

Primary Cardiac Angiosarcoma with Adrenal Metastasis

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INTRODUCTION

Cardiac angiosarcomas are rare tumours which can present with varying clinical symptoms, and are usually difficult to manage. We present a case of a young female who presented with respiratory symptoms, and was found to have a primary cardiac angiosarcoma with metastases. A discussion of primary angiosarcoma and a review of its management follows.

CASE REPORT

A 33-year old female with no known illnesses was admitted to The Queen Elizabeth Hospital with a three-month history of cough, shortness of breath and weight loss. On examination, she was found to have decreased air entry in the right mid to lower zones with dull percussion note. Laboratory investigations revealed a microcytic anaemia and mild dehydration. Chest X-ray revealed a mediastinal mass and a large right pleural effusion. Computed tomography (CT) scan of the chest, with intravenous contrast showed a large hyper-vascular mass in the right atrium, invading the superior vena cava, with collateral veins (Figs. 1 and 2).

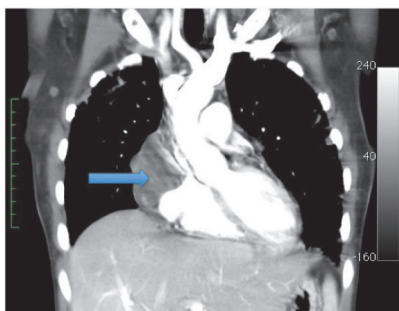


Fig. 1: Coronal computed tomography imaging of the chest showing right atrial mass.

There were multiple small nodules in both lung fields and a moderate right pleural effusion. There was also a 2 cm mass in the right adrenal gland (Fig. 3).

Echocardiography confirmed the presence of a large right atrial mass with tamponade type physiology due to the tumour occupying a significant portion of the right atrium (Fig. 4).

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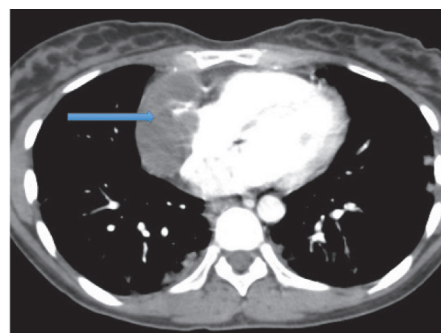


Fig. 2: Axial computed tomography imaging of the chest showing right atrial mass.

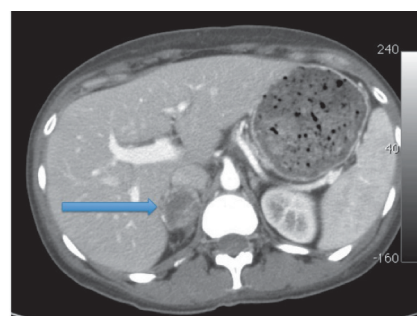


Fig. 3: Contrast computed tomography imaging of the abdomen showing large right adrenal mass.

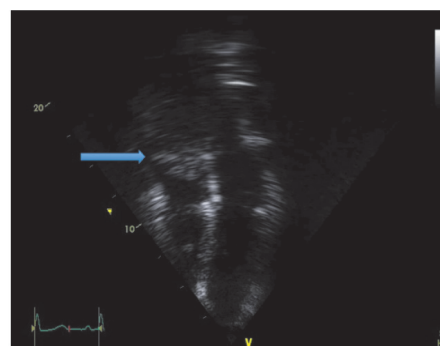


Fig. 4: Echocardiographic imaging showing large right atrial mass.

Differential diagnoses included lymphoma, thymoma, cardiac tumour or a primary mediastinal tumour. The patient underwent CT-guided core biopsy of the mediastinal mass. She was subsequently referred to the Radiation Oncology service for rescue radiation therapy, and underwent 30 Gy of radiation in six sessions, over a three-week period.

Histology of the mediastinal specimen showed predominantly blood clot and a small fragment of fibrovascular tissue with benign epithelium at one edge. The fibrovascular tissue contained rare, mildly epithelioid cells at the area of tissue fragmentation. The specimen was thought to not be

representative of the pathologic condition. Repeat CT-guided biopsies of the mass and the right adrenal mass were performed. Computed tomography scan performed post radiation treatment showed a reduction in the size of the mass.

Histology of the repeat mediastinal biopsy showed portions of cardiac muscle and thrombus (Fig. 5). There was no evidence of malignancy.

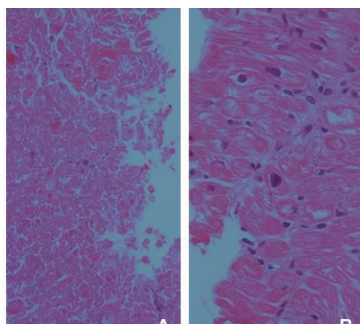


Fig. 5: Haematoxylin and eosin (H&E) staining of sections of the biopsy of cardiac muscle showing part of the thrombus (A) and normal cardiac myocytes (B).

The adrenal specimen showed portions of malignant neoplasm composed of groups and trabeculae of cells with medium sized polygonal nuclei (Fig. 6). Immunohistochemistry of the cells showed strong positivity for CD31, a vascular immunomarker (Fig. 7). The cells were negative for epithelial markers (cytokeratin 7 and cytokeratin 20) as well as desmin. A diagnosis of primary cardiac angiosarcoma with metastasis to the adrenal gland was made.

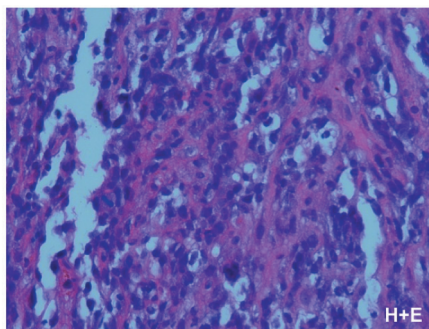


Fig. 6: Adrenal gland showing a malignant neoplasm composed of medium sized cells with pleomorphic, hyperchromatic nuclei with scattered mitoses.

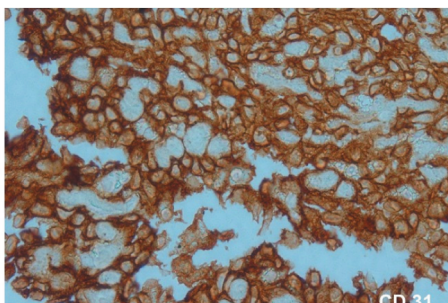


Fig. 7: Adrenal gland, showing the characteristic 'chicken wiring' on CD31 staining.

The oncologists decided that the patient should undergo 12 cycles of Taxol in an attempt to reduce the tumour burden, as she was not a candidate for surgical resection as we had no options for replacement of the right atrial wall. She completed four cycles of treatment, but developed a lower respiratory tract infection, which lead to her demise, six months after initial presentation.

DISCUSSION

Cardiac angiosarcoma is a rare clinical entity with limited information available regarding the optimal management. The disease tends to be aggressive and most patients have widespread metastases at the time of diagnosis, further worsening an already bleak prognosis. Angiosarcomas are a subtype of soft-tissue sarcoma and are aggressive, malignant endothelial-cell tumours of vascular or lymphatic origin.

Most patients present with symptoms of dyspnoea, atypical chest pain, haemoptysis and other non-specific symptoms such as nausea, emesis, fever and anorexia (1).

Nakaya *et al* reported a case of a 72-year old man with a primary cardiac angiosarcoma presenting with cardiac tamponade (2). There is no typical angiosarcoma patient, as they can present at any age, with no gender predilection. Cases of primary cardiac angiosarcomas have been described in patients as young as 23 months of age (3).

Imaging modalities used to aid in the diagnosis include CT scan, transthoracic or transoesophageal echocardiography, positron emission tomography scan and magnetic resonance imaging. Definitive diagnosis is made based on characteristic features on immunohistochemical assessment.

Treatment strategies described include resection of the tumour and reconstruction of the atrium with autologous or bovine pericardium, radiation therapy and chemotherapeutic regimens based on anthracyclines, taxanes and ifosfamide. Interleukin use has also been described.

One of the longest patient follow-ups, post diagnosis and treatment was published by Nakamichi and colleagues in 1997. They reported a 53-month survival of an eight-year old child who underwent excision of a primary right atrial angiosarcoma and extensive immunochemotherapy with dacarbazine, THP-doxorubicin, ifosfamide and interleukin-2 over a two-year period, as well as irradiation with 50 Gy in the early postoperative period (3). Unlike most other cases reported, this patient had no systemic metastases at presentation. Median survival in patients with complete surgical excision is approximately 17 months and approximately six months in those in whom surgical resection cannot be achieved (4).

CONCLUSION

The advances in imaging diagnostic methods has allowed for tissue sampling in symptomatic patients, and advances in immunohistochemical assessment and evaluation have resulted in accurate ante-mortem diagnoses being made. Screening for the condition is impossible as there is no defined patient subset at increased risk and risk factors are largely unknown. Al-

though some progress has been made regarding the treatment strategies for primary angiosarcoma, the 'optimal' treatment is yet to be described. Fortunately, primary cardiac angiosarcomas are a rare clinical entity as a cure for this condition remains elusive.

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