Annals of Surgical Case Reports

Case Report
Published: 04 Apr, 2019

Primary Cardiac Intimal Sarcoma

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Abstract

Primary cardiac intimal sarcoma is an extremely rare, aggressive malignant tumour with a poor prognosis. Intimal sarcoma arises from the sub-endothelial cells in the intima of the large arterial blood vessels. Here, we present a 44 years old female with primary cardiac intimal sarcoma, originating from the left ventricle and left atrium with involvement of the mitral valve.

Introduction

Primary cardiac tumours are rare, with approximately 75% of cases being benign, and the majority of these being myxomas [1,2]. Among the malignant tumours, sarcomas account for approximately 95%, with lymphomas making up the remainder [1]. Metastasis to the heart from a primary tumour elsewhere however is far more common and it would be prudent to rule out a metastatic lesion when investigating a cardiac tumour [3]. Primary cardiac sarcomas can have differing presenting symptoms and signs or may be diagnosed incidentally, and are often advanced at presentation. Most cases are treated with surgical resection and chemotherapy, however the prognosis remains poor and the majority of patients die within a year [4].

Case Report

We report the case of a 44 years old morbidly obese female (BMI of 44), with no other significant co-morbidities, presenting to her GP with a month's history of dyspnea, palpitations and dizziness. A CTPA was ordered to exclude pulmonary embolism, which showed a lobulated, 54 mm left ventricular mass (Figure 1). Subsequent transthoracic echocardiography revealed a mass sitting on the mitral valve and extending into the left ventricle. CT staging scan did not reveal any other distant metastasis at the time of presentation. The patient was referred for a cardiothoracic surgical opinion, with a provisional diagnosis of angiosarcoma or atrial myxoma. The patient underwent excision of the mass and mechanical mitral valve replacement. Intra-operatively, the tumour was adherent to the anterior and lateral walls of the left ventricle, with an unclear border of extension, making complete resection difficult. An intra-operative frozen section was requested on the ventricular mass (Figure 2). This showed a highly cellular lesion comprised of pleomorphic spindle cells with foci of necrosis and scattered mitoses, and was reported as a high grade malignancy favoring sarcoma (Figure 3). Combined morphological, immunological and molecular studies performed on the resection specimen were consistent with a diagnosis of primary cardiac intimal sarcoma. Cardiac transplantation was considered upon expert consultation, however due to morbid obesity

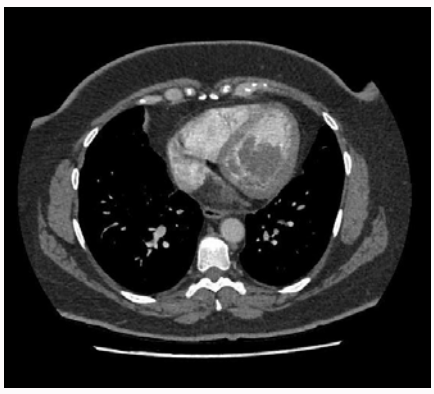

Figure 1: CTPA shows a lobulated, left ventricular mass, as well as a grossly enlarged pulmonary trunk.

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Received Date: *08 Mar 2019*
Accepted Date: *02 Apr 2019*
Published Date: *04 Apr 2019*

Citation:
Thuraisingam A, Fletcher N, McKay G, Smiles J. Primary Cardiac Intimal Sarcoma. Ann Surg Case Rep. 2019; 2(1): 1013.

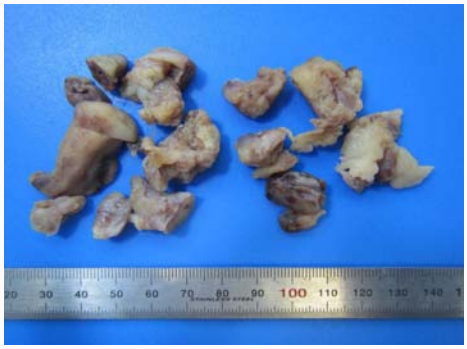

Figure 2: Post fixation image of ventricular mass.

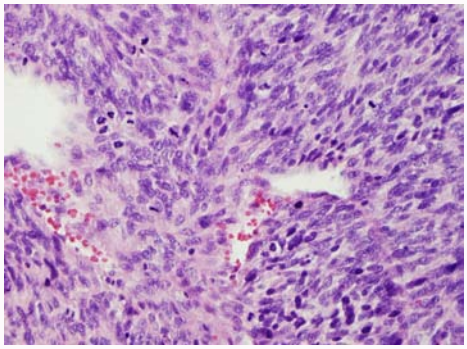

Figure 3: Formalin fixed, paraffin embedded section showing a highly cellular tumour with pleomorphism and a high mitotic rate.

the patient did not qualify. The patient was referred to an oncologist for chemotherapy. She was commenced on weekly Paclitaxel as a palliative adjunctive chemotherapy to control the disease progression. Her three months follow up echocardiography study and PET scan revealed recurrence of the tumour in the left ventricle and metastasis to the brain, bilateral adrenals, gluteus muscles and ileum. Due to rapid metastasis, she elected to have radiation therapy to the bilateral adrenal metastases, and stereotactic radiation therapy to the brain metastasis. Unfortunately, her latest re-staging PET scan revealed progression of the disease with a new solitary lesion in the liver despite ongoing chemotherapy and radiation therapy. The latest echocardiogram showed stable intimal sarcoma with left ventricular outflow obstruction. As she became more symptomatic, the palliative care teams were consulted for comfort care and pain management.

Comment

Primary intimal sarcoma of the heart is a rare and aggressive malignancy that carries a poor prognosis [2,4]. Also called spindle cell sarcomas, these are mesenchymal tumours that arise from subendothelial cells in the intima of large blood vessels and when occurring in the heart tend to involve the left atrium and mitral valve [5]. Complete surgical resection is often not possible due to the highly aggressive nature of the tumour, and the frequent involvement of critical structures. Local recurrence and metastases are common and often occur early despite adjunctive chemotherapy and radiotherapy.

In recent years, molecular studies using FISH or PCR techniques have shown intimal sarcomas to characteristically show amplification of Murine Double Minute 2 (MDM2) protein [1,2,5,6]. Positivity for MDM2 by immunohistochemistry is also typical, and can aid in initial assessment of a poorly differentiated cardiac sarcoma. Other reported genetic mutations include PDGFR (A/B), ECFR, CDK4 and HMGA1 [5-7]. In addition to use as a diagnostic tool, having knowledge of the molecular alterations around this rare tumour may allow for future targeted therapies.

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