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Pulmonary artery intimal sarcoma: A review of the literature and case report

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Statement of the Problem: Pulmonary artery intimal sarcoma (PAIS) is a rare, aggressive tumor often diagnosed at autopsy. Presentation often involves the main pulmonary arteries and pulmonary valves with resultant pulmonary embolism and chronic thromboembolic pulmonary hypertension leading to the misdiagnosis of pulmonary embolic disease. Due to the paucity of data on the clinicopathologic features of disease in PAIS, the aim of this study was to combine a clinical case report with comprehensive review of literature with meta-analysis to increase awareness and facilitate clinical recognition of PAIS.

Methods: Clinical information was collected regarding tumor site, size, location, extension, patient presentation signs and symptoms, imaging data, disease recurrence, distant metastasis, and overall survival. Progression-free survival, distant-metastasis free survival, and disease-specific survival curves and estimates were calculated using the Kaplan-Meier method with univariate and multivariate analyses of prognostic variables calculated using Cox proportional hazards model.

Results: Kaplan-Meier survival plots did not indicate statistically significant correlations between mean survival and tumor location; however, metastasis and tumor recurrence showed statistical significance in correlation with mean survival time (P=0.21, P=0.31 respectively). Chi-square values indicated tumor location in the pulmonary valve and aorta were significant factors in mean survival time (P=0.004, P=0.23 respectively), as well as metastasis and tumor recurrence (P=0.003, P=0.001 respectively). The clinical case is of a 53-year-old woman with recurrent pulmonary emboli, claudication of legs, dyspnea and hemoptysis whom was later discovered to have a large (12.0x6.5x5.0 cm) intravascular lung pulmonary artery intimal sarcoma with satellite pulmonary metastases. Tumor cells showed immunohistochemical phenotypic expression of MDM2 and fluorescence in situ hybridization amplification of MDM2.

Conclusion & Significance: PAIS is a rare tumor type, clinicians treating patients for suspected PE that is refractory to anti-coagulant or thrombolysis therapy should have a high level of clinical suspicion for PAIS to improve outcomes.

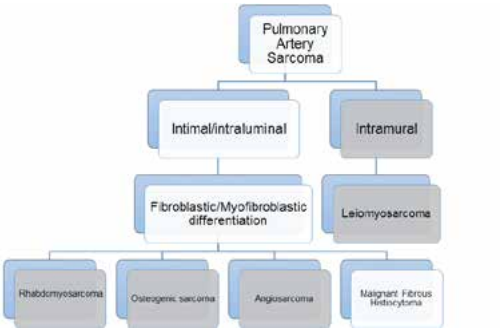

Figure 1: PAS is subdivided into intimal and intramural sarcomas. Intimal sarcomas are further classified as fibroblastic/myofibroblastic differentiation with several different subtypes. The four most common are listed here. In this paper, we focus on malignant fibrous histiocytomas, also referred to as high grade spindle cell sarcoma, or undifferentiated pleomorphic sarcoma (UPS).

Biography

Manjit K Bhandal is a second year MD student at Central Michigan University College of Medicine. Her primary interest in medicine is Cardiology in which she has several years of pre-clinical experience and is now conducting several studies in various aspects of the field. These studies include the present cardio-oncology study, outcomes of femoral artery catheterization in patients with US versus palpation techniques, as well as studying the outcomes of diabetic heart failure patients implanted with CardioMEMS.

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