype of poems syndrome: IGG4 subtype

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Objective: There is very limited data concerning the relationship between POEMS syndrome and IgG4-related disease.

Case report: A 40 year-old male patient presented with a 3 month history of progressive weakness and numbness in his lower extremities, impotence, diarrhea and weight loss. Complete blood count was as follows: WBC: 7.3×10^9/L, Hgb: 16.5 g/L, platelet 543×10^9/L. Liver enzymes, renal function, electrolytes and routine urine examination were normal. Ig G level was 14.5 g/dL (normal: 7–16 g/L). Serum immunofixation electrophoresis showed IgG λ monoclonality. Endocrine laborotory tests showed hipergonadotropic hipogonadism. Echocardiography showed pericardial effusion. Abdominal USG showed hepatomegaly and splenomegaly measuring 200 mm and 174 mm on longitudunal axis, respectively. On contrast enhanced MRİ, a 6 cm × 3.5 cm mass showing bone destruction was detected in the left sacral ala extending into the pelvis. PET CT scan demonstrated high FDG uptake (SUVmax: 10.5) for the sacral mass lesion. Based on these findings, a diagnosis of POEMS Syndrome was considered. Funduscopic examination showed no papiloedema. Vascular endothelial growth factor (VEGF) was very high (>700 pg/mL, normal: <96 pg/mL). Trucut biopsy of the mass lesion consisted of a nonneoplastic fibrous tissue and a dense infiltrate of mature plasmacytes with dense eosinophilic cytoplasm and eccentrically placed nuclei. Also, perivascular accumulation of sclerotic collagen like substance was noted. On immunohistochemical staining, neoplastic cells showed diffuse positivity for Ig G and Ig G4. Neoplastic cells were CD138(+), κ(−), λ(+), CD38(+), CD30 (−), ALK(−), CD20(−), CD10(−), CD23(−), CD45(−), CD56(−), CD57(−). Bone marrow biopsy showed a 3% monoclonal λ(+) plasma cell infiltration. Diagnosis of POEMS syndrome was confirmed. Taking into consideration high IgG4 expression in the neoplastic mass, IgG4 levels in serum was checked and found to be high 6.34 g/L (normal <1.35 g/L).

Methodology: POEMS syndrome and IgG4 related diseases show similarities including organomegaly and systemic organ damage. Polyneuropathy and bone lesions associated with IgG4 related diseases has not been reported. PET/CT detects bone lesions and lymph nodes in patients with suspected POEMS syndrome. In IgG4 related disease on the other hand, PET/CT identifies multiple lymph node enlargements/organomegaly with normal metabolic activity.

Results: Our patient had an osteosclerotic mass lesion demostrated by PET/CT and histopathological examination. Our patient had high serum IgG4 level and showed IgG4 plasmacyte tissue infiltration, yet her plasmacytes were shown to be monoclonal by bone marrow immunohistochemical staining and serum immunofixation electrophoresis. Therefore, final diagnosis was POEMS syndrome but not IgG4 related disease.

Conclusion: We propose this patient has a subtype of POEMS syndrome because he showed high serum IgG4 levels and a monoclonal IgG4 plasmacyte tissue infiltration. Monoclonal hyperglobinemia is not a feature of IgG4 related disease. It is not clear whether IgG4-positive plasma cell tissue infiltration and elevated serum IgG4 concentrations are origins or outcomes of IgG4 related diseases. To our knowledge, this is the second presumed case of POEMS syndrome-IgG4 subtype. Further research and collecting more cases are essential. We suggest every suspected POEMS patient should be tested for their serum IgG4 concentration.

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Monoclonal gammopathy of undertemined significance and solitary plasmacytoma: progression factors in population of gomel region in belarus

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Objective: To define progression factors of MGUS and SP in population of Gomel region in Belarus.

Case report: Solitary plasmacytoma (SP) and monoclonal gammopathy of undetermined significance (MGUS) are characterized by the presence of less than 10% of tumor cells in the bone marrow and the absence of CRAB criteria. Both diseases have a high risk of progression to multiple myeloma due to certain factors.

Methodology: The study included 106 patients: MGUS ($n = 90$) and SP ($n = 16$) of Gomel region (Belarus) in 2017–2019. The average age was 60.5 years; female patients prevailed All patients underwent aspiration biopsy with IPT and FISH, trepanobiopsy of the ilium wing with immunohistochemical examination of the bone marrow. (Bone marrow aspirates IPT and FISH, and biopsies were obtained for cytological and histopathological evaluation of PC infiltration, including immunohistochemical). The determination of the ratio of light chains of immunoglobulins (kappa/lambda) in blood serum was carried out. Results were assessed after 3 years of observation. The signs of progression include the appearance of any one of the CRAB-criteria.

Results: There were no statistically significant differences between groups of patients with MGUS and SP according to signs (presence of tumor plasma cells, CD95+, CD200+, CD27+,