Abstract: Ebstein’s anomaly (EA) is a rare congenital anomaly of the heart characterized by apical displacement of the septal and posterior tricuspid valve leaflet leading to atrialization of the right ventricle with variable degrees of malformation and displacement of the anterior leaflet. Importantly, atrial communications are present in 80% of patients with Ebstein's anomaly, through which a paradoxical embolus may occur. A case report of a 11 year old girl with Ebstein's anomaly presented with supra ventricular tachycardia (SVT) with right sided hemiplegia with UMN facial palsy in Pediatric department in Coimbatore Medical College Hospital, diagnosed as a case of paradoxical embolus causing left middle cerebral artery territory infarction.

Keyword: Ebstein's anomaly, paradoxical emboli.

INTRODUCTION

Ebstein’s anomaly (EA) is a rare congenital malformation of the heart characterized by apical displacement of the septal and often posterior tricuspid valve leaflets, leading to “atrialization” of the right ventricle with a variable degree of malformation and displacement of the anterior leaflet. The prevalence of EA is about 1 in 20,000 live births with no gender predilection and it accounts for less than 1% of all congenital heart disease. It is estimated that approximately 5% of the patients survive beyond the age of 50 years. The clinical presentation varies widely from asymptomatic to severe symptoms according to the degree of anatomic variation and heart failure. Severe disease leads to foetal or neonatal death. Mild forms are often undiagnosed until adulthood due to its benign course and survival has been reported until the ninth decade. We presented this case because paradoxical embolus with cerebro vascular accident (CVA) is a rare presentation in paediatric age group. Only few cases of Ebstein’s anomaly with paradoxical emboli have been reported in adults.

CASE REPORT

11yr old developmentally normal girl, 3rd born of non consanguineous marriage admitted with altered sensorium and inability to use right upper limb (UL) and lower limb (LL) for 2 days. Child had history of palpitation and vomiting on and off for past 1 week duration. History of bluish discolouration of fingers, toes and lips since 5 yrs of age and dyspnoea on exertion with easy fatigability since then. Further history revealed child was diagnosed as Ebstein’s anomaly at the age of 5yrs but not on any medications. Antenatal, natal and postnatal histories were uneventful. No history of maternal drug intake.

On arrival to emergency room, Child was in altered level of consciousness (ALOC), pain responsive. Respiratory rate was 24/min with adequate tidal volume. Heart rate (HR)-172/min, pulsevolume +/-, peripheries cool, BP:70/7 mmHg in left UL, liver span normal, SPO2 – 86% in all 4 limbs with 6l/min oxygen through mask, ECG showed supraventricular tachycardia (fig:1). Planned for DC cardioversion. Mean while adenosine was tried, which failed. Child was treated with synchronized DC cardioversion (0.5 Joules/kg) followed by amiadarone infusion. Rhythm reverted to normal. Shock was corrected with fluid bolus and ionotropes. Heart rate became 86/min, BP-100/70mmHg in left UL and 120 /70 mmHg in left LL and we could do complete cardiovascular examination which revealed central cyanosis with grade-3 clubbing. Apical impulse Lt 6th, Inter costal space lateral to mid clavicular line – hyper dynamic in nature, visible epigastric pulsation was present. Tricuspid area - S1 split, loud, Grade 3/6 Pan systolic murmur heard in lower left sternal border with multiple clicks. Mitral area – grade 3/6 middiastolic murmur heard. Central nervous system: Right side hemiplegia with ipsilateral upper motor neuron type facial palsy. Electrocardiogram (ECG) showed tall P-wave with wide PR interval (fig: 2). Chest X-ray showed right side heart enlargement (fig: 3). Echo showed Ebsteins anomaly with ostium secundum atrial septal defect (ASD) and severe tricuspid regurgitation (TR). MRI brain revealed acute infarct involving the perisylvian, left frontoparietal region, caudate nucleus, lentiform nucleus (fig:4) and MRA brain revealed occlusion of left middle cerebral artery and its branches. Lab parameters showed WBC and platelet count normal, Hematocrit- 47%, Hemoglobin -14.7gm/dl, ABG-normal, electrolytes, liver and renal function test were normal. Child was treated with aspirin and low molecular weight heparin and physiotherapy started. Child’s general condition improved. Child was put on oral amiadarone and aspirin, and referred to higher center for Ebstein’s anomaly correction and advised further follow up.
DISCUSSION

Ebstein's anomaly accounts for <1% of all congenital heart disease with equal sex preponderance. Pathology involving downward displacement of septal and posterior tricuspid leaflets associated with Tricuspid regurgitation and right ventricular outflow tract obstruction (RVOT), fibrosis and hypoplasia of right ventricle (RV). Left ventricular (LV) dysfunction is due to geometric distortion of LV, decreased end diastolic volume, increased fibrous tissue, decreased cardiac myocytes. Most commonly associated with Wolff Parkinson White syndrome (WPW) which leads to SVT. Other associated defect are pulmonary stenosis, pulmonary atresia, ventricular septal defect. 80% of patients have inter atrial communication.

Clinical presentation depends upon age at presentation, anatomical severity, hemodynamics, degree of shunting. Foetal & newborn period are more vulnerable. Neonate with severe disease present with severe cyanosis may show initial improvement due to fall in pulmonary vascular resistance (PVR) and develop cyanosis later. Infants present with cyanosis, congestive cardiac failure - tachypnoea, tachycardia and failure to thrive. Children with less severe disease are asymptomatic where congestive cardiac failure - tachypnoea, tachycardia and failure to thrive. Adolescences & adults as significant lesion present with persistent or intermittent cyanosis, dyspnoea, easy fatigability. Paradoxical emboli though rare presentation in adult with Ebstein's anomaly, it can occur in paediatric age as in our case. Some time it may be the initial presentation which may lead to the diagnosis of congenital heart disease. Surgical correction must be performed if the patient present with paradoxical embolism.

REFERENCES

6. Paradoxical embolism in Ebstein’s anomaly Cihangir Uyan, MD, Mehmet Yazici, MD, Ayten Pamukcu Uyan, MD, and Barbaros Dokumaci, MD Pediatric Department, Düzce Medical School, Abant Izzet Baysal University, Düzce.