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An Unexpected Finding

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Clinical Image

We report a case of a 70 year old woman with previously diagnosed JAK-2 positive myeloproliferative neoplasm meeting morphological criteria for essential thrombocythemia (Figure 1A). Ten months from time of diagnosis she presented with anemia and thrombocytopenia: haemoglobin concentration of 107 g/L (130-180 g/L), white cell count of 46×10^9/L ($4\text{-}11 \times 10^9$/L) and platelet count of 90×10^9/L ($150\text{-}400 \times 10^9$/L). The blood film showed leucoerythroblastic features. Clinically the patient had ascites and weight loss. The blood film changes and bicytopenia raised the possibility of progression to post-ET myelofibrosis. A repeat marrow was performed. Bone marrow aspirate was blood dilute but otherwise non-diagnostic. The trephine was markedly hyper cellular with clusters of hyperlobated megakaryocytes in keeping with previously diagnosed myeloproliferative neoplasm. In addition, marrow spaces contained sheets of spindle shaped cells (Figure 1B). On a background of markedly increased reticulin (MF3) suggestive of progression to myelofibrosis (Figure 1C) further immunohistochemical staining of the spindle cells on the trephine with CD31 (Figure 1D) as well as CD34 and vimentin revealed a network of anastomosing irregular vascular channels confirming the diagnosis of metastatic angiosarcoma. The patient progressed rapidly and on autopsy had angiosarcoma in the liver and spleen.

There are only few case reports of angiosarcoma with bone marrow involvement and there appears to be a unique association with primary tumor of the spleen. This case illustrates the challenge of distinguishing the angiosarcoma from the possibility of more likely secondary myelofibrosis in the context of pre-existing ET.

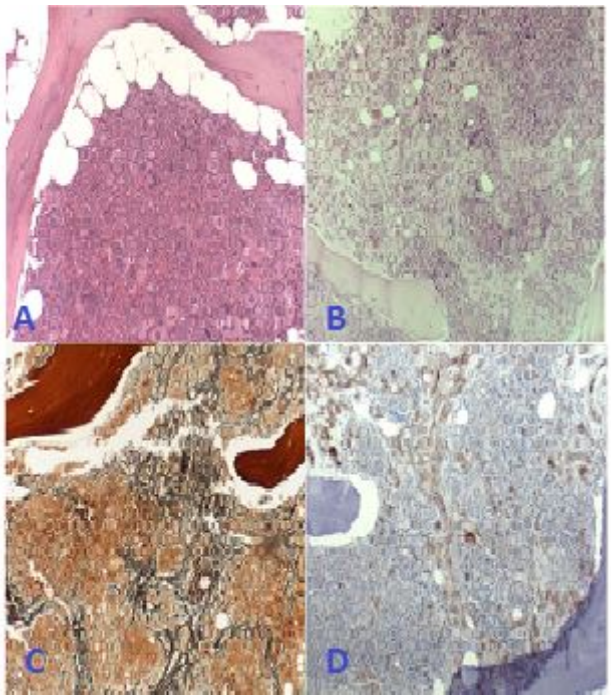

Figure 1: A: HE stain X10; B: HE stain X10; C: Reticulin stain X10; D: CD31 Stain X10.