Right Ventricle Outflow Tract Intimal Sarcoma Extending into the Main Pulmonary Artery: A Case Report

Rashid AL Umairi[1], Khadija AL Adwai[1], Nahid AL Qanubi[2], Ruqaiya AL Shamsi[3], Adel Mohamed Kamel[4] and Uday Arun Gokhale[3]

[1]Radiology Department, The Royal Hospital, Muscat, Oman
[2]Radiology Residency Training Program, Oman Medical Specialty Board, Muscat, Oman
[3]Histopathology Department, The Royal Hospital, Muscat, Oman
[4]Cardiology Department, The Royal Hospital, Muscat, Oman

Received: 10 January 2021
Accepted: 16 May 2021

*Corresponding author: alumairi1@yahoo.com

DOI 10.5001/omj.2022.28

Abstract

Intimal sarcomas are rare malignant mesenchymal tumors that arise in the large arteries and very rarely in the heart. Herein we report a 55-year-old man who was referred for further assessment of a Right Ventricle Outflow Tract (RVOT) mass. Assessment with cardiac MRI revealed an enhancing mobile mass arising within the RVOT and extending into the main pulmonary artery. The initial diagnostic possibilities included thrombus and myxoma. The patient underwent surgical recession of the mass and the histopathology examination confirmed the diagnosis of cardiac intimal sarcoma.

Introduction

Primary cardiac tumors are rare with an estimated incidence of approximately 0.3-0.7% in surgery and autopsy reports.[1] Approximately 25 % of primary cardiac tumors are malignant.[2] Angiosarcomas and undifferentiated pleomorphic sarcoma were previously thought to represent the most common primary malignant cardiac tumors. However, analysis of 100 primary cardiac sarcomas using fluorescence in situ hybridization (FISH) for mouse double minute 2 (MDM2) amplification, as a classification parameter, has suggested that intimal sarcomas are the commonest primary cardiac sarcomas (42/100 cases).[3]

Cardiac intimal sarcomas are frequently misdiagnosed as benign myxoma and thrombus. Cardiac MRI (CMR) is a non-ionizing imaging modality that can be utilized to diagnose cardiac tumors pre-operatively.[4] Herein, we present a 55-year old man who presented with a

history of dizziness and found to have a right ventricle outflow tract (RVOT) mass on echocardiogram. Further assessment with CMR revealed a right ventricle outflow tract well-circumscribed enhancing mobile mass that was protruding into the main pulmonary artery through the pulmonary valve during the systolic phase resulting in complete obliteration of the main pulmonary artery. The radiological features were favoring a cardiac myxoma. The patient underwent surgical resection of the mass and the histopathology results confirmed the diagnosis of cardiac intimal sarcoma. The information provided in this article aims to increase the awareness of radiologists regarding this rare entity that can simulate benign lesions. To our knowledge this is the first case of cardiac intimal sarcoma diagnosed in Oman.

Case report

A 55-year-old male patient, known to be hypertensive and diabetic, presented to the Emergency department at a local hospital with a two-week history of dizziness, which was exacerbated by exercise and relieved by rest. He denied any history of chest pain, fever, cough or shortness of breath.

Electrocardiogram (ECG) was done and showed T-wave inversion in the anterior leads. His chest radiograph was unremarkable. Laboratory investigations including troponin, complete blood count, liver function test and renal function were normal. An echocardiogram was requested and showed a right ventricular outflow tract mass, which was extending into the main pulmonary trunk and measuring 5 x 2.5 cm. The mass was obstructing the main pulmonary trunk giving a maximal gradient of 30 mmHg. In addition, there were mild concentric left ventricular hypertrophy and a small pericardial effusion. Otherwise, there was no evidence of valvular disease and the ejection fraction was normal; 65%. In view of the echocardiogram findings, a pulmonary angiography CT was requested, which confirmed the presence a right ventricular outflow tract well-defined hypodense mass extending to the main pulmonary trunk. The impression was a pulmonary embolism versus a myxoma with pulmonary embolism was favored over a tumor.

A cardiac MRI was requested for further characterization of the mass, and revealed a well-defined mass arising within the right ventricular outflow tract. The mass was protruding into

the main pulmonary trunk during systole phase and causing almost a complete occlusion of main pulmonary artery. It was isointense on steady state free precession (SSFP) images, hyperintense on T2-weighted images, hypointense on T1 and measuring 3.6 x 4 x 5.5 cm (Figure 1). The mass demonstrated an early first-pass perfusion and intense post-contrast enhancement on the delayed images (Figure 2). The findings on MRI were in-keeping with a neoplasm and confidently excluding thrombus. Differential diagnosis of the mass included myxoma and angiosarcoma. The patient underwent surgical excision of the RVOT mass, and it was sent for histopathological examination.

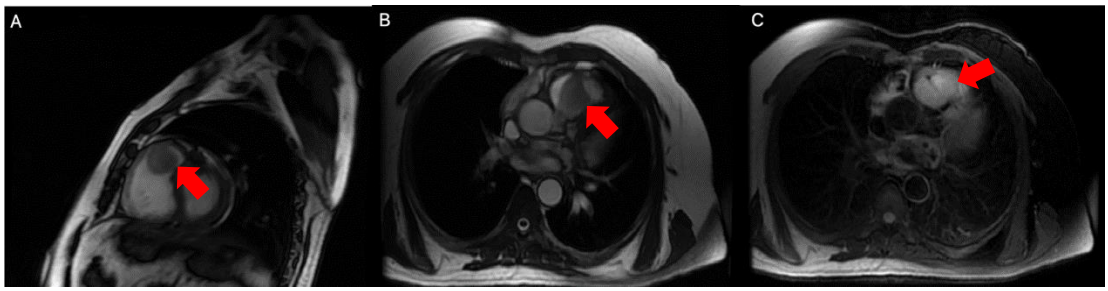

Figure 1: Cine steady state free precession MR images (A, C) showing a well-defined isointense mass located within the right ventricle outflow tract(Red arrow). The mass is hyperintense in T2-wighted image ©.

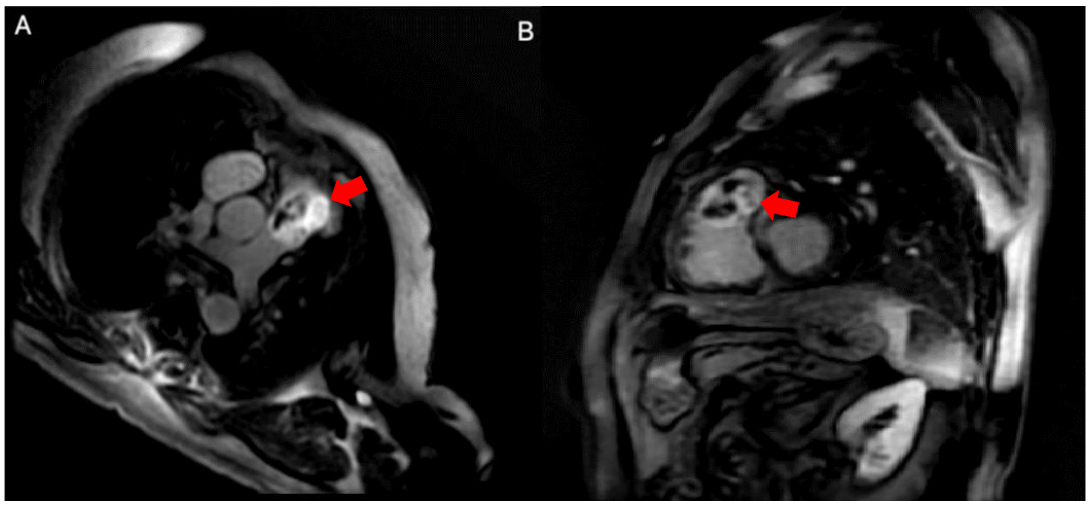

Figure 2: Late gadolinium axial (A) and short axis oblique (B) views showing delayed enhancement of the tumor with areas of non-enhancement representing necrosis (red arrow).

The resected specimen was a circumscribed light brown firm nodule weighing 25.4 gm and measuring 5.7 cm in maximum dimension. The cut surface was grey, white with a few yellow brown areas, focal myxoid areas and tiny cystic spaces (Figure 3). Microscopically, the tumor was composed of sheets of malignant cells with hyperchromatic and markedly pleomorphic

nuclei. Some of the cells showed clear or valcuolated cytoplasm. The tumor contained many variably sized vascular spaces. Areas of myxoid degeneration, hemorrhage and necrosis were also seen. The malignant cells were focally positive for MDM2, p16, SMA, and CD31. Other vascular markers (CD34, factor VIII, D2-40, and FLI-1) were all negative. Epithelial and other muscle markers were also negative. S100 was negative (Figure3). The histological findings were consisting with an intimal sarcoma.

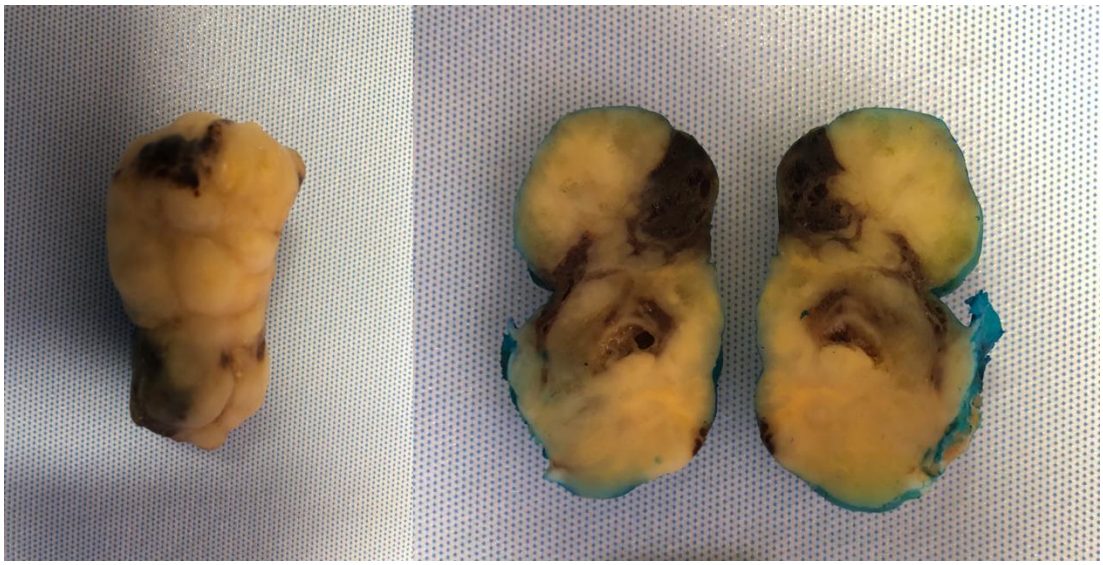

Figure 3: A resected nodular, fairly well circumscribed, grey white tumor.

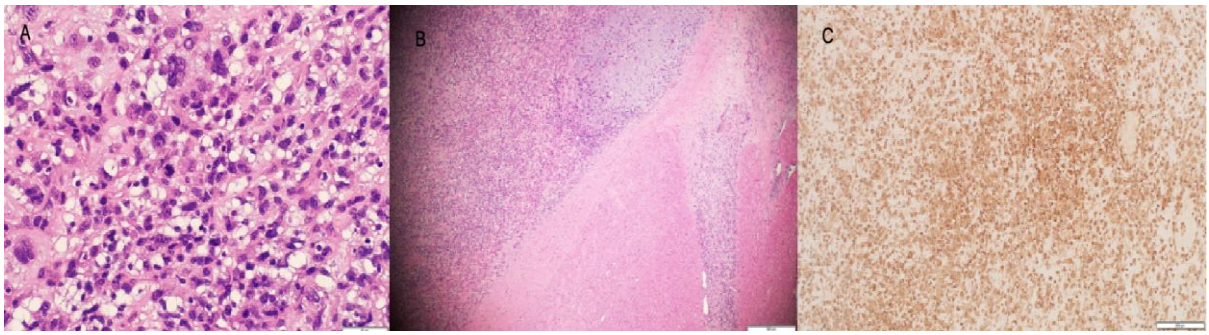

Figure 4: (A): Sheets of malignant cells with marked nuclear pleomorphism and scattered mitotic figures. (B): The malignant tumour invades the myocardial. (C): Immunohistochemistry profile shows positivity for S100.

After the diagnosis of intimal sarcoma, the patient had a PETCT that revealed no evidence of distal metastasis. The patients received an external beam radiotherapy using Volumetric Modulated arch Therapy (VMAT) technique. A follow up CT of the neck, chest, abdomen and pelvis after 6 months from treatment showed no evidence of local recurrence or distant metastasis. The patient was referred to the medical oncology clinic for further evaluation and

he underwent four cycles of chemotherapy with cyclophosphamide, Doxorubicin, and vincristine in combination

Discussion

Cardiac intimal sarcomas are extremely rare tumors with few reported cases in the literature.[5] The preoperative diagnosis of cardiac intimal sarcomas is very challenging, and they are usually misdiagnosed with thrombus or myxomas. Patients with cardiac intimal sarcomas can be asymptomatic or present with variable non-specific symptoms, depending on the location of the tumor, including valvular disease, heart failure, transit ischemic attack and chest pain.[5-8] In our case, the tumor was located in the RVOT and protruding into the main pulmonary artery during systole. This explains the clinical presentation of our patient with dizziness that was exacerbated by exercise and relieved by rest.

Histologically, intimal sarcoma composed of atypical spindle cell that have variable degrees of atypia, mitotic activity, necrosis and nuclear polymorphism.[3] Intimal sarcomas can rarely show areas with morphological features similar to angiosarcoma, rhabdomyosaroma, and osteosarcoma.[5] Immunohistochemically, tumor cells are usually positive for MDM2, vimentin and osteopontin. Variable positivity is seen for BCL-2, CD117, P53, CD68, SMA and desmin. Although, the tumor cells are typically negative for Factor VIII, CD34 and CD31, areas with angiosarcomatous differentiation can be positive.[5]

Different imaging modalities are used for assessment of cardiac tumors. Echocardiogram is a readily available non-invasive diagnostic tool that is usually used as a first-line imaging tool for the assessment of cardiac tumors. It allows anatomical localization of cardiac tumors and the assessment of their relation to cardiac structures. However, echocardiogram is operator dependent with restricted filed of the view and limited ability to the assess the right side of the heart. Moreover, echocardiogram is less sensitive for tissue characterization, and sometime the differentiation between cardiac tumors and thrombi can be challenging.[9] MRI is a well-established imaging tool of cardiac masses.[4] It allows the differentiation of tumors from thrombi and further characterizing tumors based on their signals in different sequences. Inversion recovery technique acquired with a prolonged inversion time (>600ms) after contrast administration can accurately differentiated tumors from thrombus. Tumors usually have

intermediate signal intensity to the myocardium whereas thrombus appears dark. On late gadolinium enhancement sequence, thrombus does not enhance as it is avascular structure. On the other hand, the majority of cardiac tumors are usually hyperintense owing to their vascularity.[4, 10]

Intimal sarcomas are highly aggressive, rapidly growing mesenchymal tumors with a very poor prognosis. The mean survival rate of patients with intimal sarcoma is 3 to 12 months.[5, 8] Surgical resection with clear margins is the main stay in the management of cardiac sarcomas.[7] Patients with complete resection live twice as the patients without resection. However, complete resection is not always achievable as tumor might infiltrate vital structures.[7, 8] Postoperative chemotherapy has been reported to be effective in some cases but its role in the treatment of PA intimal sarcoma is still not clearly defined. The same is true for radiation therapy and postoperative anticoagulation therapy. Our patient underwent resection of the tumor follow by radiation therapy and three cycles of chemotherapy including cyclophosphamide, Doxorubicin, and vincristine in combination. Follow up after 6 months revealed no evidence of recurrence of distance metastasis.

Conclusion

Intimal sarcomas are very rare, highly aggressive and rapidly growing mesenchymal tumor that can mimic benign cardiac tumors. Awareness of this rare entity and high index of suspicion can help in early diagnosis and prompt surgical intervention.

References

1. Leja MJ, Shah DJ, Reardon MJ. Primary cardiac tumors. Tex Heart Inst J. 2011;38(3):261-2.
2. Paraskevaidis IA, Michalakeas CA, Papadopoulos CH, Anastasiou-Nana M. Cardiac tumors. ISRN Oncol. 2011;2011:208929- DOI: 10.5402/2011/208929.
3. Neuville A, Collin F, Bruneval P, Parrens M, Thivolet F, Gomez-Brouchet A, et al. Intimal sarcoma is the most frequent primary cardiac sarcoma: clinicopathologic and molecular retrospective analysis of 100 primary cardiac sarcomas. Am J Surg Pathol. 2014;38(4):461-9 DOI: 10.1097/pas.0000000000000184.
4. Motwani M, Kidambi A, Herzog BA, Uddin A, Greenwood JP, Plein S. MR imaging of cardiac tumors and masses: a review of methods and clinical applications. Radiology. 2013;268(1):26-43 DOI: 10.1148/radiol.13121239.
5. Ibrahim A, Luk A, Singhal P, Wan B, Zavodni A, Cusimano RJ, et al. Primary Intimal (Spindle Cell) Sarcoma of the Heart: A Case Report and Review of the Literature. Case Reports in Medicine. 2013;2013:461815 DOI: 10.1155/2013/461815.

6. Valecha G, Pau D, Nalluri N, Liu Y, Mohammad F, Atallah JP. Primary Intimal Sarcoma of the Left Atrium: An Incidental Finding on Routine Echocardiography. Rare Tumors. 2016;8(4):6389 DOI: 10.4081/rt.2016.6389.
7. Li Z, Hsieh T, Salehi A. Recurrent Cardiac Intimal (Spindle Cell) Sarcoma of the Left Atrium. Journal of Cardiothoracic and Vascular Anesthesia. 2013;27(1):103-7 DOI: https://doi.org/10.1053/j.jvca.2011.07.027.
8. Grant L, Morgan I, Sumathi V, Salmons N. Intimal sarcoma of the left atrium presenting with transient ischaemic attack — A case report and review of the literature. Journal of Cardiology Cases. 2020;21(3):89-92 DOI: https://doi.org/10.1016/j.jccase.2019.10.006.
9. Malik SB, Chen N, III RAP, Hsu JY. Transthoracic Echocardiography: Pitfalls and Limitations as Delineated at Cardiac CT and MR Imaging. RadioGraphics. 2017;37(2):383-406 DOI: 10.1148/rg.2017160105.
10. The European Association of Cardiovascular Imaging (EACVI). EACVI CMR Pocket Guides [updated August 7, 2020. Available from: http://www.cmr-guide.com/Page102.html.