A Rare Case of Cardiac Myxofibrosarcoma

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Background

- Primary cardiac tumors are rare, with an incidence of 0.001-0.3% [1].
- 25% of cardiac tumors are malignant with angiosarcoma being the most common subtype [2].
- Cardiac myxofibrosarcomas are exceptionally rare and aggressive malignant tumors with poor prognosis [2].
- Embryologically, myxofibrosarcomas arise from mesenchyme and can be found in the atria, ventricles and vessels. Patients tend to present with symptoms when the tumor is large enough to obstruct blood flow [3].

Patient Presentation

A 66 year-old-man initially presented to the emergency department with dyspnea, chest pain, and palpitations. He was found on transthoracic echocardiography (TTE) to have a large multilobed left ventricular (LV) mass protruding into both the LV outflow tract and mitral valve (MV) orifice during contraction. He underwent R2 resection of the mass with pathology revealing a high-grade myxofibrosarcoma with immunochemistry positive for Actin (SMA), CD-31, and KI-67 40%. The patient was readmitted 2 months later with symptoms of heart failure. A repeat TTE demonstrated recurrence of the LV mass and an ejection fraction (EF) of 28% with LV outflow tract obstruction. Staging CT scans revealed thoracic spine and pelvic osseous metastasis.

Exams and Imaging

Figure 1: Cardiac MRI demonstrated a large 4.7 x 4.5 cm intraluminal mass attached to the lateral wall of the LV.

Treatment Plan

After a multidisciplinary discussion involving Oncology and Cardiothoracic Surgery, a decision was made to proceed with chemotherapy as opposed to surgical resection given high intra-operative risks and high risk of recurrence of the tumor. He was started on a chemotherapy regimen of gemcitabine/docetaxel as opposed to an anthracycline-based regimen given his reduced EF.

Patient Outcome

In summary, this patient with primary myxofibrosarcoma complicated by recurrence and osseous metastasis after R2 resection has had a good response thus far on gemcitabine/docetaxel. He has now completed 4 cycles of chemotherapy with his most recent CT scan showing shrinkage of the mass from 8.7 to 5.6 cm. His performance status has improved and he receives close follow up.

Discussion

Primary cardiac myxofibrosarcomas have an average survival of approximately 11 months owing to their highly aggressive and metastatic nature [4]. Early detection is ideal, and surgery is the first-line treatment. But following surgery, approximately 43% of patients present with local recurrence and 19% with distant metastasis [4]. Complete resection may lead to nearly twice the life expectancy compared to incomplete excision [5]. While adjuvant chemotherapy and radiation may be helpful, regular long-term follow up with imaging is necessary to detect recurrence.

References


Disclosure

No authors have financial conflicts of interests to report.