A middle-aged man presented with vague abdominal and chest pain. Laboratory results suggested anemia. Computed tomography of the chest and abdomen revealed mediastinal lymphadenopathy, sclerotic bone lesions, hypointense liver masses, and pancreatic body/tail mass. Left iliac bone biopsy revealed patchy bone marrow involvement by cords of atypical cells with eccentrically placed nuclei and plump eosinophilic cytoplasm (Figure A and B), associated with extensive “stromal injury” and residual trilineage hematopoeisis. The differential diagnosis included both hematolymphoid and non-hematolymphoid neoplasms. The concurrent liver biopsy revealed tumor cells with similar morphology but were embedded in a prominent myxohyaline stroma, indicating that the presence of this distinctive myxohyaline stromal matrix was previously interpreted as secondary stromal injury in the bone marrow. The overall findings raised consideration for a vascular neoplasm, and immunohistochemistry revealed positive staining for vascular endothelial markers including CD31, ETS-related gene (ERG) (Figure C), friend leukemia integration 1 (FLI1), and focal pankeratin. Calmodulin-binding transcription activator 1 (CAMTA1) exhibited diffuse nuclear and cytoplasmic staining in the tumor cells (Figure D), suggestive of WWTR1-CAMTA1 fusion (for expansion of gene symbols, use search tool at www.mayoclinicproceedings.org).

FIGURE. Light microscopy revealing cords of epithelioid endothelial cells in a background of myxohyaline stroma involving the bone marrow (A and B, hematoxylin and eosin). Immunostains were positive for ETS-related gene (ERG) (C, antibody clone: EPR3864; commercial source: Ventana) and calmodulin-binding transcription activator 1 (CAMTA1) (D, antibody clone: NBP1-93620; commercial source: Novus Biologicals). CAMTA1 positivity is suggestive of WWTR1-CAMTA1 fusion. Magnification: (A, C, and D) ×200; (B) ×600.
Epithelioid hemangioendothelioma is a rare vascular neoplasm with an estimated prevalence of less than 1 per 1,000,000 individuals.¹ It has a heterogeneous presentation from localized to widespread metastatic disease.¹ About 90% of cases harbor WWTR1-CAMTA1 fusions and 10% YAP1-TFE3 fusions. Epithelioid hemangioendothelioma involving bone marrow is extremely rare and presents with cytopenias, mimicking hematopoietic neoplasms.² Given the lack of standardized treatments, management should be pursued in sarcoma reference centers by a multidisciplinary team.¹

POTENTIAL COMPETING INTERESTS
The authors report no competing interests.

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