The Rarest of the Rare: A Case of Primary Cardiac Osteosarcoma

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INTRODUCTION

- Most cardiac tumors are metastatic tumors, which are 20–40 times more common than primary tumors of the heart.
- Primary cardiac tumors are extremely rare with an incidence of less than 0.1 percent.
- Virtually all types of sarcomas have been reported in the heart as isolated case reports. We present a rare case of biatrial high-grade osteosarcoma.

CASE DESCRIPTION

- A 54-year-old Hispanic female presented with shortness of breath and was cyanotic on the exam while visiting Mexico.
- Due to abnormal chest x-ray, echocardiogram concerning for bilateral atrial myoma she was referred to a cardiothoracic surgeon.
- She underwent bi atrial intracardiac tumor resection in Mexico.
- Post resection staging PET-CT showed hypermetabolic mixed lytic and sclerotic lesion of T10 concerning for metastasis disease.
- Surgical pathology showed extensive undifferentiated spindle cell proliferation with multifocal osteoid production and foci of osseocartilaginous differentiation. This is consistent with a primary cardiac high-grade osteosarcoma.

CASE DESCRIPTION Contd.

- She received approximately 6 cycles of adriamycin and ifosfamide chemotherapy. Adriamycin was discontinued due to left ventricular dysfunction with an ejection fraction of 30-35%, multiple segmental abnormalities, diffuse left ventricular hypokinesis, and moderate to severe mitral valve regurgitation.
- A follow-up PET-CT five months after cessation of treatment reveals no significant evidence of uptake other than abnormalities in the T10 vertebra.

PATHOLOGY

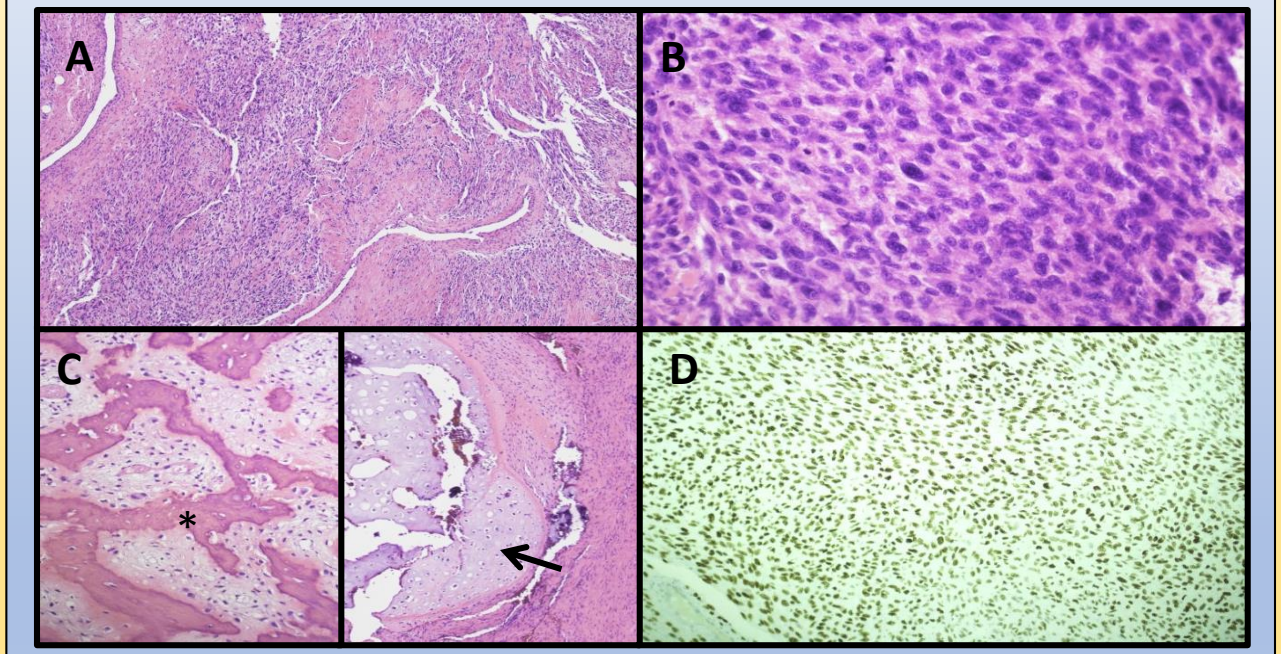

Figure 1. H&E stain at 40x magnification showing a high-grade spindle cell neoplasm infiltrating cardiac muscle **(A)**. At 400x magnification the tumor is composed of atypical ovoid cells with inconspicuous cell borders, eosinophilic cytoplasm, hyperchromatic nuclei, prominent nucleoli, and scattered mitoses **(B).** Osteoblastic (asterisk) and chondroblastic (arrow) foci are seen **(C)**. Tumor cells show strong, diffuse, nuclear positivity with SATB2 immunohistochemical stain, supporting the diagnosis of osteosarcoma **(D)**.

DISCUSSION

- Osteosarcomas are aggressive with a high incidence of recurrence and metastasis. Fewer than 50 cases of primary cardiac osteosarcomas have been reported in the literature.
- Currently, it is postulated that they arise from undifferentiated mesenchymal stem cells in the endocardium that transform into active osteoblasts.
- Even though complete resection can be achieved in some cases, long-term results are usually poor. No standard therapy has been established due to the tumor's low incidence rate and lack of clinical trial data.
- Our case highlights the importance of evaluating common symptoms thoroughly since it may be a harbinger of rare and serious disorders.
- This case reflects the heterogeneous nature of sarcoma histology, the consequent tumor biology and hence varied clinical course and prognosis.

REFERENCES

- Primary osteosarcoma of the heart: experience of an unusual case.Karagöz Özen DS, Oztürk MA, Selcukbiricik F, Esatoglu SN, Turna ZH, Beyaz P, Dervisoglu S, Ozgüroglu M.Case Rep Oncol. 2013 Jan; 6(1):224-8. Epub 2013 Apr 19.