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Extramedullary hematopoiesis manifesting as pleural effusion in a patient with post-polycythemic myelofibrosis

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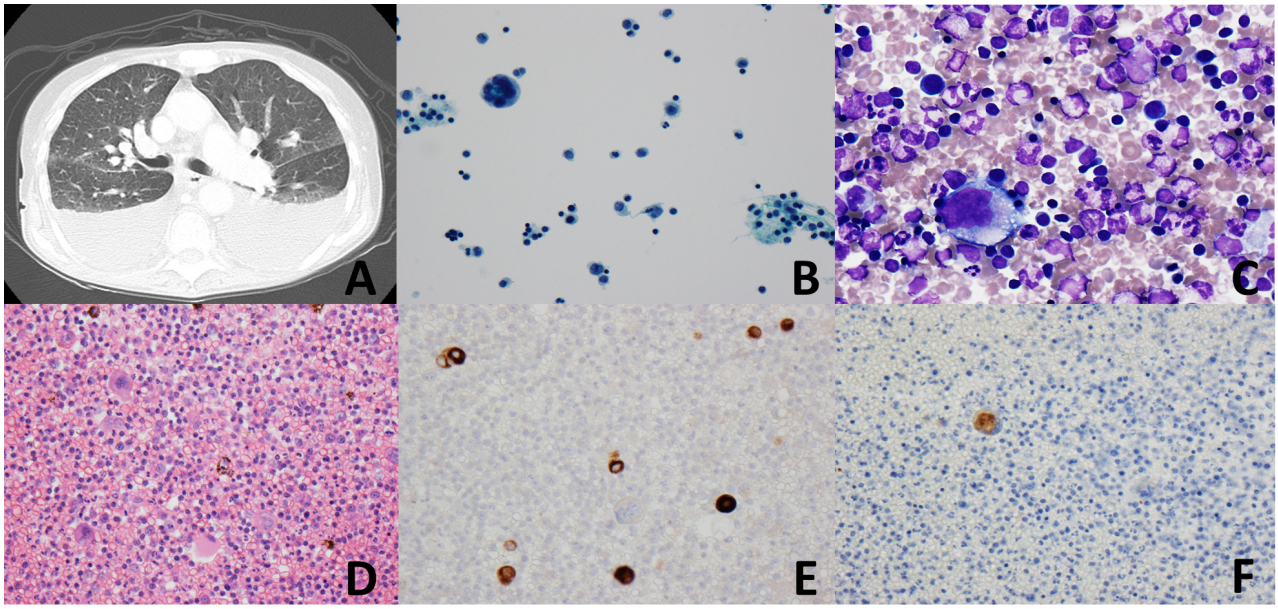

A 79-year-old woman with a long history of polycythemia vera (PV) with *JAK2* Exon 12 mutation (N542-E543 del) treated with biannual phlebotomy presented with recurrent pleural effusions (panel A), weight loss and hepatosplenomegaly for the past few months. Complete blood count showed neutrophilia with elevated immature granulocytes (ANC 15.6 x 10^9/L, immature granulocytes 0.2 x 10^9/L), microcytic anemia (HGB 11.4 g/dL, MCV 67 fL) and elevated hematocrit (50.9%). Nucleated red blood cells were also present. At the time of presentation, she had undergone thoracentesis 5 times in the past 6 weeks. The most recent sample of pleural fluid contained scattered megakaryocytes in a background of maturing erythroid and myeloid precursors (panel B, ThinPrep slide, Papanicolaou stain, x400; panel C, Wright-Giemsa stain, x600). Also present were abundant macrophages and reactive mesothelial cells (panel D, hematoxylin and eosin stain, x400) which stained positively with calretinin, D2-40 and CK7 (panel E, x400). The megakaryocytes were highlighted with CD61 (panel F, x400). Flow cytometry of the fluid showed no increased blasts or abnormal B or T cells.

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Follow up studies included a bone marrow biopsy which revealed a markedly hypercellular bone marrow with erythroid predominant trilineage hematopoiesis and moderate myelofibrosis (WHO MF-2), consistent with post-PV myelofibrosis. Iron studies revealed severe iron deficiency. Thalassemia and hemoglobinopathy work-up was negative. After initiation of treatment with ruxolitinib, the patient no longer required thoracentesis. Extramedullary hematopoiesis usually involves liver and spleen but can rarely manifest as pleural effusion in patients with primary or post-PV/ET (essential thrombocythemia) myelofibrosis.

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