A Case of Primary Pericardial Synovial Sarcoma

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Background
- Primary pericardial malignancies are rare causes of pericardial effusion and tamponade.
- Of these malignancies pericardial sarcomas are a group of related malignancies of which several histologic subtypes exist.
- We describe here a case of primary pericardial synovial sarcoma in an otherwise healthy young male and the diagnostic and clinical dilemmas encountered during his care.

Patient History/Presentation
- 29 yo obese M patient who presented initially with large pericardial effusion. CT chest with no evidence to suggest malignancy.
- Underwent pericardiocentesis. Cytology and flow cytometry showed no evidence of malignancy. Thought potentially to be due to recent COVID infection.
- 6 months later seen for repeat TTE which showed accumulation of pericardial effusion (Fig 1) in addition to effusive-constrictive physiology.
- Sent to ED and ultimately admitted with concern for cardiac tamponade.

Exam
- BP 120/90 HR 90 Respiratory 97% on RA
- Bibasilar rales and moderate LE edema
- Pulsus paradoxus present with 15mmHg drop in BP with inspiration

Hospital Course/Treatment Plan
- CMR ordered due to concern for malignancy with constriction and high density effusion on TTE
- Following CMR, attempted VATS biopsy, converted to open biopsy and pericardial window. Intrapericardial contents covered by "lobulated gel like mass"
- Biopsy consistent with high grade synovial sarcoma with SS18 Gene mutation

Patient Outcomes
- Planned for doxorubicin based chemotherapy at 6 weeks from thoracotomy followed by photon beam radiation
- Patient had two subsequent admissions for diuretic resistant volume overload
- Ultimately transitioned to comfort care following first round of chemotherapy

Clinical Implications
- Primary pericardial tumors can be missed on standard evaluation for pericardial effusions.
- A high index of suspicion and close surveillance is required to rule out life threatening etiologies of recurrent effusion such as neoplasm.
- Recurrent or atypical presentations of pericardial effusions warrant further evaluation with CMR to characterize the process and assess the need for biopsy.

Disclosures: The authors listed above have no relevant disclosures.