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CASE REPORT

Monophasic synovial sarcoma: a rare cause of a primary cardiac malignancy

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Introduction

Primary cardiac monophasic synovial sarcomas are exceedingly rare tumours. This article describes a case of a poorly differentiated synovial sarcoma presenting in a young woman. As definitive diagnosis may be difficult, treatment for such patients is usually delayed. This case outlines the clinical picture, the investigations needed for diagnosis, and the management of such rare tumours. A multidisciplinary team approach is crucial in ensuring optimal patient care.

Case report

A previously healthy 19-year-old woman presented to the emergency department with a 4-week history of bilateral intermittent cramp-like flank pain, with each episode lasting 2–3 hours. The patient complained of associated nausea and vomiting and was unable to keep down anything she ate or drank.

She had experienced four syncopal episodes over a 1-week period without preceding pre-syncopal features or dizziness. The patient was otherwise haemodynamically stable. Examination was unremarkable except for mild tenderness in the flanks.

Electrocardiogram showed a right bundle–branch block. Chest X-ray was unremarkable. Bloods revealed an elevated D-dimer level of 2239 ng/ml (normal range 0–500 ng/ml) and NT-proB-type natriuretic peptide level of 5686 pg/ml (normal range 0–125 pg/ml).

A computed tomography pulmonary angiography scan showed a massive filling defect in the right ventricle extending across the tricuspid valve into the base of the right atrium, causing this to dilate. The filling defect extended into the right ventricular outflow tract to fill the main pulmonary trunk. The right ventricular outflow tract was dilated, measuring 3.6 cm (2–3 cm), and the left ventricular outflow tract was normal. The impression at this point was of a massive right intra-ventricular thrombus with right intra-atrial and right ventricular outflow tract extension resulting in severe right heart strain.

Echocardiography confirmed the presence of a 5 cm mass in the right ventricle, extending into the right ventricular outflow tract and also prolapsing across the tricuspid valve and into the right atrium. There was evidence of right ventricle pressure overload as pulmonary pressures were estimated at 60 mmHg (17–30 mmHg). Mild tricuspid regurgitation and severe pulmonary regurgitation were present.

Urgent cardiac surgery was performed to remove the suspected thrombus. Upon assessing the cardiac chambers, a large soft rubbery mass was apparent in the right ventricle. This was associated with significant involvement of the pulmonary trunk, such that the pulmonary valve was barely visible. The right atrium and right ventricle were highly enlarged with severe right ventricular hypertrophy of 8 mm (normal right ventricle thickness 3–5 mm).

The macroscopic surgical findings raised the possibility of an intra-cardiac tumour, so a cardiac magnetic resonance scan was performed. This showed a dilated right ventricle with severely impaired systolic function. Tissue characterisation sequences were highly suggestive of an intra-cardiac tumour, which was poorly perfused in the right ventricle apex.

Histology of the excised intracardiac mass was in keeping with a monophasic synovial sarcoma (Figure 1). Cardiac magnetic resonance findings indicated that the excision was incomplete (Figure 2).

Immunohistochemistry analysis was also performed. The Ki-67 proliferation index was significant throughout the tumour, with certain areas having a Ki-67 proliferation index of 50% (<20%) (Figure 3).

Case report (continued)

The patient underwent a second, debulking operation, during which the right ventricular outflow tract was noted to be free of tumour. However, multiple tumour sites were now observed, involving the right ventricle apex and papillary muscles. Histology was in keeping with areas of poorly differentiated round cell synovial sarcoma, French grade 3 (Figure 4).

Following the second operation, the patient was given 12 cycles of immunotherapy. Despite the immunotherapy, a computed tomography scan of the chest performed at the end of immunotherapy showed further progression, with bilateral pulmonary metastatic nodules and mediastinal lymphadenopathy present. The patient is being followed up closely by the oncologists.

Discussion

Primary cardiac synovial sarcomas are vanishingly rare. Poorly differentiated monophasic cardiac synovial sarcomas are even rarer, with very little literature describing such cases. Primary cardiac tumours account for 0.002–0.33% of all tumours, with 75% being benign

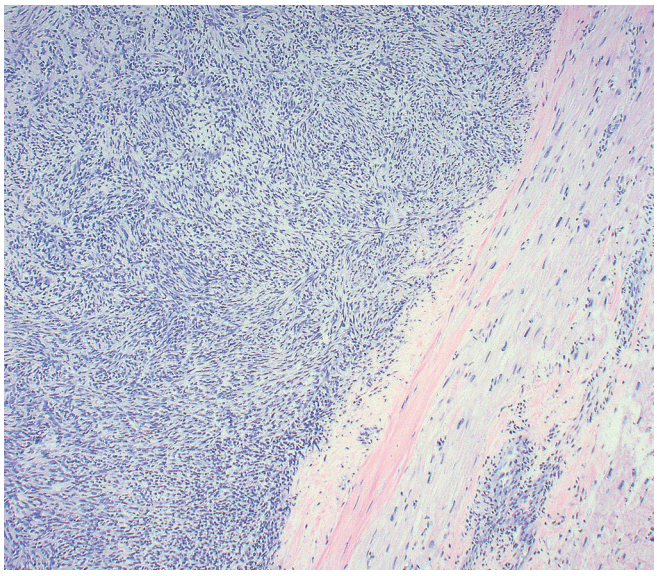

Figure 1. Histology showing a cellular spindle cell neoplasm (left) infiltrating the cardiac muscle (right) (haematoxylin and eosin x 200).

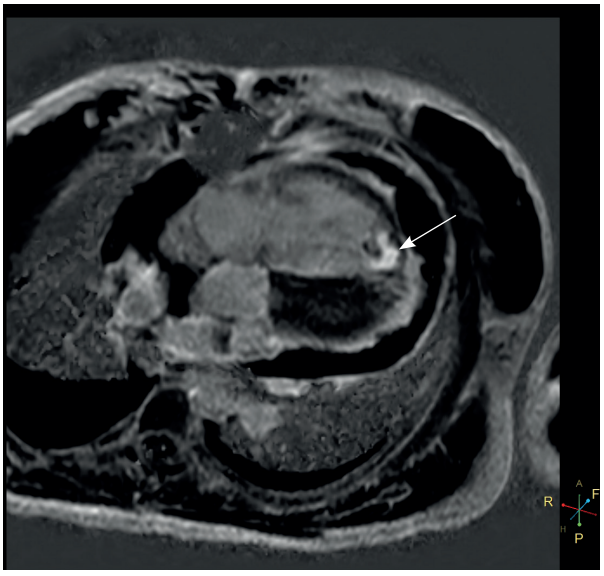

Figure 2. Cardiac magnetic resonance showing late gadolinium enhancement (white area, arrow), signifying that the mass is vascular in nature.

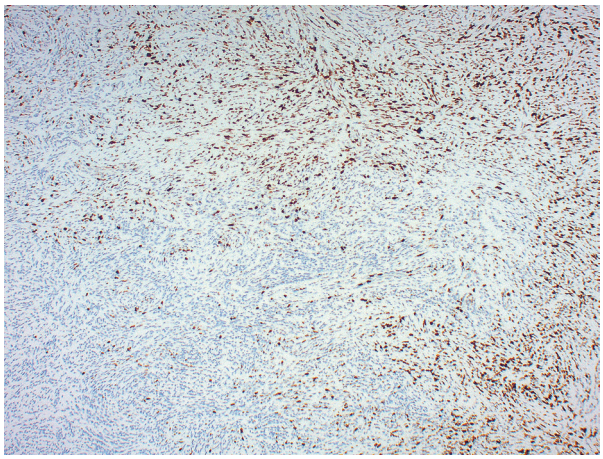

Figure 3. Immunohistochemistry showing a Ki-67 proliferation index which is relatively low in the conventional monophasic area (left) but much higher in the poorly differentiated area (right).

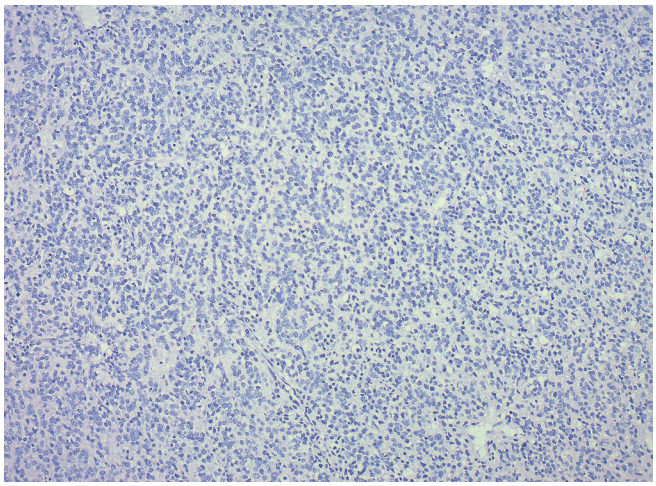

Figure 4. Area of poorly differentiated synovial sarcoma showing a 'small round blue cell tumour' morphology with very brisk mitotic activity (haematoxylin and eosin x 400).

Learning points

- Primary cardiac synovial sarcomas are rare tumours, which can be difficult to diagnose and hard to treat successfully.
- They have a relatively poor prognosis, owing to their aggressive nature and propensity to metastasise.
- More research is required to better understand these tumours and improve overall management and patient survival.

(Hoffmeier et al, 2014). Therefore, primary malignant cardiac tumours account for around 0.008% of all tumours, and 64.8% of these are sarcomas (Oliveira et al, 2015). Synovial sarcomas make up approximately 5% of all cardiac sarcomas. The prognosis of such tumours is very poor, as malignant synovial sarcomas tend to behave in a very aggressive manner, particularly if incompletely excised.

Cardiac synovial sarcomas tend to have a very non-specific presentation (Coli et al, 2018). In this case, the main symptomatology was seemingly gastrointestinal in nature, presumably secondary to hepatic congestion as a result of right-sided heart failure. The only feature in keeping with possible cardiac pathology was the recurrent episodes of unexplained syncope.

The Ki-67 proliferation index was estimated to be 50% in certain cells. This implies that these cells have a high proliferation rate, and is thus an indirect marker of prognosis – a high index is associated with a more aggressive tumour and a poorer prognosis. Consensus varies because of the lack of literature, but a Ki-67 proliferation index of more than 20% is generally considered abnormal, and a proliferation index of 50% definitely considered pathological (Skytting et al, 1999).

Two other important markers of prognosis are the age of the patient and the use of chemotherapy as a treatment option (Wang and Li, 2013). Patients diagnosed in the first three decades of life have a better prognosis, which is further improved with the administration of chemotherapy, irrespective of whether or not surgical excision is performed (Coli et al, 2018). The overall median survival post diagnosis is 5–27 months (Wang and Li, 2013).

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