Olena Dobenska Mary N Sheppard

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Cardiac Sarcoma

Background and methods.

Cardiac sarcomas are extremely rare primary malignant tumours of the heart. Recent research focuses on cardiac intimal sarcomas arising from the left side of the heart as the most common cardiac sarcoma types. This study aims to provide clinico-pathological correlations of the reported cardiac intimal sarcomas exemplified by an unusual case of the right atrial/extracardiac intimal sarcoma with divergent differentiation, diagnosis of which was supported by molecular studies.

Results.

47y.o. man, presented with acute dyspnoea, was found to have large pericardial effusion and a right atrial mass on routine transthoracic echocardiography. Cardiac magnetic resonance imaging showed 59mm mass with heterogeneous enhancement involving the right atrium in connection with 45mm extracardiac mass adherent to inferior cardiac surface in pericardial space. The patient underwent resection of the mass. Several lobulated haemorrhagic and white solid tumour masses were submitted for histological examination, which revealed high grade pleomorphic sarcoma with a variety of morphological features, including foci of floret cell rhabdomyosarcoma with cambian layer and strong nuclear expression of desmin and myogenin. Angiosarcoma was suspected on morphology, but not supported by immunohistochemistry, including ERG expression. CDK4 immunohistochemistry was equivocal and p16 was positive. FISH revealed MDM2 gene amplification at 12q15, supporting the diagnosis of an intimal sarcoma. Due to complex involvement of the heart the tumour was not completely excised.

The patient was treated with chemotherapy and is well at 4 months after the diagnosis.

Conclusions.

Cardiac sarcomas often present late without overt symptoms, precluding effective tumour eradication. Neoadjuvant therapy targeting MDM2 or PDGFRA if attempted prior to excision may facilitate achievement of complete resection. These rarities should be targeted by 100KGP for better understanding of the pathology.