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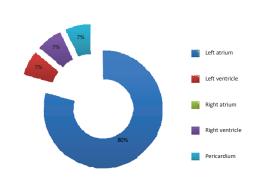
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The heart of the matter: A unique convergence of cardiac neoplasm, hereditary nonpolyposis colorectal cancer, and spindle cell sarcoma

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Primary cardiac tumors are exceedingly unusual and aggressive; they often develop in younger patients and present with advanced disease. The rarity and heterogeneity of primary cardiac tumors challenge the standardization of therapeutic guidelines. Undifferentiated primary cardiac spindle cell sarcomas, a distinct subset of primary cardiac sarcomas, are especially unique with fewer than 20 cases reported worldwide—the majority of which are of left atrial origin. We present a review of the etiology, pathophysiology, and therapy of undifferentiated primary cardiac spindle cell sarcomas. In conjunction, we present a unique case of a woman with hereditary nonpolyposis colorectal cancer (Lynch syndrome) who presented with a primary cardiac spindle cell sarcoma



Anatomic Distribution of Primary Cardiac Undifferentiated Spindle Cell Sarcoma

of left ventricular origin, the first case of this type and location of cardiac tumor reported in a patient with Lynch syndrome. While some malignancies are more common in patients with Lynch syndrome, sarcomas are not one of them. The absence of metastases at the time of diagnosis is atypical for left-sided cardiac sarcomas, the overwhelming majority of which have metastases at the time of diagnosis. In addition to anatomic and pathophysiologic distinctions of this case from other primary cardiac spindle cell sarcomas, it also demonstrated unique immunohistochemistry as the first reported case of MDM-negative (murine double minute homolog, a principle diagnostic marker of spindle cell sarcoma) ever reported.

Biography

Emily Bryer is a physician in Philadelphia, Pennsylvania USA. She attended the Schreyer Honors College of The Pennsylvania State University for her undergraduate training where she served as President of Global Medical Brigades and established medical clinics in areas of Ghana, Honduras, Nicaragua, and Panama with limited access to medical care and healthcare resources. She received her medical degree from The Philadelphia College of Osteopathic Medicine before starting her Internal Medicine residency at Pennsylvania Hospital of the University of Pennsylvania. Her research interests include venous thromboembolism in chemotherapy-induced anemia and gestational trophoblastic disease.

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