中文題目：案例報告之罕見的骨髓增生性腫瘤（MPNs）和多發性骨髓瘤（MM）同時存在

英文題目：A case report of coexistence of myeloproliferative neoplasms (MPNs) and multiple myeloma (MM)

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Abstract:

The coexistence of myeloproliferative neoplasms (MPNs) and multiple myeloma (MM) is rare. MPNs represent a heterogeneous group of clonal hematopoietic disorders. Philadelphia chromosome-negative MPNs, majorly including polycythemia vera (PV), essential thrombocythemia (ET), and primary myelofibrosis (PMF), not otherwise specified MPN, unclassifiable (MPN-U). As for MM, a neoplastic proliferation of plasma cells in bone marrow, produces monoclonal immunoglobulin, causes hypercalcemia, renal insufficiency, anemia and bone pain. Coexistence is rare, therefore we report a case and literature review.

A 60-year-old Taiwanese male presented with persisted asymptomatic leukocytosis while health-examination in January 2018. The white cell count was 42.58×10^9/L with normal hemoglobin (Hb) and platelet (PLT), without blast cells. Bone marrow examination revealed hypercellularity (cellularity 90%) with increase of megakaryocytes, increased myeloid/erythroid (M/E) ratio, prominent granulocytic proliferation, but silver stain reveals no reticulin fiber. Abdominal sonography revealed splenomegaly. Cytogenetics showed a normal karyotype. There was no JAK2V617F mutation, nor BCR/ABL-translocation. Under the impression of MPN, he had hydroxyurea therapy with improvement of leukocytosis [WBC:15.81×10^9/L] 1 months later, but anemia and thrombocytopenia. However, in May 2018, he suffered from diplopia, the neck pain and rigidity, weight loss (about 11kg in 5 months), and fatigue. Brain CT showed lobulated, soft tissue mass (4.55 x 4.88 x 4.3 cm^3) at the skull base. Laboratory data revealed severe hypercalcemia, renal failure and A/G reverse. Serum and urine immunofixation electrophoresis (IFE) showed monoclonal of IgGκ gammopathy. Bone marrow survey showed cellularity around 30% with atypical plasma cells, highlighted by CD138 but negative to CD79a. These plasma cells are kappa light chain restricted. The final diagnosis was multiple myeloma, monoclonal IgGκ gammopathy, ISS: II, with CNS plasmacytoma. Hence, he started myeloma treatment-VTD (bortezomib, thalidomide, and dexamethasone) since June 2018, and image guided radiotherapy (IGRT) of skull base. Treatment outcome was partial response. Brain MRI on December 2018 revealed decreasing tumor volume, and his clinical condition improved, with mild elevated free kapp. After 8 courses of therapy, VGPR was achieved, his clinical symptoms and lab data all improved.

Conclusions:

1. The coexistence of MPN and MM is rare, and the relationship with these different myeloid malignancy and lymphoid dyscrasias is unclear. The real mechanism is still unclear, but some suggested coexistence of 2 distinct clonal diseases or the biclonal evolution from a common hematopoietic stem cell and developing independently.
2. Our patient had a short interval from hydrea therapy to MM diagnosis. Through the risks is not high, we could not exclude the influence of hydrea on the unstable marrow microenvironment. In summary, we report here an interesting case and highlight the importance of suspicious relationship for second malignancies in these kinds of status.