Cardiology in the Young

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Rare haematogenous sarcoma metastasis to the heart in a child

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Abstract

Haematogenous non-contiguous metastatic spread of remote solid tumours to the heart is rare. We describe a previously healthy 5-year-old girl who presented with extensive intracardiac involvement by metastatic pelvic sarcoma.

Case report

A previously healthy 5-year-old girl was hospitalised with a 1-month history of progressive left knee pain with limp, spreading to the thigh and then to the right lower extremity, with eventual refusal to walk. There were no other complaints apart from intermittent low-grade fever of 38–39°C. There was no significant family history, traumatic injury, or infectious exposure. Initial investigations at another institution, including MRI, were interpreted as showing pelvic and sacral osteomyelitis. Her symptoms worsened during 12 days of intravenous antibiotic therapy with cephamycin and clindamycin.

On admission to our hospital, she was alert but in obvious discomfort, with stable vital signs including sinus tachycardia at 146 beats/minute, respiratory rate 24 breaths/minute, blood pressure 93/59 mmHg, temperature 37.7°C, and oximetric saturation of 98% in air. Significant findings were limited to marked generalised tenderness of both lower extremities. There was no hepatosplenomegaly. Laboratory investigations revealed mild anaemia (haemoglobin 9.7 g/dl), leucocytosis (WBC 13.6 × 10^3/μl), and increased C-reactive protein (132 mg/L) as well as ferritin (824 ng/ml). Liver enzymes were mildly and non-specifically elevated.

The electrocardiogram showed sinus tachycardia with rightward QRS axis deviation. The PR interval was 160 ms with QRS duration of 80 ms, and there were non-specific T-wave changes with a QTc of 475 ms, with no other significant abnormality noted. Chest x-ray showed moderate cardiomegaly with bilateral pulmonary venous congestion, and several very small parenchymal nodules of uncertain significance were seen in both lungs.

Transthoracic 2D and Doppler echocardiography demonstrated multiple large, homogeneous, sessile intracavitary masses adherent to walls and valve leaflets in all four cardiac chambers (Fig 1), with one mass most notably almost filling the left atrium (Fig 2). No masses were seen in the inferior caval vein. There was no apparent interatrial or interventricular shunting lesion based on colour Doppler septal interrogation. Mitral valve colour Doppler inflow was distorted and turbulent, with a peak velocity of 2 m/second measured by continuous wave Doppler. Tricuspid inflow was also qualitatively distorted, but no significant acceleration was apparent. There was a moderate-sized, non-circumferential pericardial effusion with a maximal depth of just over 2 cm, whose haemodynamic significance could not be conventionally assessed due to the large atrial masses as well as those adherent to AV valve leaflets (Fig 1).

CT of the chest and abdomen with and without contrast revealed a solitary lesion in the right adrenal gland, suspicious for malignant metastasis. There were multiple large intracardiac masses corresponding to those documented echocardiographically as already described. The main pulmonary artery and its right and left branches appeared patent and free of intraluminal masses. The inferior caval vein was similarly widely patent throughout its length, with no intraluminal masses observed. There was generalised pulmonary oedema, with scattered small parenchymal nodules bilaterally corresponding to those noted on chest x-ray. The left upper, left lower, and right lower pulmonary veins were essentially normal in their courses and caliber; the right upper and middle pulmonary veins were not visualised. CT-guided needle biopsy of the left ischium revealed bone invasion by cells histopathologically characterised as undifferentiated sarcoma.

Within days of her hospitalisation, the patient developed progressive respiratory distress. The family accepted a recommendation for comfort care based on assessment that her malignancy would not be amenable to rapid debulking with either chemotherapy or radiation, and she expired 9 days after admission. There was no post-mortem examination.

Brief Report

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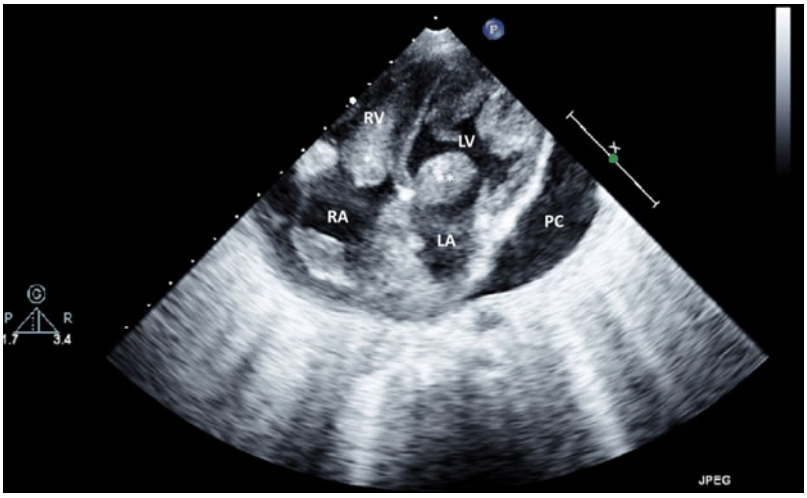

Figure 1. In this apical four-chamber two-dimensional echocardiogram, multiple large tumour masses are seen in all four cardiac chambers (RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle), notably including one that traverses the tricuspid valve (*), and another enveloping the anterior mitral valve leaflet (**). These lesions were associated with turbulent atrioventricular valve inflow, with increased peak velocity of 2 m/second through the mitral valve. A moderate-sized pericardial effusion is apparent (PC).

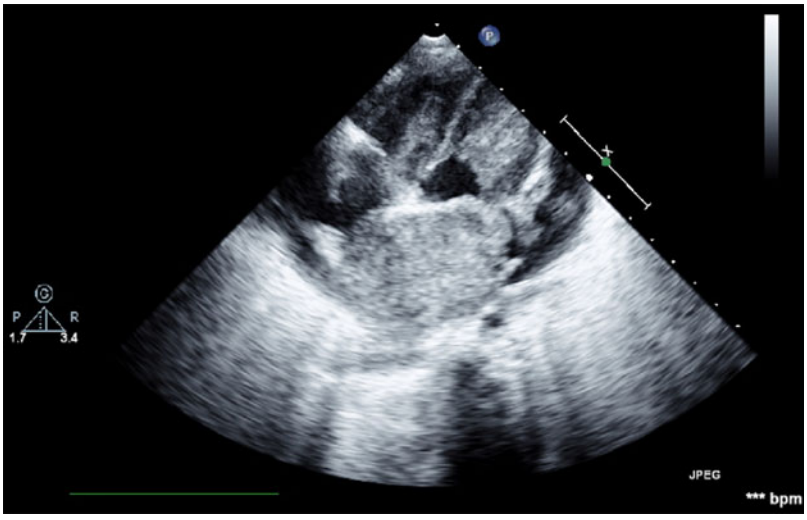

Figure 2. The left atrium is essentially occupied by a very large tumour mass.

Discussion

This patient represents a dramatic example of non-contiguous haematogenous cardiac involvement with metastatic sarcoma, which is a recognised but distinctly uncommon phenomenon. Yedururi et al recently reviewed the literature and summarised their own findings in 20 young patients known to have cardiovascular involvement with osteosarcoma.[1] Only 3 (15%) of their 20 patients had non-contiguous cardiac tumour involvement, while the other 17 exhibited direct intravascular extension from primary or pulmonary arterial metastatic lesions. They point out that osteosarcoma has a known predilection for vascular rather than lymphatic spread and suggest that the proximity of pelvic primary tumours to large central vessels could explain the observation that cardiac involvement is more frequent with pelvic than with distal femoral primary lesions. We are aware of at least one additional case reported since then.[2] Presumably, our patient experienced haematogenous tumour spread to the right heart from her primary pelvic tumour, and to the left heart from small pulmonary metastases that were suspected from her chest x-ray and CT scan but not formally diagnosed. Alternatively, her left heart might have been seeded through a small interatrial or interventricular septal communication which escaped our detection. Unfortunately, the rapid and aggressive spread of her disease foreclosed the possibility of any potentially life-prolonging intervention.

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Conflicts of Interest. None.

Ethical Standards. This work involved no human or animal experimentation, and we are unaware of any requirement for institutional ethics committee approval for publication of de-identified case reports. We did obtain the patient's parental consent for publication, and would be happy to include a statement to that effect pending your further advice.

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