

## Case Report

## Salvage One and One-Half Ventricular Repair and Resection of Ewing's Sarcoma of Cardiac Origin

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A 4-year-old boy with primary cardiac Ewing's sarcoma presenting with congestive cardiac failure is reported for its rarity. Its surgical importance is highlighted.

**P**rimitive neuroectodermal tumours and Ewing's sarcoma of cardiac origin are rare entities among primary cardiac neoplasms. Our extensive search of the literature revealed 3 cases of primitive neuro-ectodermal tumour of the myocardium and 2 cases of Ewing's sarcoma of cardiac origin, treated surgically.<sup>1-3</sup> We report another case of Ewing's sarcoma of right ventricular origin, treated by resection of the tumor mass, right ventricular endocardectomy, and salvage one and one-half ventricular repair (1.5 VR).

### Case presentation

A 4-year-old boy weighing 14 kg was admitted to the hospital with increasing dyspnoea (New York Heart Association Class IV) and palpitation of six months' duration. Significant findings included tachypnoea, tachycardia, *pulsus paradoxus*, hypotension, a loud pulmonic component of the second heart sound, and tender hepatomegaly. Chest radiography revealed cardiomegaly with a cardiothoracic ratio of 0.8 and normal lung fields. The electrocardiogram showed right axis deviation with right ventricular hypertrophy and non-specific ST-T changes in the precordial leads.

Two-dimensional Doppler echocar-

diography revealed a large mass obliterating the entire right ventricular (RV) cavity, global reduction of biventricular contractility (ejection fraction 0.20), and thickening of the interventricular septum without any significant valvular abnormalities, clot, or pericardial effusion. A metastatic workup, including abdominal/pelvic ultrasonography and computed tomography (CT) scan of the chest, was negative. Considering the patient's moribund clinical status, magnetic resonance imaging (MRI) and endomyocardial biopsy examinations were not undertaken and the patient was subjected to surgical exploration.

Intraoperatively, there was sanguinous pericardial effusion, with distended right atrium, right ventricle and *venae cavae*. On moderately hypothermic cardiopulmonary bypass (CPB), the heart was arrested using cold hyperkalaemic blood cardioplegia and the tumour was approached via trans right-atrial-pulmonary artery approach. There was a massive intracavitary tumour obliterating the entire RV cavity, variably infiltrating the RV free wall, interventricular septum and tricuspid chordopapillary apparatus (Figure 1A & B).

After its malignant nature had been confirmed by frozen section, the tumour mass was excised as radically as possible.

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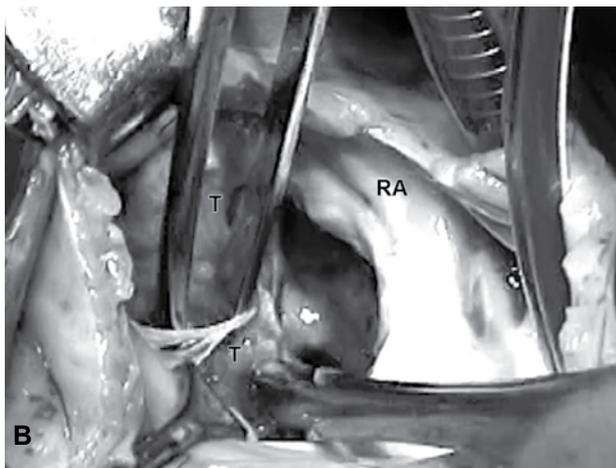
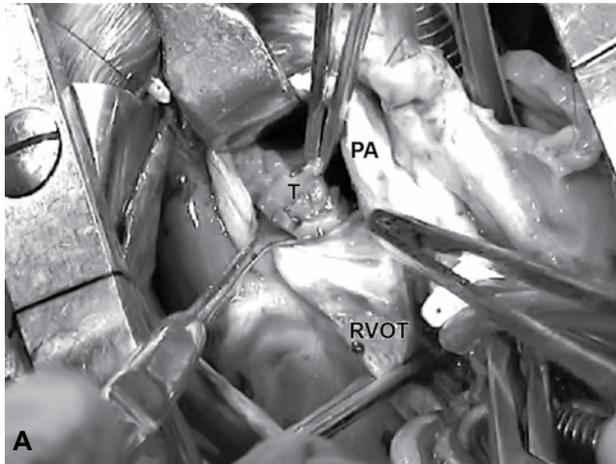
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**Figure 1.** A. Operative view from the pulmonary arteriotomy (PA) showing the tumour mass (T) occupying the entire right ventricular outflow tract (RVOT). B. A view from the right atriotomy (RA) showing the tumor mass (T) occupying the entire RV cavity and partially involving the tricuspid valve apparatus.

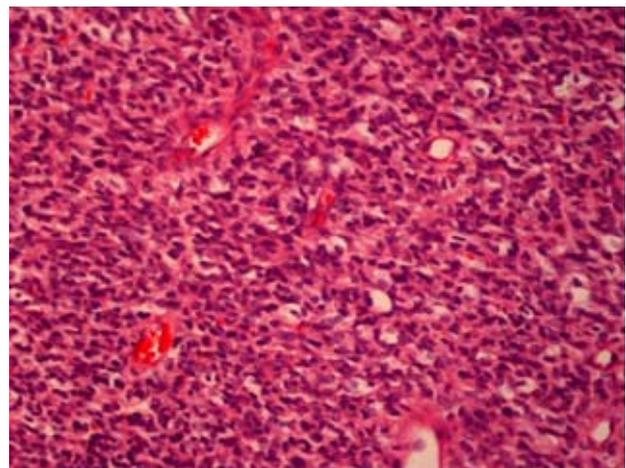
Following resection, we were unable to wean the patient off bypass in spite of management protocol. Consequently, we decided to add a bi-directional Glenn and convert into a 1.5 VR circuit, following which he was successfully weaned off bypass.

Postoperatively, he had stable haemodynamics with minimal inotropic support; mean pressures in the superior *vena cava* and right atrium were between 16-18 mmHg and 8-10 mmHg respectively. Transoesophageal echocardiography revealed unobstructed flow through the bi-directional cavopulmonary connection to the lungs, pulsatile flow from the RV to pulmonary arteries and grossly hypokinetic ventricles with an ejection fraction of 0.40.

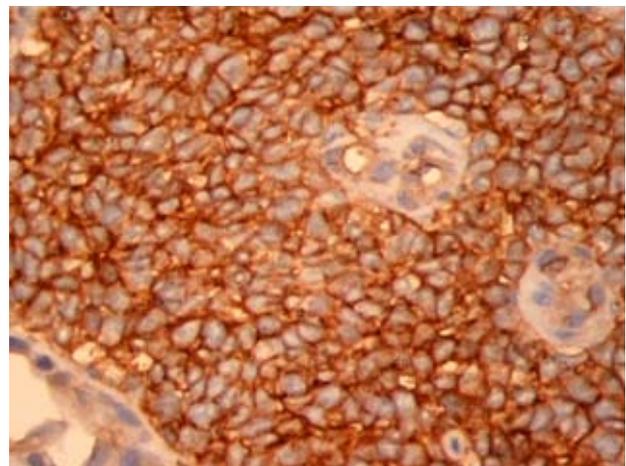
Following extubation on the first postoperative day, the patient developed intractable ventricular arrhythmias unresponsive to medical management

and succumbed thereafter despite all resuscitative measures. Permission for autopsy was denied.

Histopathological examination of the resected specimen revealed a tumour with islands and nests of relatively monomorphous small blue cells with round hyperchromatic nuclei and scanty cytoplasm, diffusely infiltrating the collagenous tissue (Figure 2). Rosettes were present. Immunohistochemistry showed strong cytoplasmic membrane positivity for CD99 in the tumour cells (Figure 3). Immunostaining for chromogranin, desmin, leukocyte common antigen and cytokeratin were all negative. Hence, a diagnosis of Ewing's sarcoma of cardiac origin was made.



**Figure 2.** Photomicrograph of the resected tumour (haematoxylin & eosin stain  $\times 200$ ), showing diffuse sheet of patternless, darkly staining, small malignant tumour cells containing scanty cytoplasm and inconspicuous nucleoli.



**Figure 3.** Photomicrograph of the resected tumour, highlighting the immunohistochemical pattern of the tumour cells with distinct, crisp membranous positivity for MIC2 (CD 99) antibody (original magnification  $\times 400$ ), characteristic of Ewing's sarcoma.

## Discussion

Primary cardiac neoplasms are rare, with an autopsy incidence of 0.001-0.28%, and 25% of these are malignant.<sup>1-3</sup> Angiosarcoma, rhabdomyosarcoma and fibrosarcoma are the most common cardiac neoplasms.<sup>1-3</sup>

There have been several reports of surgical excision of Ewing's sarcoma with cardiac metastases.<sup>1,4</sup> In two other reported cases of primary cardiac Ewing's sarcoma, the first patient expired intraoperatively after attempted surgical resection.<sup>2</sup> The second patient underwent sarcoma-based chemotherapy with 18 weeks of vincristine, ifosafamide, doxorubicin and etoposide, followed by 2 additional weeks of vincristine, doxorubicin and ifosafamide. Subsequently, the patient underwent resection of a primary Ewing's sarcoma with reconstruction of the superior *vena cava* using a bovine pericardial tube.<sup>3</sup> Our report not only demonstrates the third case of Ewing's sarcoma of cardiac origin but also the first case of resection of Ewing's sarcoma of cardiac origin with concomitant one and one-half ventricular repair.

The Ewing's sarcoma family of tumours includes Ewing's sarcoma, primary neuroectodermal tumour, neuroepithelioma and Askin's tumour, and is thought to derive from neural crest cells. In contrast to the osseous variety, the extra-osseous variety is more common during the second decade of life, is equally distributed between the sexes, and commonly affects the soft tissues of the trunk, retroperitoneum and extremities. Histopathological features include chromosomal reciprocal translocation (t11; 22) (q24; q12), cell surface antigen positivity, neuron-specific enolase positivity, well-defined Homer-Wright pseudo rosettes, sheets and lobules, cords and trabeculae, and spindled areas.<sup>4</sup>

Clinical manifestations are generally non-specific and are due to dysfunction of the myocardium, pericardium or valves.<sup>1-4</sup> Common clinical manifestations include congestive heart failure, myocardial ischaemia, thromboembolism, arrhythmias, or invasion of adjacent mediastinal structures. While the previously reported cases had a subacute history, with exertional dyspnoea, fatigue, weight loss, and symptoms of myocardial ischaemia, our patient was in a moribund state, with orthopnoea, hepatomegaly and congestive heart failure.

Improvements in diagnostic technology have increased the referrals for surgical management. Chest radiography frequently demonstrated cardiomegaly. Although echocardiography is the investigation of choice for initial evaluation and is sensitive in predict-

ing the aetiology of most intracavitary cardiac masses, it is less reliable in determining the nature of intramural or extra-myocardial lesions. CT and MRI are complementary to each other in determining the presence, site, and nature of a cardiac mass, in predicting extracardiac extension of the tumour and in establishing the amount of myocardial and great vessel involvement.<sup>5</sup>

The basic premise for the treatment of primary malignant cardiac tumours has not changed during the history of cardiac surgery. Resection is the only available form of curative therapy, debulking the tumour mass and relieving the pathway obstruction. Adjuvant chemo-radiotherapy may be helpful to palliate symptoms.

However, a borderline, dysfunctional, and dilated RV was an indication for the adjunctive approach of one and one-half ventricle repair in this patient, as described in our earlier publication.<sup>6</sup> Although extracorporeal membrane oxygenation is a useful tool to manage acute severe postoperative right ventricular failure, it was not used in this case because of the impression that the RV was inadequate to handle the biventricular repair.

In conclusion, an accurate preoperative diagnosis is desirable to allow planning of multimodality therapy, while the initiation of treatment should be expeditious for Ewing's sarcoma of cardiac origin. Decision making for 1.5 VR should ideally be done preoperatively, so as to reduce the cardiopulmonary bypass time and myocardial ischaemia. A wider appreciation of this entity is warranted.

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