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## CASE REPORT

## Acute myeloid leukaemia relapse presenting as cardiac myeloid sarcoma

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**SUMMARY**

A 50-year-old woman previously diagnosed with acute myeloid leukaemia presented with a 3-month history of shortness of breath and a right-sided facial rash. A chest CT revealed an intracardiac mass in the right atrium extending into her superior and inferior vena cava. Surgery was performed to remove the mass and pathology was consistent with myeloid sarcoma. After surgery, adjuvant radiation therapy was directed to the residual disease. The patient eventually relapsed in other sites not including the right atrium and eventually succumbed to her disease.

**BACKGROUND**

Acute myeloid leukaemia (AML) is a relatively uncommon malignancy representing approximately 1.2% of new cancer cases in the USA.<sup>1</sup> Myeloid sarcoma (MS) is a rare cancer type that is commonly associated with AML with an incidence of 2.5%–9.1% in AML patients.<sup>2</sup> Due to the rarity of MS, little is known about the treatment of this disease. The malignancy commonly involves the skin, bone or lymph nodes, but can affect many locations in the body.<sup>3</sup> Intracardiac MS is a very rare disorder and only a few cases have been published about this malignancy; as such a treatment protocol has yet to be determined.<sup>4–8</sup> We present a case of an intracardiac MS treated with surgery and adjuvant radiation therapy.

**CASE PRESENTATION**

A 50-year-old woman presented in January 2016 with a 3-month history of shortness of breath on exertion and a rash on the right side of her face. Her medical history was significant for AML with trisomy 8. She had previously undergone a sibling donor stem cell transplant for the treatment of her AML in January 2014 and was in complete remission. Her skin rash was attributed to graft versus host syndrome. Her current medications include tacrolimus 0.5 mg every 12 hours on an empty stomach.

**INVESTIGATIONS**

Imaging studies including a CT scan was performed of her chest revealing a mass involving the right atrium with possible associated thrombus formation. She was admitted to the hospital for further evaluation and workup. The cardiac mass measured 4.4×5.6×6.8 cm and was situated along the margin of the right atrium with a septal attachment

extending to the superior and inferior vena cava (figures 1–3). A transthoracic echocardiogram (TTE) identified a centrally located hypodense mass that was thought to be an associated thrombus. An atrial septal defect was also noted in the TTE. A pulmonary function test was performed which was unremarkable.

A myocardial perfusion echocardiogram was performed and the soft tissue mass displayed some areas of vascularisation with areas of hypoperfusion. After a review of the case by a cardiologist and cardiothoracic surgeon, resection of the right atrial mass was recommended. A cardiac catheterisation showed normal coronary arteries, clearing the patient for surgery.

A tissue sample of the cardiac mass was sent to pathology undergoing immunohistochemical staining with CD34, CD117, CD68, terminal deoxynucleotidyl transferase, myeloperoxidase, CD3 and CD20. Proper controls for the staining were used and the neoplastic cells were positive for myeloperoxidase, CD68 and CD117. The tumour pathology was consistent with a MS due to an AML relapse.

**TREATMENT**

The patient underwent excision of the cardiac mass. At the time of the surgery, the tumour was noted to be invading into the lateral wall of the right atrium. The tumour extended into the superior and inferior vena cava, the septal wall and the roof of the left and right atria. The right atrial wall, atrial septum, roof of the right and left atrium and approximately 95% of the tumour was resected. The right atrium was repaired with bovine pericardial patches.

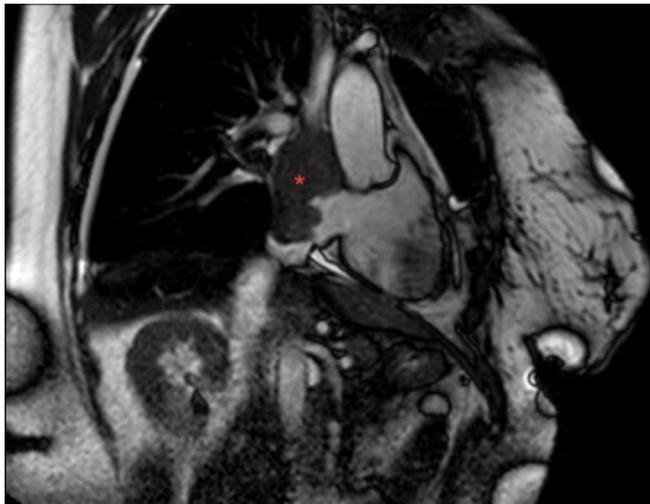
The surgery was completed without any complications, but the patient remained in ventricular escape. She later developed anisocoria and both a neurologist and ophthalmologist were consulted. A pacemaker was placed to correct the ventricular escape rhythm. Approximately 2 weeks after the pacemaker was placed, she experienced heart failure due to tricuspid regurgitation and was treated with furosemide until the regurgitation was resolved.

After she recovered from her surgeries, she underwent a bone marrow biopsy, which was negative for leukaemia. Adjuvant radiation therapy of 2000 cGy to the residual disease involving the right atrium was recommended and she underwent 10 fractions of 200 cGy to the residual disease. The volume of lung tissue receiving 200 cGy (V20) for the treatment was <1% with a mean lung dose of 425 cGy. The radiation was tolerated well and the



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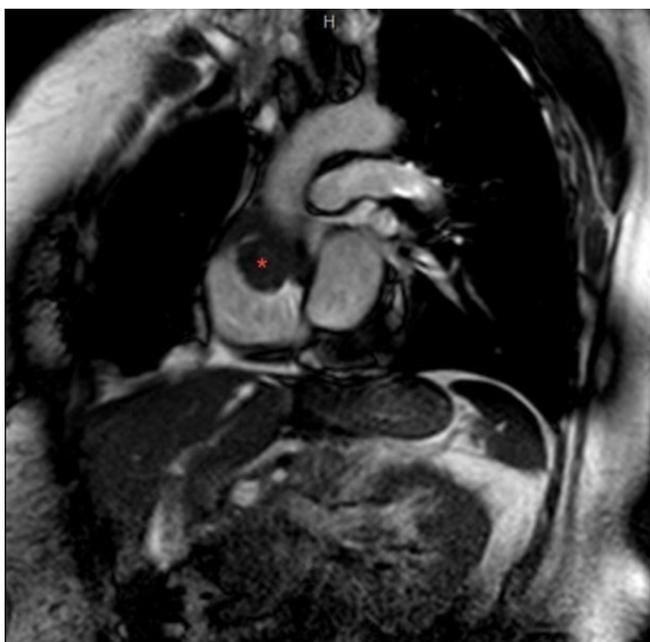


**Figure 1** A sagittal MRI scan of the heart showing the cardiac mass in the right atrium. The mass (red asterisk) is attached septally and extends from the superior vena cava to the inferior vena cava.

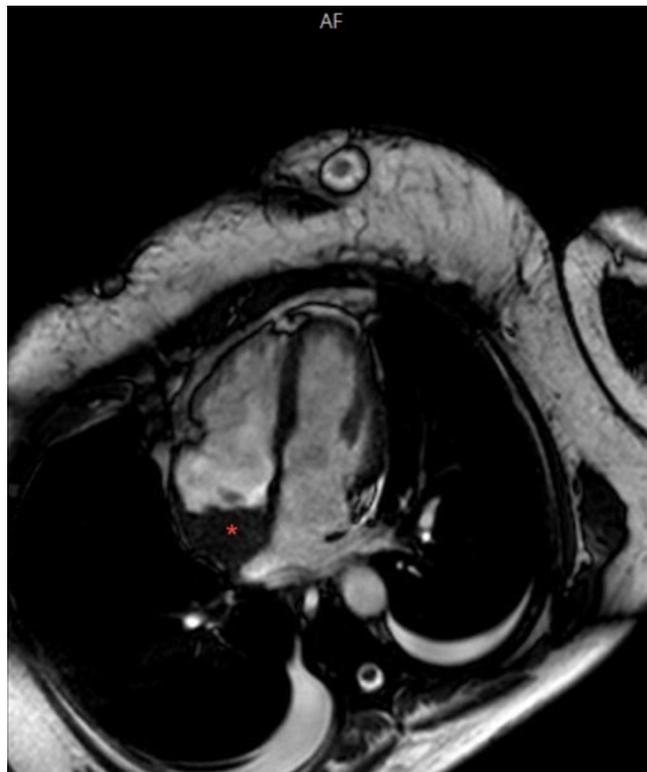
course was completed without any interruptions. The patient developed mild radiation esophagitis, which was treated with lidocaine and Mylanta.

#### OUTCOME AND FOLLOW-UP

A month after the completion of treatment, she developed extreme shortness of breath with hypoxaemia. She was diagnosed with radiation pneumonitis and placed on a 4-day course of steroids. After the course of steroids, the patient developed severe pain and swelling in the right eye and right maxillary region. The radiation pneumonitis recurred for the next 2 weeks, so she was started on a course of 60 mg of prednisone daily for a week and then 40 mg daily for a week after that. Her shortness of breath was treated and she denied any chest pain or coughing. She later developed multiple MS recurrences involving the right



**Figure 2** A frontal MRI scan of the heart showing the cardiac mass in the right atrium. The red asterisk marks the tumour.



**Figure 3** A transverse MRI scan of the heart showing the cardiac mass in the right atrium. The red asterisk marks the tumour.

orbit, right humerus and right brachial plexus for which radiation was given at another hospital. The patient relapsed again in November 2016 with dissemination of AML to the central nervous system and she expired a month later.

#### DISCUSSION

AML is a heterogeneous malignancy of the myeloid line of cells. Depending on the gene mutations and patient characteristics, prognosis and treatment options will vary.

MS is most commonly associated with AML and can precede or succeed AML.<sup>9</sup> MS is an extramedullary mass of myeloid cells that can occur almost anywhere in the body. MS may be difficult to diagnose because of its similarity to other neoplasms and a biopsy is often necessary.<sup>10</sup>

Prognosis for MS that is associated with AML or another lymphoproliferative disorder is usually poor, while de novo MS is responsive to both radiotherapy and chemotherapy. Patients that responded to initial therapy, overall had a better prognosis than those who did not respond. In a study of 92 MS patients, the longest survival post-treatment was seen with patients who underwent stem cell transplant.<sup>9</sup>

MS is a rare tumour regardless of the location in which it is found, making a standardised treatment difficult. In one case of MS, a patient had a thoracic dorsal epidural mass compressing his spinal cord resulting in weakness and numbness in his lower extremities. The mass was surgically resected, but recurred 13 days later compressing the spinal cord. Emergent radiotherapy was initiated consisting of 3000 cGy in 10 fractions over 10 days, with immediate improvement in the patients' lower extremity weakness. Imaging of the patient's spine revealed decompression of the spinal cord.<sup>11</sup> Another patient presented with an MS mass in his gastric body and was treated with idarubicin and cytosine arabinoside. An endoscopy showed the gastric mucosa was

clear of nodular lesions both at 3 months after chemotherapy and 1 year after chemotherapy.<sup>12</sup> In a case of a patient with an MS mass in the superior mediastinum, the treatment was daunorubicin and cytosine arabinoside. The patient presented with pleural effusion, which quickly resolved after starting chemotherapy. After the initial therapy, consolidation therapy with cytosine arabinoside was given. Radiological imaging showed complete resolution of the tumour mass and the patient is being followed-up for symptom improvement.<sup>13</sup>

In a study of 335 patients diagnosed with MS, only 32% of the patients received radiation therapy and this varied depending on the location of the malignancy.<sup>14</sup> In patients younger than 60 years of age, 70%–80% are expected to achieve complete remission after chemotherapy.<sup>15</sup>

There have been few case reports on intracardiac MS, precluding identification of the best treatment for this malignancy.<sup>4–8</sup> A case of a patient with a cardiac presentation of MS which was treated with both chemotherapy and radiotherapy.<sup>4</sup> The patient responded well, but relapsed 2 years later. Further relapse was seen 1 year after the second treatment and the patient ultimately died from sepsis-associated respiratory failure. In another case of a cardiac MS, a woman diagnosed with MS was in remission from AML for 10 years before relapse. She underwent chemotherapy for both the AML and MS. This patient died 4 months after treatment due to a transplant complication.<sup>6</sup> Compared with the previously mentioned patient who also underwent chemotherapy, this second patient had a much shorter survival after treatment, highlighting the difficulty in deciding treatment in these small number of cardiac MS cases.

As can be seen from our patient and the multiple cases described above, MS is a rare disease especially intracardiac MS, which does not have a standardised method of treatment. Multiple treatment options exist for the treatment of an intracardiac MS and MS including surgery, radiation and chemotherapy or a combination of the three. The purpose of this case report was to review the different treatment options for intracardiac MS. Unfortunately, although our patient discussed in this case report expired secondary to systemic disease, local control of her intracardiac MS was obtained with the addition of radiation therapy with minimal side effects. In our patient, radiotherapy after surgical resection of the tumour was adopted in an effort to prevent tumour recurrence in the heart.

Although, our patient was diagnosed with radiation pneumonitis at an outside hospital, the volume of her lung tissue that received greater than 200 cGy was less than 1% making this diagnosis less likely. Radiation pneumonitis is an adverse side effect of radiation therapy to the lungs and can be minimised by limiting the lung V20 to less than 22%.<sup>16</sup> In addition, our patient also developed radiation esophagitis which was easily treated with conservative measures.

Which options to choose to treat intracardiac MS should be based on available data and after a thorough discussion in a multidisciplinary tumour board as well as a discussion with the patient and family members. In our patient, radiation therapy seemed to be an effective component of her treatment. The radiation therapy was well tolerated with minimal side effects. It also appeared to control her intracardiac disease and her recurrent disease that developed at other sites. In conclusion, radiation therapy should be considered in the treatment of intracardiac MS since it appears to be well tolerated with minimal side effects and provides durable local control of disease.

## Learning points

- ▶ Myeloid sarcoma rarely presents as an intracardiac mass and a treatment protocol has not been developed. The treatment for intracardiac myeloid sarcoma should be determined on a case-by-case basis.
- ▶ Treatment used for intracardiac myeloid sarcoma has been surgery, radiation, and chemotherapy.
- ▶ A variety of treatments have been used because no single method has been proven to be more effective than another for myeloid sarcoma of the heart.

**Contributors** AW was responsible for gathering the patient data, reviewing the pertinent articles related to the topic under supervision of SM and MJ, as well as a major contributor to the writing of the paper. MJ provided knowledge and guidance of the biology underlying the treatment as well as assistance in the writing and editing of the paper. SM is the senior author as well as the physician who developed the treatment plan for this patient.

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