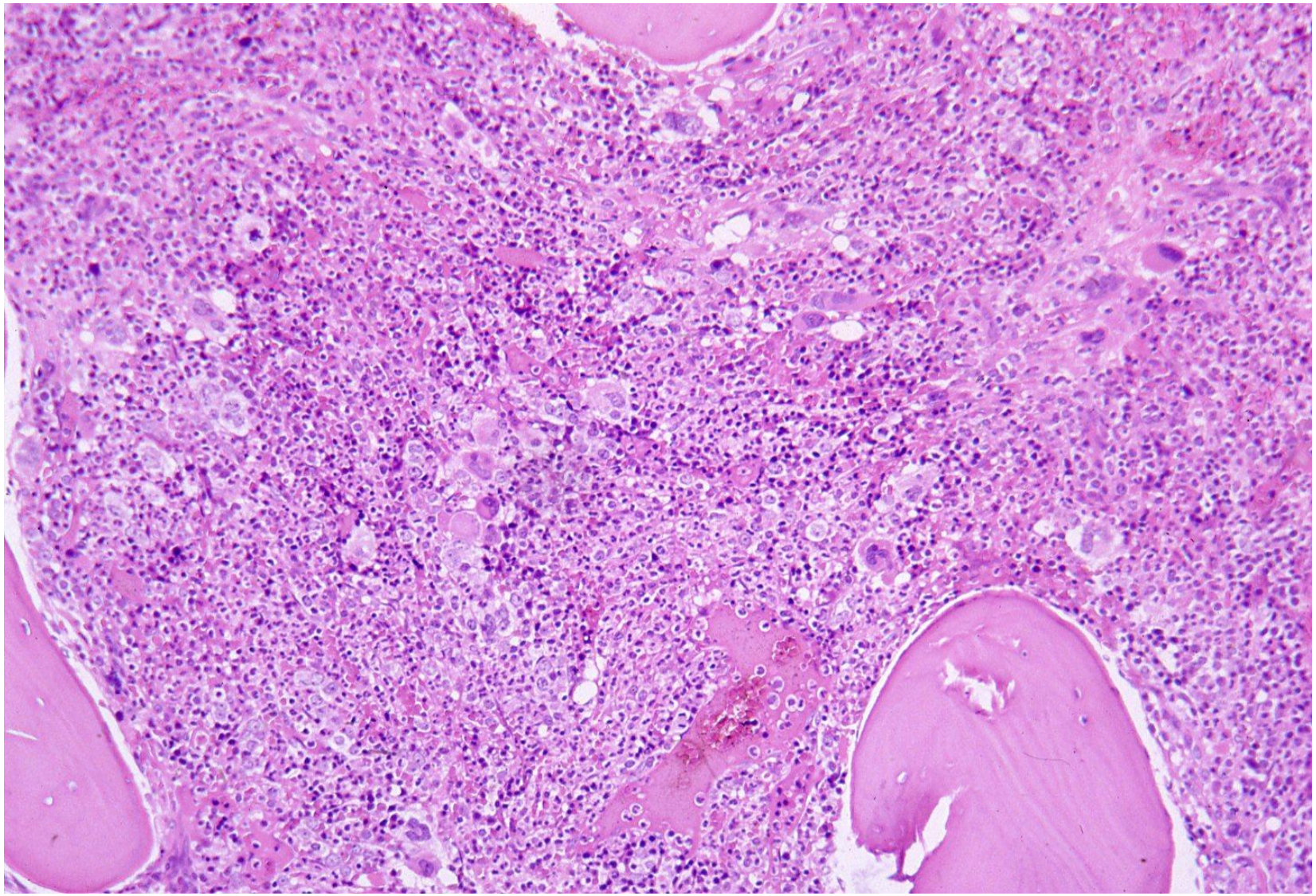
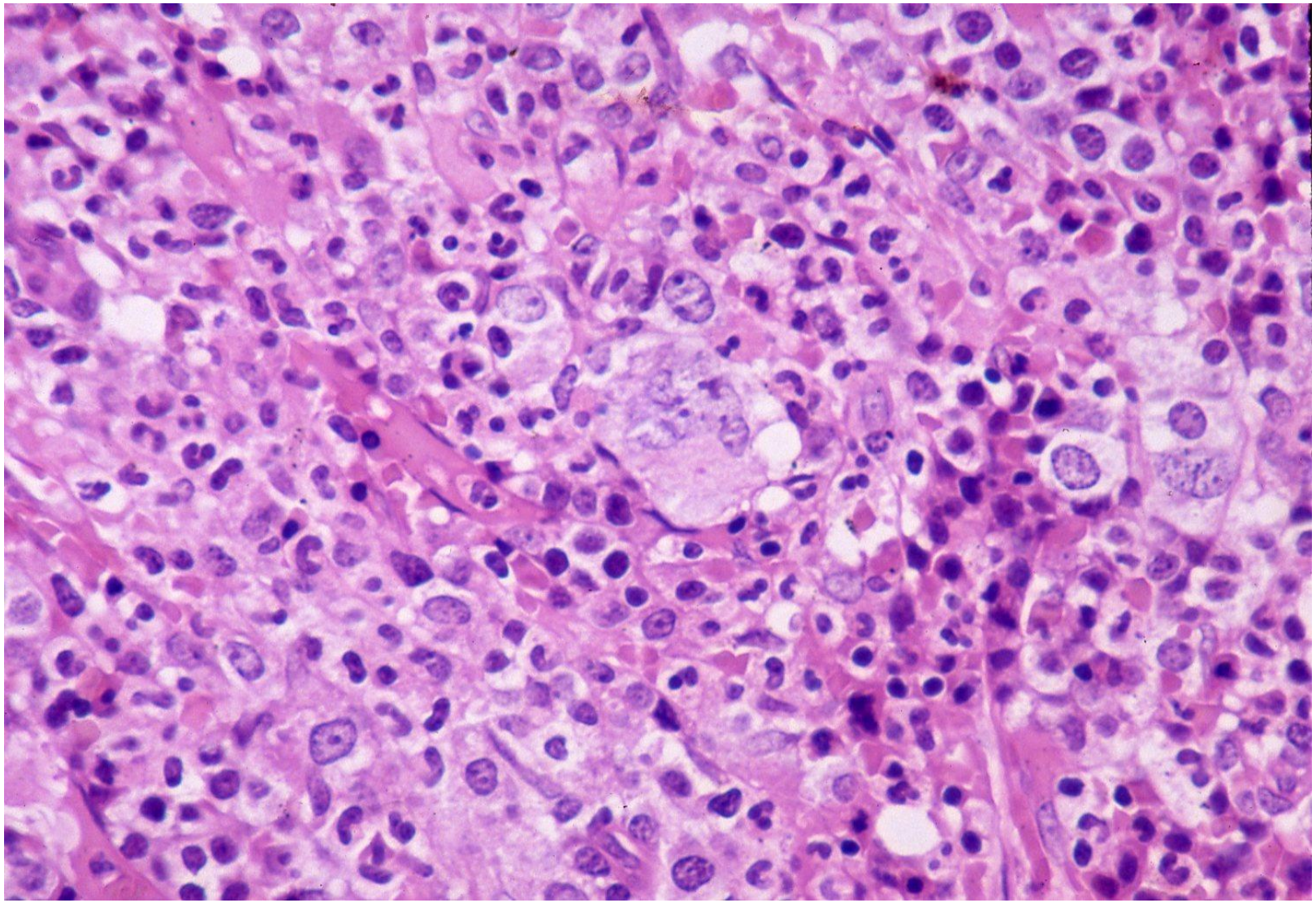
Primary myelofibrosis

Primary myelofibrosis is a myeloproliferative disease featured by neoplastic proliferation of abnormal megakaryocytes and granulocytes in the bone marrow, associated with reactive reticulin fibrous and extramedullary hematopoiesis. The microscopic features are common with chronic myeloid leukemia, polycythemia vera and essential thrombocythemia. Myelofibrosis is caused by neoplastic megakaryocytes releasing platelet derived growth factor, basic fibroblast growth factor, transforming growth factor beta or other cytokines. Osteosclerosis may also be associated. Secondary “sea blue histiocytosis” may be seen. Angiogenesis in the bone marrow and spleen is due to increased serum levels of vascular endothelial growth factor. Teardrop red blood cells and leukoerythroblastosis are provoked by extramedullary hematopoiesis.

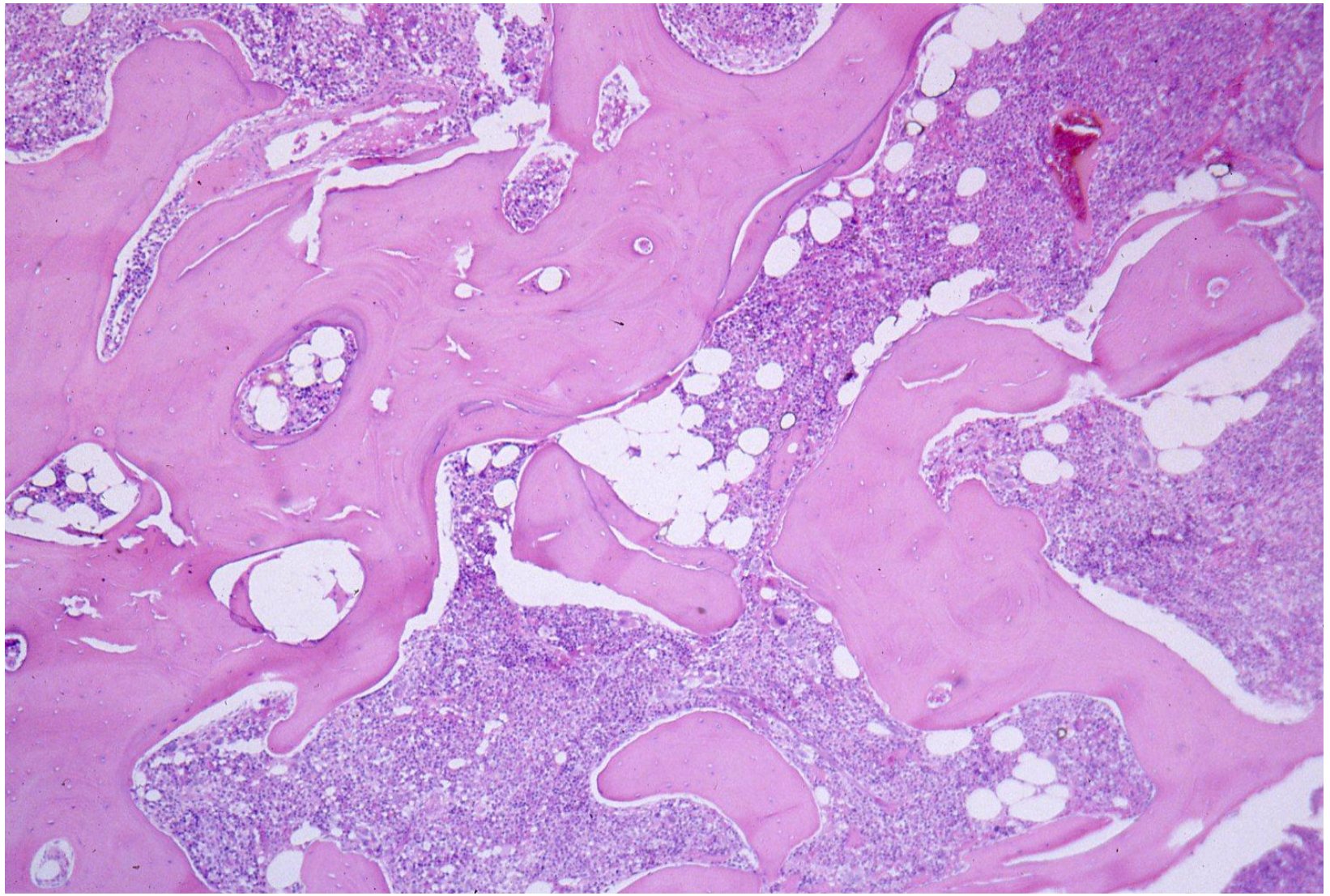
Ref.: Ng ZY, et al. Morphology of myeloproliferative neoplasms. Int J Lab Hematol 2023; 45(Suppl 2): 59-70. doi: 10.1111/ijlh.14086



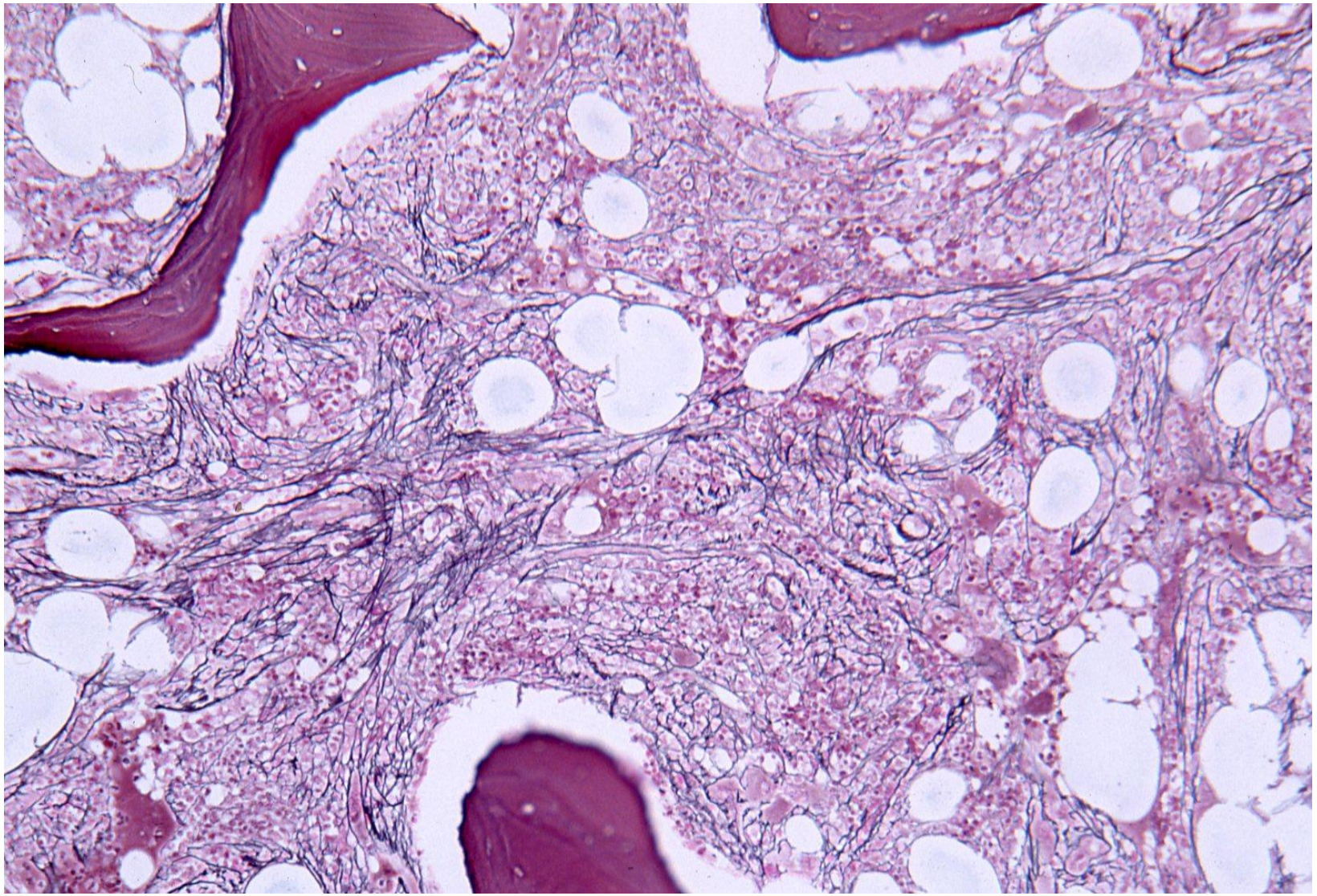
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow biopsy reveals 100% hypercellularity with abnormal increase of megakaryocytes (H&E-1).



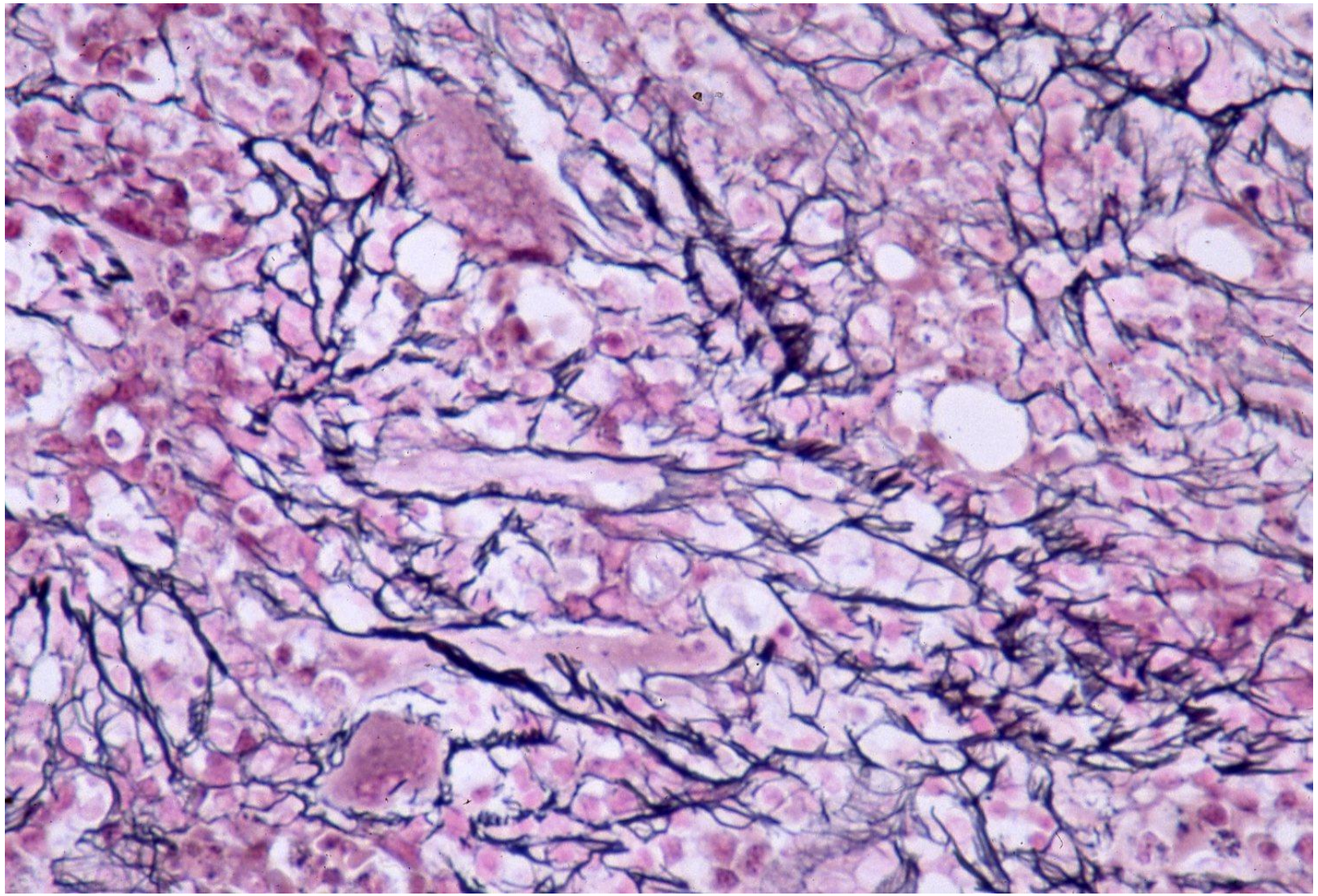
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow biopsy reveals 100% hypercellularity with abnormal increase of megakaryocytes. Myeloid series is also increased (H&E-2).



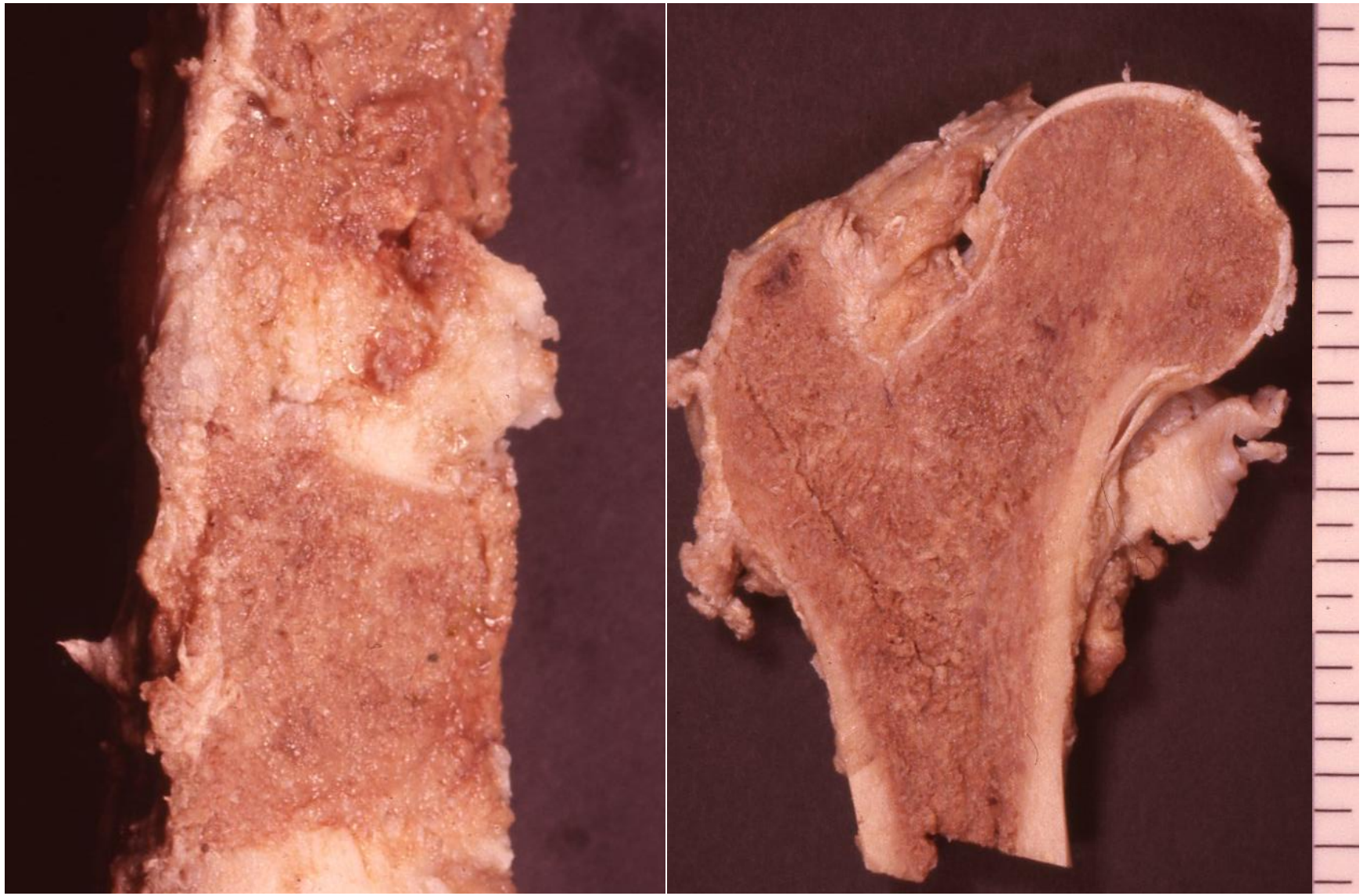
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow biopsy reveals 100% hypercellularity with abnormal increase of megakaryocytes. Osteosclerosis is focally associated (H&E-3).



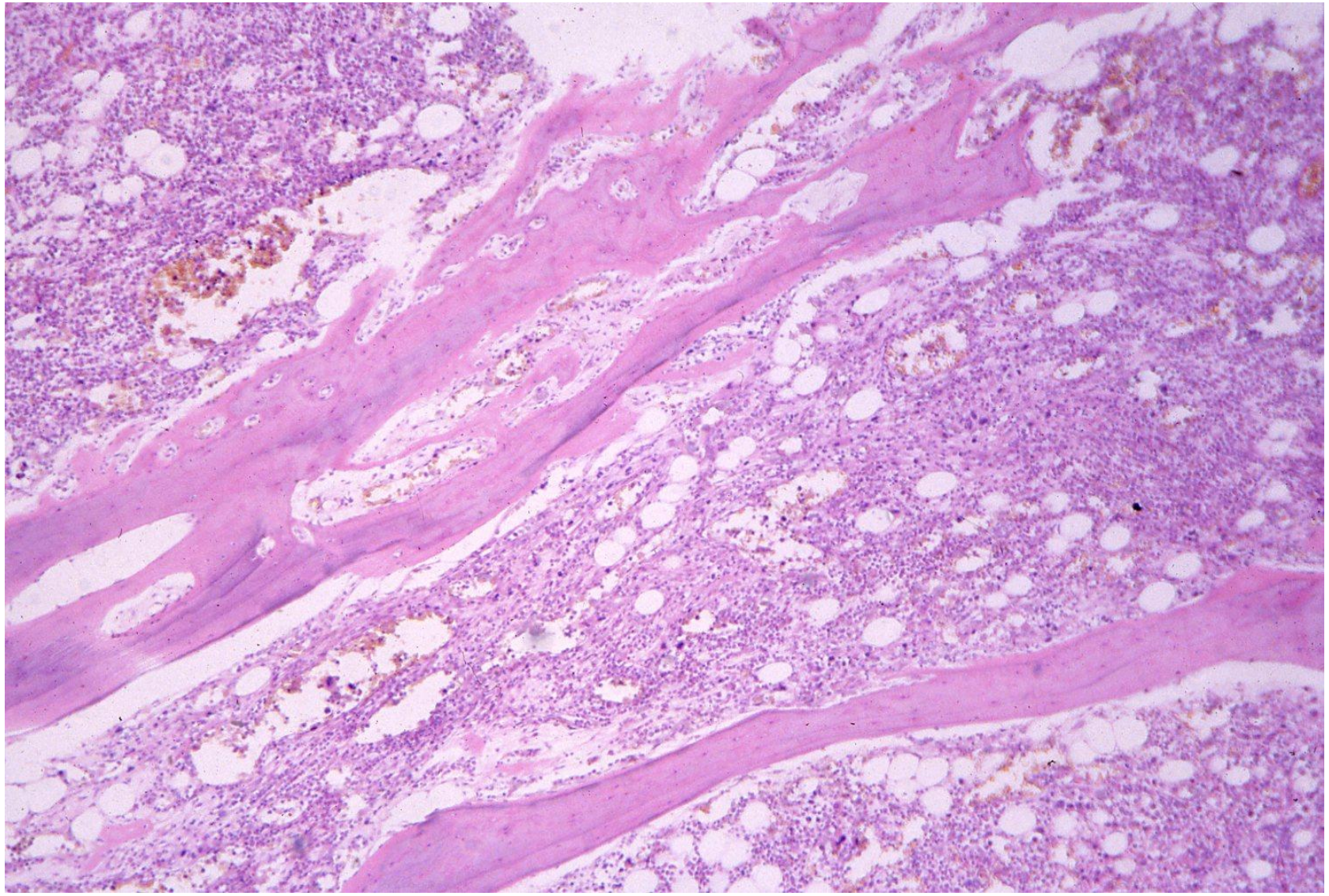
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow biopsy reveals 100% hypercellularity and dense reticulin fibrosis. Bone marrow fails to be tapped (silver impregnation-1).



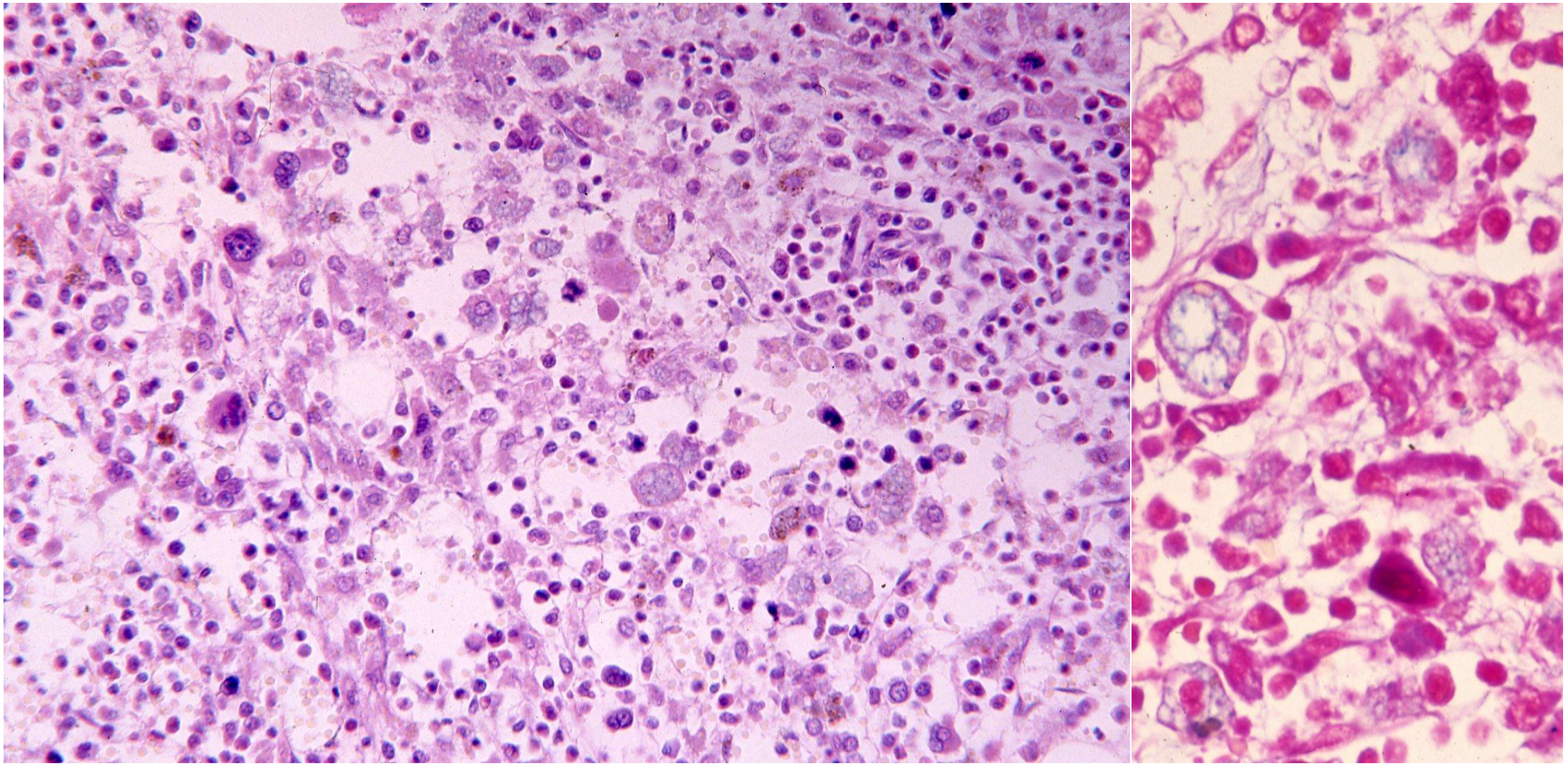
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow biopsy reveals 100% hypercellularity and dense reticulin fibrosis. Bone marrow fails to be tapped (silver impregnation-2).



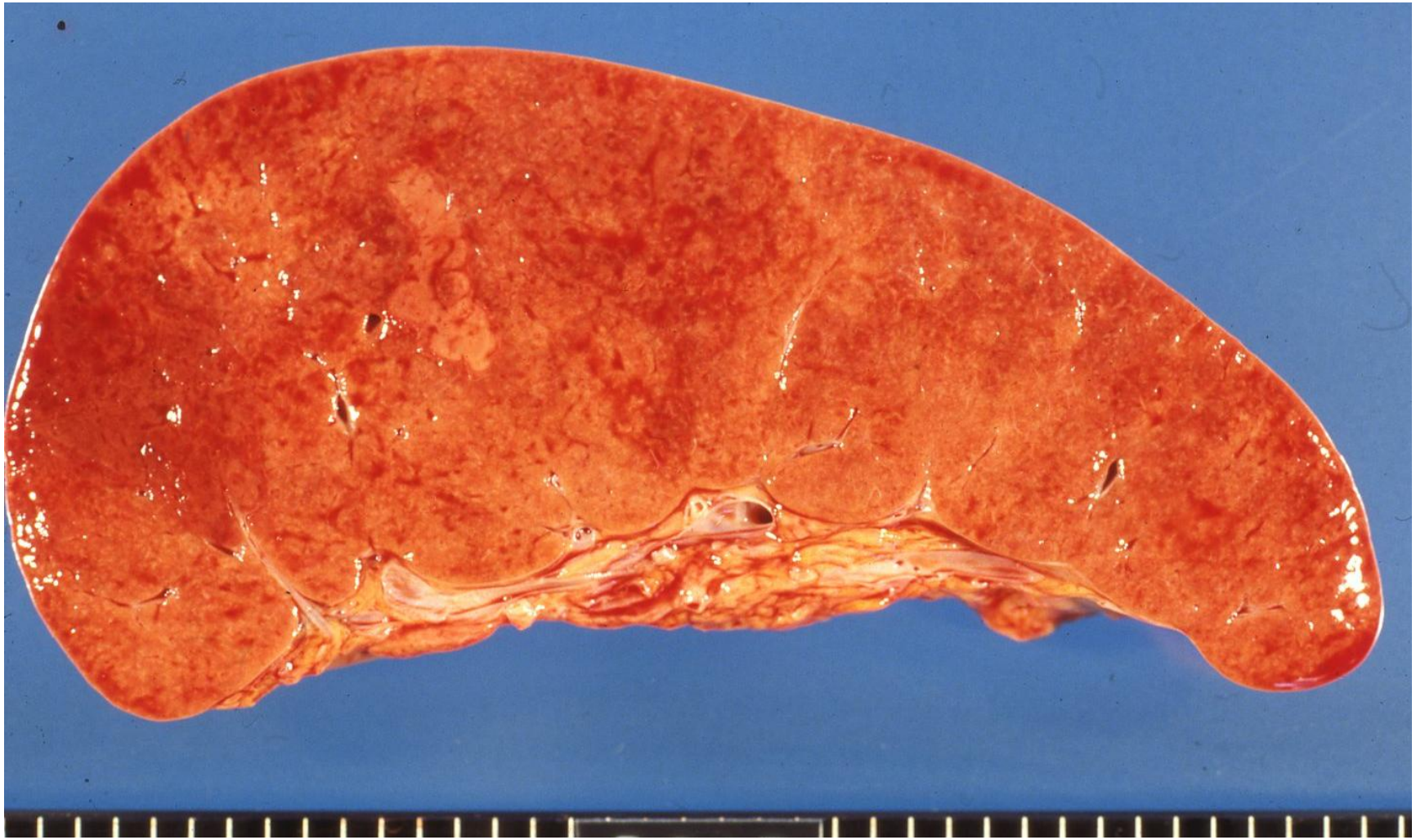
Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, the bone marrow is highly cellular and hard in consistency (left: spine). The femoral bone marrow is totally hematopoietic (right).



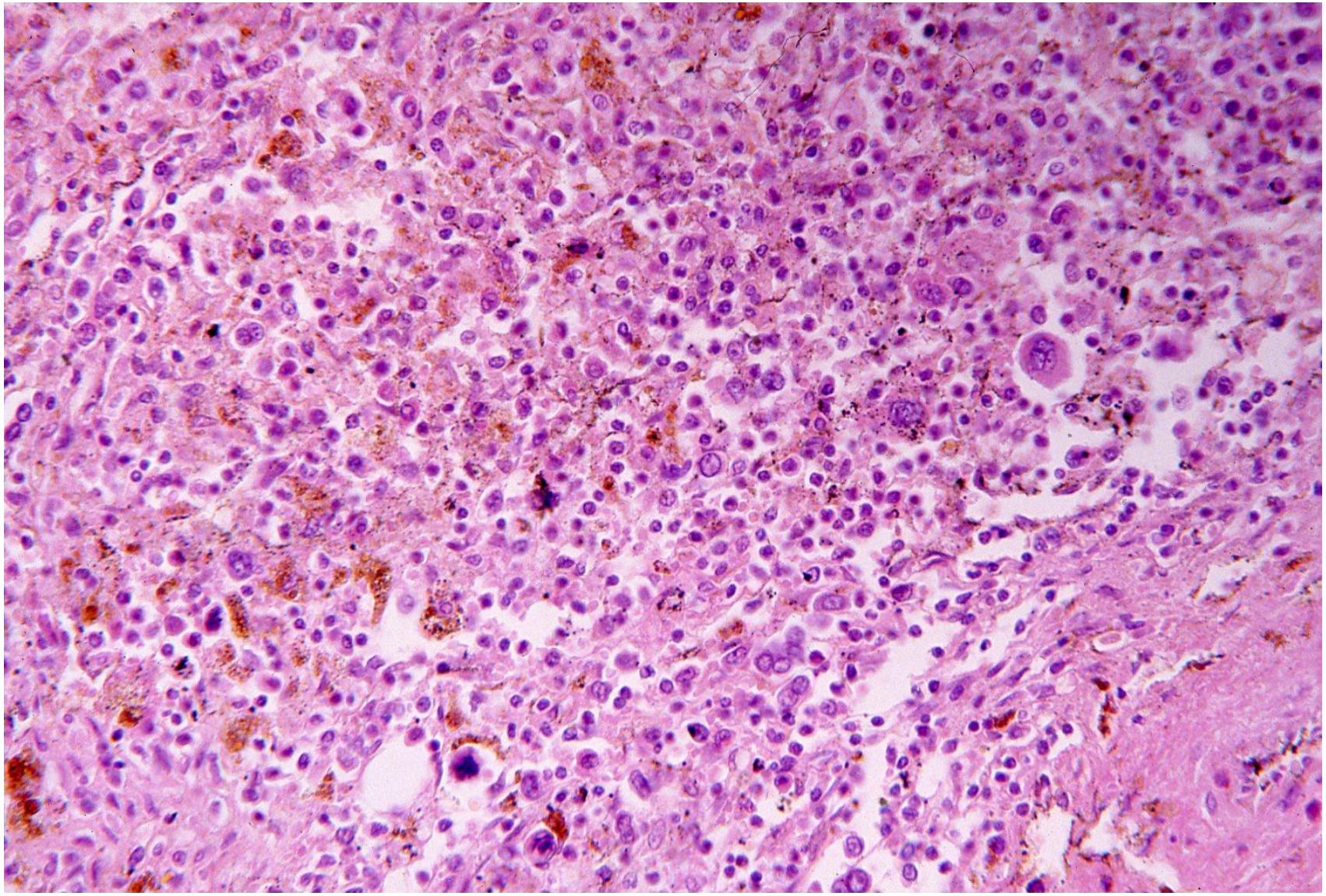
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow at autopsy reveals high-degree hypercellularity with osteosclerosis (H&E-a).



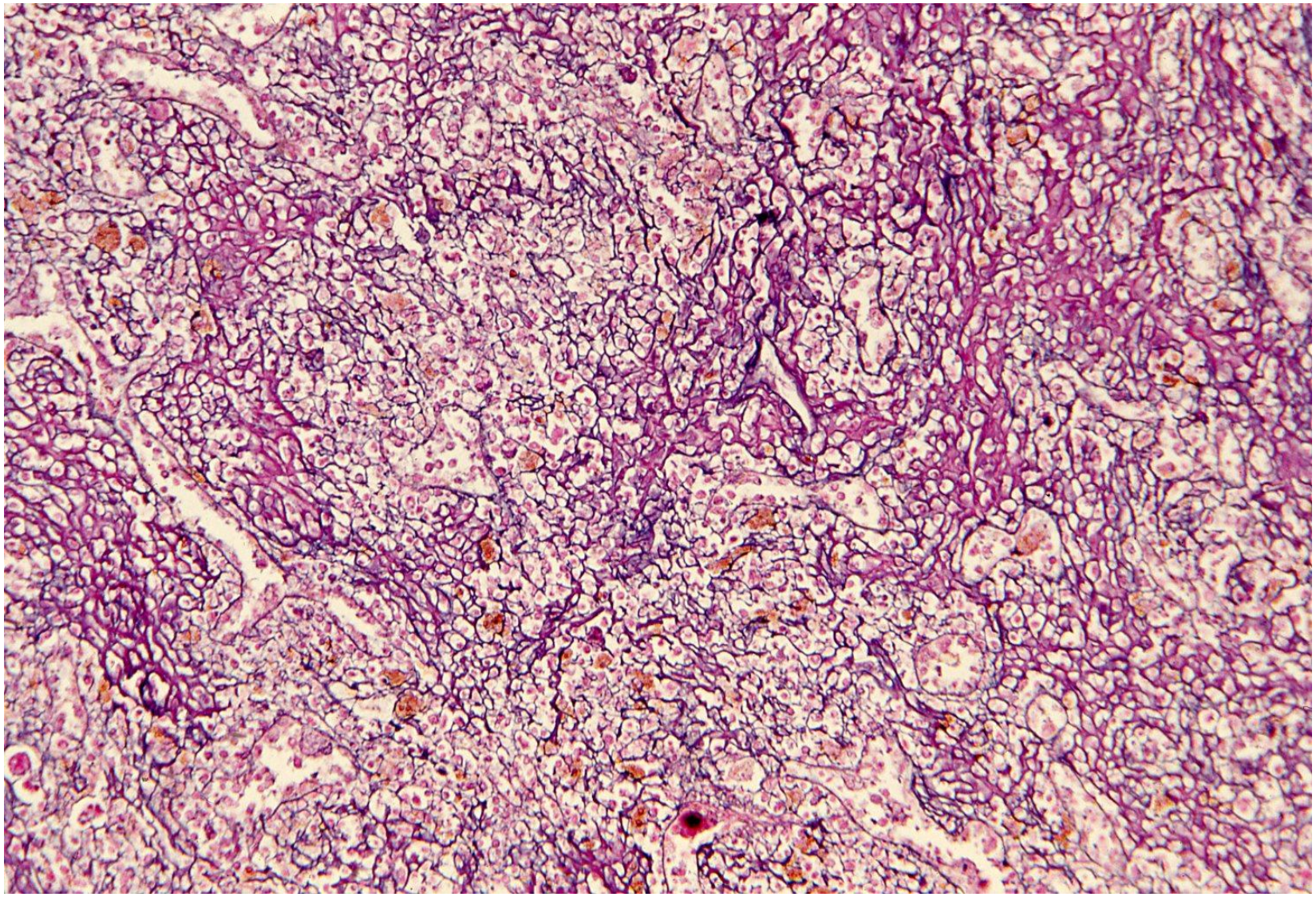
Primary myelofibrosis seen in a 51 y-o female patient. Bone marrow at autopsy reveals hypercellularity. Of note is the distribution of sea blue histiocytes in the marrow space. Myeloproliferative disorders such as AML, CML and MDS may secondarily accompany sea blue histiocytosis (left: H&E-b, right: Azan).



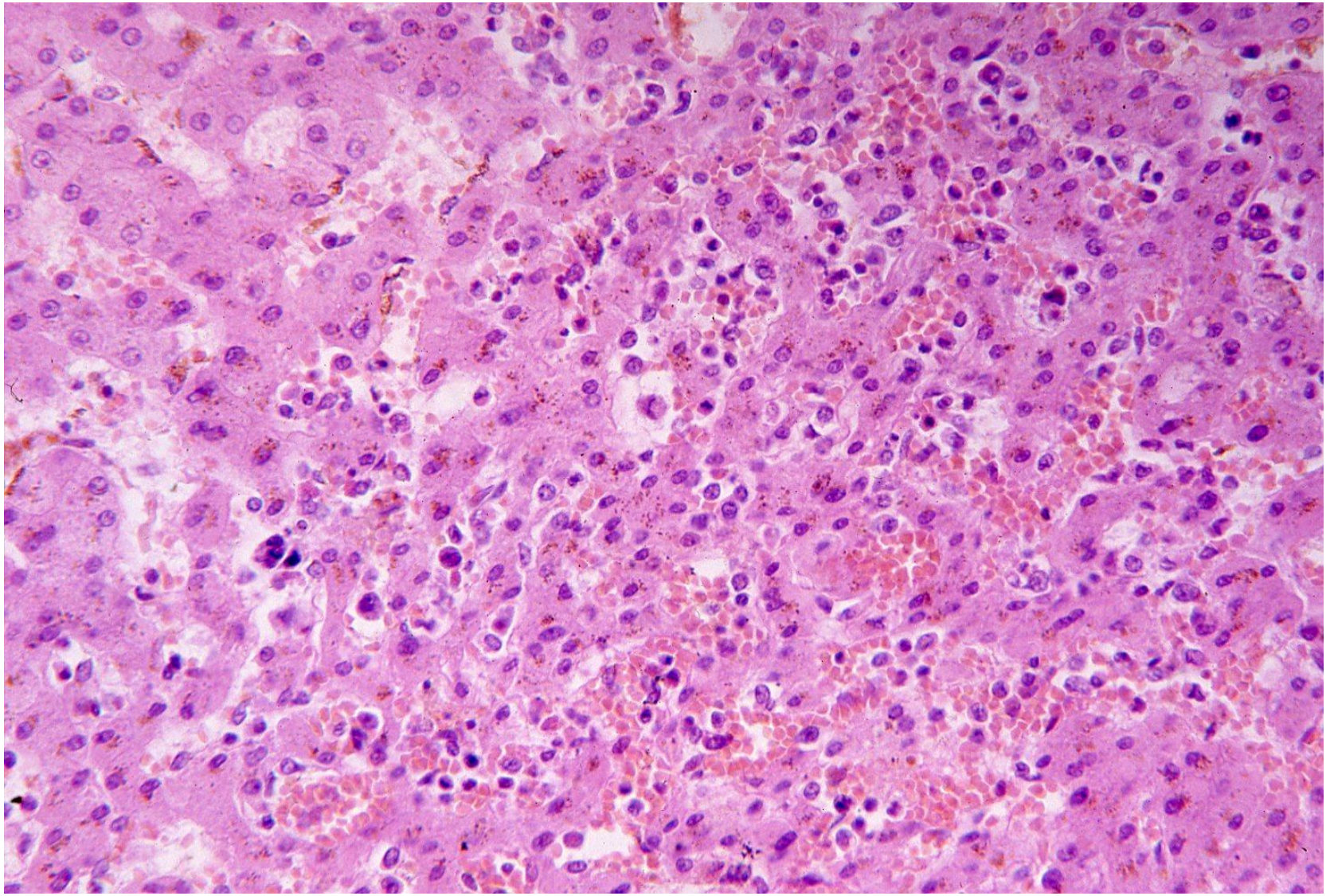
Primary myelofibrosis seen in a 51 y-o female patient. The splenomegaly is characteristic of chronic myeloproliferative disorders, including primary myelofibrosis. At autopsy, the spleen sized 18 cm in length weighs 550 g.



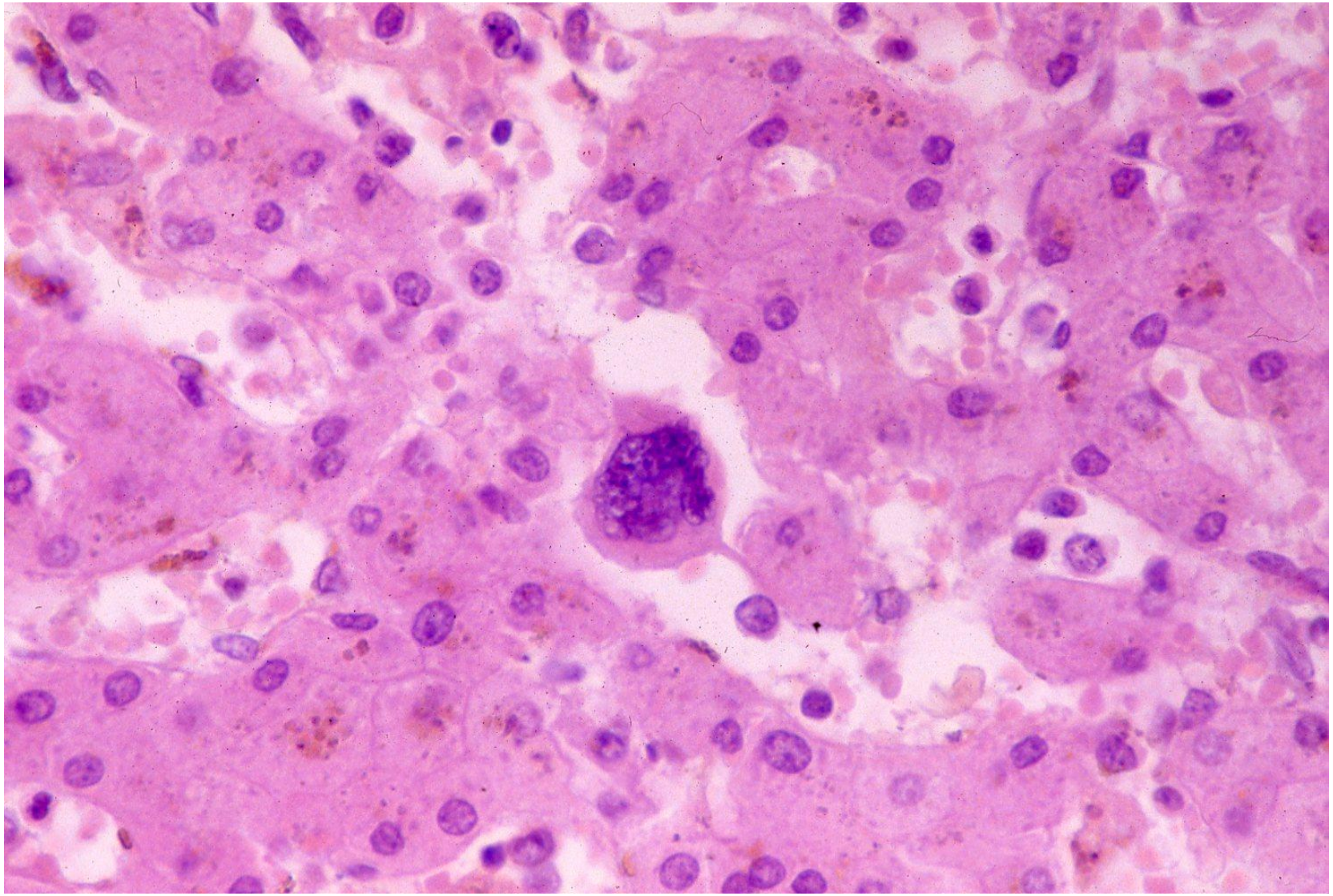
Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, extramedullary hematopoiesis is observed in the red pulp of the spleen. Megakaryocytes are scattered. Hemosiderosis is associated (H&E-c).



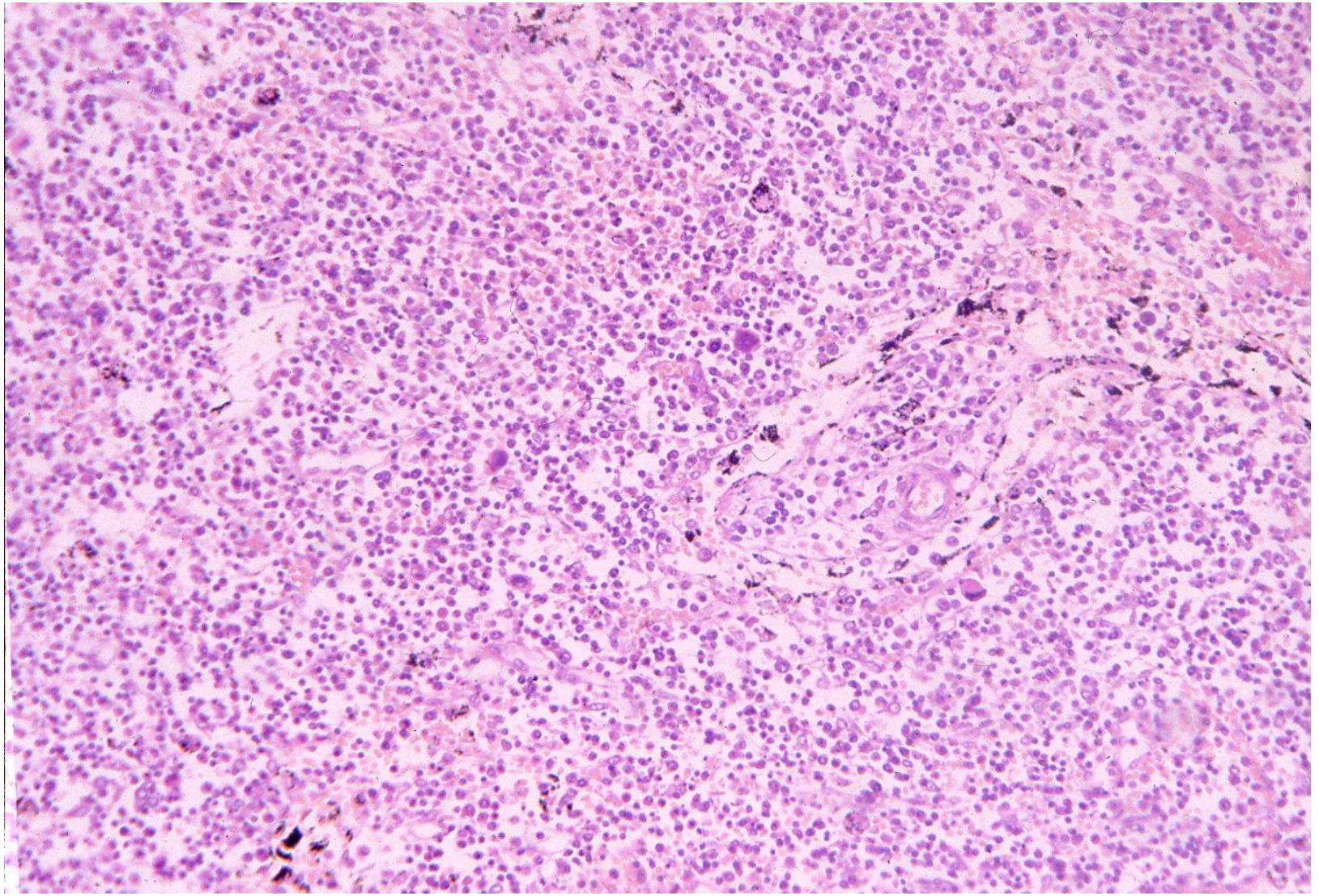
Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, reticulin fibrosis is seen in the red pulp of the spleen, where extramedullary hematopoiesis is observed (silver impregnation).



Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, extramedullary hematopoiesis is observed in the congested sinusoid of the liver. Megakaryocytes and myeloid cells are dispersed (H&E-d).



Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, extramedullary hematopoiesis is observed in the sinusoid of the liver. A megakaryocyte is seen. The hepatocytes are hemosiderotic (H&E-e).



Primary myelofibrosis seen in a 51 y-o female patient. At autopsy, extramedullary hematopoiesis is observed in the parenchyma of the lymph node. Megakaryocytes are dispersed (H&E-f).