Primary intracardiac germ cell tumor

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ABSTRACT

We present the echocardiography images in a 6 year old girl who presented with a history of scalp swelling after trivial trauma which was subsequently diagnosed as metastases from primary intracardiac germ cell tumour.

Keywords: Cardiac tumors, echocardiography, intracardiac germ cell tumor, magnetic resonance imaging, pediatric cardiac tumors

A 6-year-old girl presented with persistent swelling in the right side of her head after trauma 2 days ago. On physical examination, a soft swelling was palpated along the right parietal region on the head with a normal overlying scalp. Magnetic resonance (MR) imaging brain was done suspecting a cephalohematoma, however, it demonstrated multiple intracranial and extracranial enhancing soft-tissue lesions [right parietal; Series 1 and 3; Figure 1 and left frontal; Series 2 and 3; Figure 1] eroding inner and outer table of the skull. A possibility of metastases from unknown primary or multifocal involvement in lymphoma was suggested. On ultrasound abdomen (done to rule out abdominal masses such as neuroblastoma), no abnormal finding was seen. Echocardiography showed a large well-defined isoechoic mass lesion in the right ventricle (RV) involving the septum and the tricuspid valve. Tricuspid regurgitation was seen secondary to incomplete closure of the valve. A similar smaller lesion was also seen on the lateral wall of the RV [Figure 2]. Intracardiac primary versus metastases was considered differential. Contrast-enhanced computed tomography of the chest and abdomen to look for primary and skeletal survey revealed no other lesions apart from those previously described. Thus, a possible diagnosis of unknown primary with skull and heart metastases with a differential of primary cardiac mass with intracardiac and skull metastases was considered. Biopsy from the skull lesion showed immature mesodermal and endodermal cells with scattered rhabdomyoblasts, immature adipose tissue, and immature metanephrogenic cells. Immunohistochemistry showed moderate positivity for FLI-1, SALL-4, and CK and mild positive for CD56, CD99, desmin, myogenin, and WT-1. Routine blood investigations were normal. Serum beta-human chorionic gonadotropin (HCG) levels were slightly raised (5.5 IU/l; Normal value <5IU/l), lactate dehydrogenase (LDH) levels were very high (1833 U/L; Normal range:140-280 U/L):, and alpha-fetoprotein (AFP) levels were normal (2.8 ng/ml; Normal value:. <40ng/ml). Diagnosis of primary immature teratoma of the RV with rhabdomyoblastic and nephroblastic differentiation was made. The patient was started on chemotherapy of cisplatin, etoposide, and bleomycin. After three cycles, repeat serum makers were beta-HCG level <2 mIU/ml, LDH – 493 μ/l, and AFP – 1.97 ng/ml. Echocardiography on follow-up showed a significant decrease in the size of the septal lesion by approximately 60% with better movement of the tricuspid valve resulting in decreased intensity of tricuspid regurgitation. The lesion in the lateral wall of the ventricle was not seen. A positron emission tomography (PET) scan was done at follow-up to look for bone metastases, residual lesion was detected

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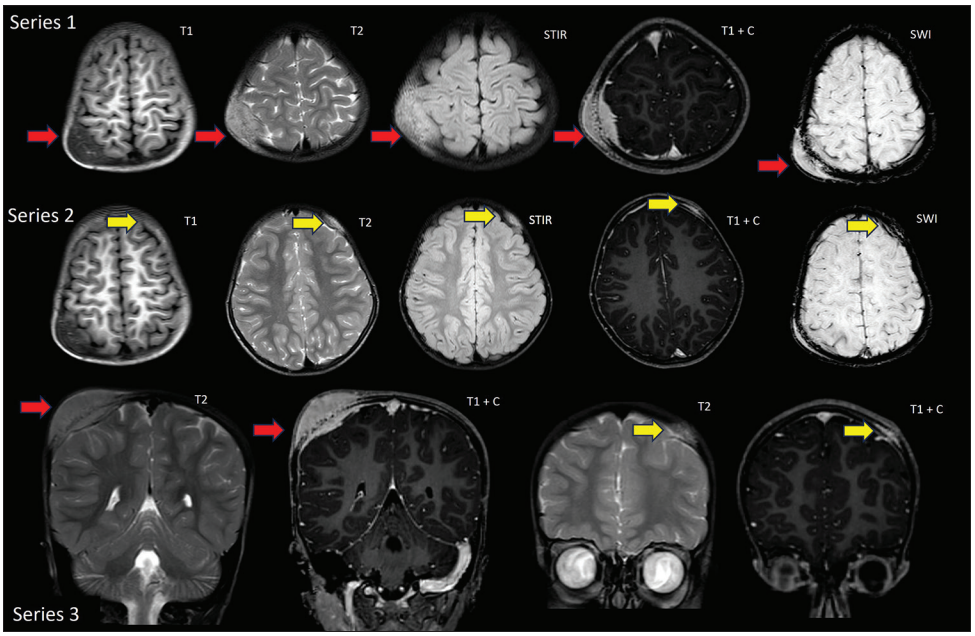

Figure 1: Sequential axial and coronal T1, T2, short tau inversion recovery, postcontrast T1 weighted, and susceptibility weighted image weighted magnetic resonance imaging images show multiple enhancing lesions in the skull (red arrow in Series 1 and 3) and (yellow arrow in Series 2 and 3). STIR: Short tau inversion recovery, SWI: Susceptibility weighted images, MRI: Magnetic resonance imaging

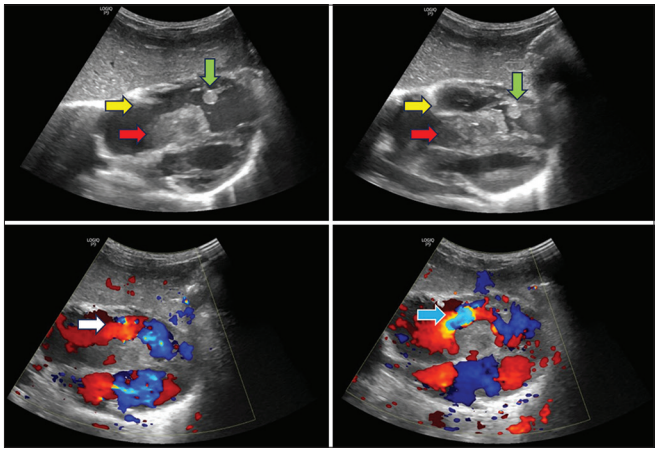

Figure 2: Echocardiography shows an echogenic mass along the septal wall of the right ventricle involving the septal leaflet of the tricuspid valve (red arrow). Another similar lesion is in the free wall of the right ventricle (green arrow). A free leaflet of the tricuspid valve is seen (yellow arrow). On color Doppler, forward flow is seen on atrial systole through the tricuspid valve (white arrow) and tricuspid regurgitation is seen as aliasing on the ventricular systole phase (blue arrow)

in the RV with maximum standardized uptake value of 4.23. No other metastatic lesions were seen.

Primary pediatric cardiac tumors are rare (prevalence of 0.0017 to 0.28 in autopsy series) and the majority of them are benign, and ~10% are malignant.[1] The most common cardiac tumor is rhabdomyoma (40%–60%) with teratomas being the second most common tumor with an incidence of 15%–19%.[1] Most teratomas arise from pericardium, unlike our case which was intracardiac and detected incidentally.[2] Symptomatic cases can present with outflow tract obstruction, cyanosis, respiratory distress, myocardial dysfunction, arrhythmias, and sudden death.[3] Most pericardial teratomas are easily removed surgically, however, surgery of intracardiac ones is more difficult, and hence chemotherapy is the mainstay of treatment.[4] Various studies have shown the utility of ^{18}F-fluorodeoxyglucose PET-computed tomography/PET-MR in such patients to know the extent of myocardial and pericardial involvement, to stage the tumor, and to assess the treatment response.[5]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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