JAK2 V617F

Polycythaemia vera (PV) is a clonal proliferative disorder of the bone marrow characterized by autonomous hematopoiesis, which results in a panmyelosis in the peripheral blood. It is typically characterized by an acquired mutation in JAK2 V617F. Progression to myelofibrosis (MF), characterized by worsening cytopenias and the development of constitutional symptoms, is seen in up to 10% of cases. Extramedullary hematopoiesis (EMH) in the spleen is a common finding in myelofibrotic transformation, but elsewhere in the body, it is extremely unusual. Randhawa et al. reported the case of a 69-year-old male whose PV progressed to secondary MF and who presented with compression of the thoracic spinal cord directly as a result of EMH. Cytogenetic and molecular findings in the bone marrow were in keeping with evolving myeloid disease. He was managed by surgical laminectomy with an excellent outcome. Extramedullary hematopoiesis may be seen in both PV and on transformation to MF. This very rare complication should be borne in mind when managing patients with myeloproliferative disorders. [1].

1)
Randhawa MS, Harrison L, Walkden J, Watson H. Spinal cord compression secondary to extramedullary haematopoiesis in transformed polycythaemia rubra vera. J R Coll Physicians Edinb. 2022 Mar;52(1):24-26. doi: 10.1177/14782715221088912. PMID: 36146966.