REDELVE INTERNATIONAL PUBLICATIONS

Annals of Case Reports
Volume 2018, Issue 01
CASE REPORT: RD-CRP-10005

Infantile Scimitar and Its Associated Cardiac Malformation: A Tertiary Centre Experience in Tanzania

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Citation: *Theophylly Ludovick Mushi, Naizihijwa Majani, Sullende Kubhoja, Godwin Sharau, Stella Mongella, Viviani Mlawi, Zawadi Edward, Neema Muro, Alexander J Mroso, Abubakari Sudi, Frederic R Lymo, Pedro Parangyo (2018) Infantile Scimitar and Its Associated Cardiac Malformation: A Tertiary Centre Experience in Tanzania. Annals of Case Reports | ReDelve: RD-CRP-10005.*

Received Date: *23 November 2018;* ***Accepted Date:*** *28 November 2018;* ***Published Date:*** *28 November 2018*

Abstract

Background

Infantile scimitar syndrome is a rare pattern of congenital cardio-pulmonary anomalies comprise of anomalous pulmonary vein draining into systemic vein/ or right atrium, it is associated with different form of cardiac lesion has high mortality rate among infants.

Case presentation

We're reporting a case of 4 months old female in Tanzania complaints of recurrent respiratory infection and failure to thrive. A routine 2D-echocardiogram revealed anomalous pulmonary venous drains to inferior vena cava with another intra-cardiac lesion.

Conclusion

Scimitar syndrome is a rare congenital anomaly, associated with increased risk of morbidity and mortality early treatment before development of pulmonary hypertension offers good outcome.

Abbreviation

ASD : Atrial Septal Defect
CT : Computed Tomography
LPA : Left Pulmonary Artery
MPA : Main Pulmonary Artery

PAH : Pulmonary Arterial Hypertension
PDA : Patent Ductus Arteriosus
RPA : Right Pulmonary Artery
VSD : Ventricular Septal Defect

Keywords: Scimitar Syndrome; Pulmonary Hypertension

Background

Infantile scimitar syndrome is a rare pattern of congenital cardio-pulmonary anomalies comprise of anomalous pulmonary vein draining into systemic vein/ or right atrium in a child of less than 1 year old [1]. This account for 1 to 3 per 100,000 live births [2]. Frequently, associated with recurrent respiratory tract infection and premature death [3]. The word scimitar "Turkish sword" is derivative of a curved vascular configuration appearing on the chest-radiogram [4].

In developing countries, it is less documented, and its management imposes great challenge due to scanty of pediatric cardiac surgery series. In our setting, for the past three years out of 6000's pediatric population attended in our outpatient clinic, we're reporting a case of 2 months old female baby with recurrent respiratory infection and failure to thrive. Routine 2D-echocardiogram revealed anomalous pulmonary venous drainage which necessitated detailed evaluation, and henceforth scimitar syndrome was identified at first time in Tanzania.

Case Presentation

H.A.A is a 2nd twin product of premature birth at 36 weeks gestation age to a 48 years old female, P4 L3+2. During index pregnancy she had pre-existing hypertension at 30's weeks, later premature labor and intrauterine growth restriction (IUGR) for the 1st twin noted on routine Obstetric USS. She delivered first twin 1.8kg scored 8/10 (asymptomatic) and second twin 2kg scored 7/10.

The index child, who is a 2nd twin at age of 1 day of life she developed difficulty in breathing with desaturation in room air, which necessitated admission in neonatal ward for 1 week. She had persisted feature of respiratory distress and interrupted breast feeding, no bluish discoloration or convulsion was reported. 2-Dimension echocardiogram was done, and a child was diagnosed to have congenital heart disease at 7 weeks of life; due to features of congestive heart failure she was kept on furosemide 2mg BID and spironolactone 2mg OD orally; for reactive airway disease nebulization with salbutamol 2.5mg and budesonide 100mcg were given. Mother reported no history of consanguinity and no family history of congenital heart disease.

On arrival to our cardiology department she was not dysmorphic, alert, afebrile, not dyspnoeic, no jaundiced, no finger clubbing, had apparent cyanosis; she weighed 3.4 kg, height of 52cm. (Z score - 2). Blood pressure 98/54; PR – 134 b/min full and equal throughout, well perfused, RR 26 b/min, SPO_2 92% in room air. Reduced air entry on right, no crepitation, no rhonchi, apex beat was heard to the right with systolic murmur grade 3/6 on right lower sternal border, No hepatomegaly.

Echocardiography identified dextroposition of the heart, with leftward pointing apex, coatriatum, right atrial dilation with large secundum atrial septal defect, partial anomalous vein drains with right lower pulmonary vein draining to the inferior vena cava. Right ventricular

hypertrophy, large ventricular septal defect ~9mm. Normal pulmonary valves, confluent peripheral pulmonary with hypoplastic right pulmonary artery measuring ~ 3.5 mm:

CT scan cardiac confirmed dextroposition, large atrial septal defect and ventricular septal defect, partial anomalous venous connection (middle lobe vein into right atrium). Hypoplastic right lung with abnormal arterial supply (upper zones supplied by branch small right pulmonary artery 3.8 mm hypoplastic right pulmonary artery, mid and lower are supplied by branch from origin of superior mesenteric artery from celiac trunk (celiac-mesenteric trunk), aberrant retro-esophageal right subclavian artery from two vessel branching aortic arch.

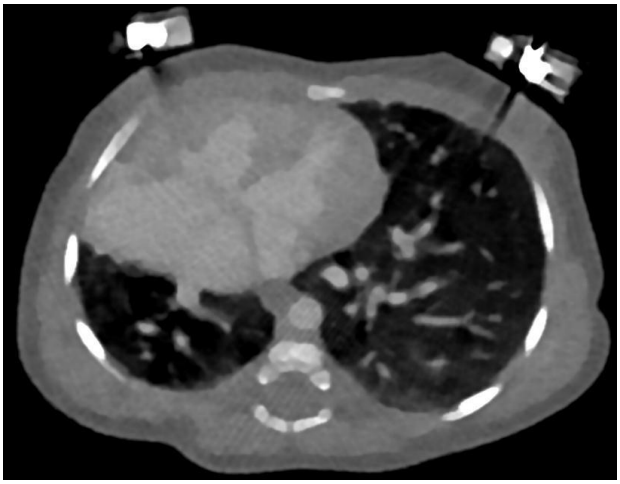

Figure 3: CT scan axial view showing dextroposition of the heart

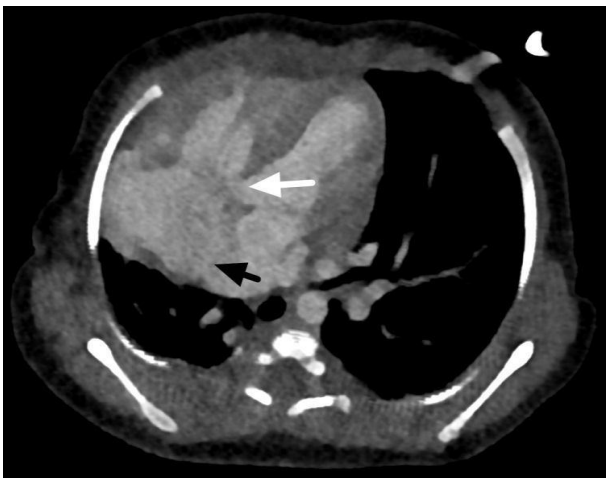

Figure 2: CT cardiac axial view showing Dextroposition of the heart, VSD (white arrow) ASD (black arrow), Hypoplastic right lung.

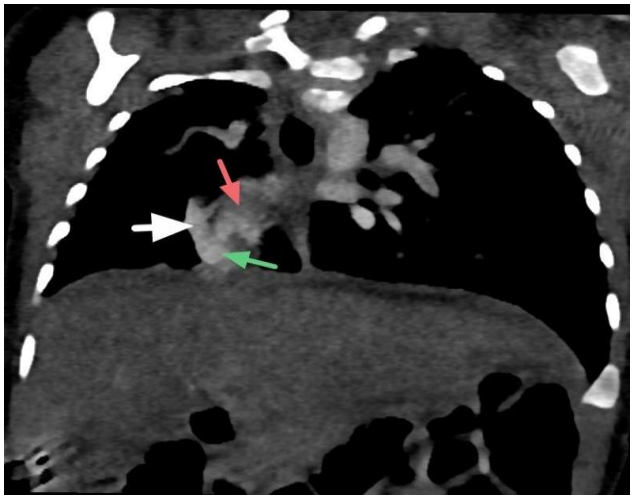

Figure 1: CT cardiac coronal reformatted view showing middle lobe pulmonary vein (white arrow) draining into inferior vena cava (green arrow).

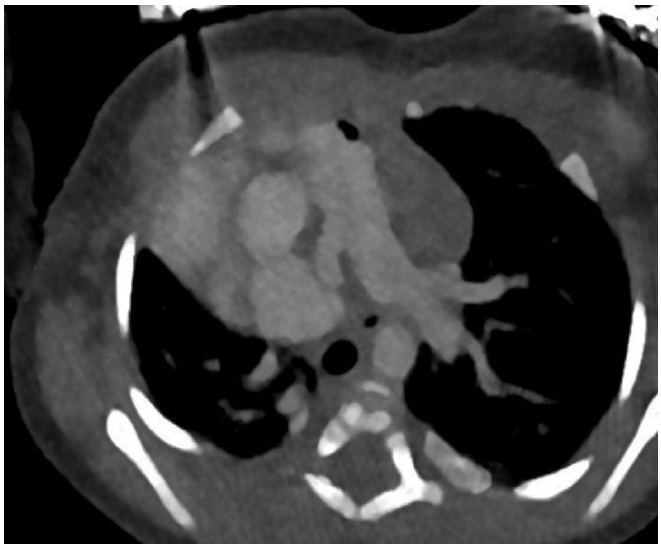

Figure 4: CT cardiac axial view showing MPA 1.23mm, hypoplastic RPA-3.8 supplying Rt upper lung zone (hypoplastic); normal (LPA-8.4mm) No (PDA) seen.

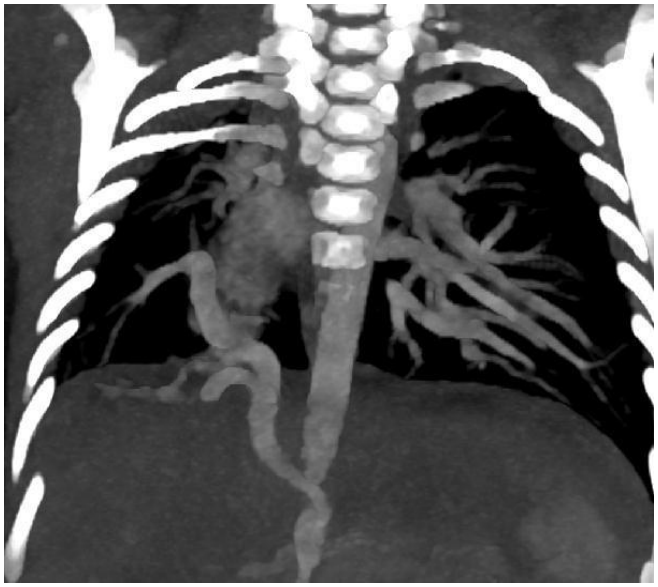

Figure 5: CT scan of the chest coronal reformatted view showing a systemic artery arising from common celiacomesenteric trunk supplying the right lower lobe.

Discussion

Due to unavailability of comprehensive pediatric cardiac care, patient was sent abroad for repair, at the age of 4 months and die 2 weeks after surgery from what was suspected to be pulmonary hypertension crisis. To our knowledge less number of studies has been reported on this syndrome in Africa. It is rare and occurs in both children and adults with varying clinical presentation with regards to age. Infants and young children suffer severe form of this disease and its usually fatal when compared to adults who preponderances are asymptomatic [4]. Our case, 7 weeks old child presented with tachypnea, respiratory distress, failure to thieve, and features of congestive heart failure like other studies done elsewhere [5,6]. Other studies have reported that diagnosis after age of 1 year is mainly incidentally found on chest radiograph and are associated with less severe clinical presentation [7].

Infants with scimitar syndrome have high rate of respiratory problems such as infection, wheezing and difficulty in breathing which was apparent to our patient [8]. Diagnosis should be suspected on chest radiograph with appearance of curved Turkish sword, which describes anomalous pulmonary vein draining to inferior vena cava and/or 2D echo with partial or anomalous veins draining to the right atrium [9]. A few studies have shown pulmonary hypertension being a severe complication of this condition and is associated with high mortality this is like other publication. Our child was in heart failure and already developed pulmonary hypertension, like other studies reported elsewhere [10].

Our patient had abnormal middle lobe pulmonary vein draining into right atrium leading to partial component of anomalous pulmonary return, which is like other study elsewhere. The right lung is mostly affected and is underdevelopment due to poor supply of blood which may results from pulmonary stenosis, hypoplasia of arterial and venous systemic circulation [11]. Nevertheless, association of other cardiac lesion such that atrial septal defect, ventricular septal defect, like other study reported elsewhere. Moreover, other congenital lesion has been reported, presented pulmonary sequestration, hypoplastic lung (right), dextrocardia, pulmonary

valve stenosis, coarctation of aorta (CoA), and patent ductus arteriosus, these features were similarly found into our patients except CoA, dextroposition instead of dextrocardia, and PDA.

In developed counties successful surgical repair for scimitar syndrome have been reported, on for reimplantation scimitar vein onto left atrium. Nevertheless, it has been associated with left pulmonary arteries stenosis and obstruction. High mortality has been associated with severe PAH, re-do for restenosis for some patients has been indicated. Others reported to be asymptomatic with their scimitar draining to left atrium [12]. In United State, less than 1 year had aorto-pulmonary collaterals and underwent surgery, despite other risk of development of PAH [13].

Scimitar syndrome is rare congenital anomalies, associated with an increased risk of morbidity and mortality, early identification is the key for early intervention before pulmonary hypertension develops (PAH) and other complications. Use of diagnostic tools such as Chest X ray, echocardiography, CT scan cardiac and Angiography are essential tools and are available for early diagnosis.

Conclusion

Clinician need to have high index of suspicion regarding the diagnosis as early treatment before development of pulmonary hypertension offers advantage in term of outcome.

Author's Contributions

TLM wrote initial draft of the manuscript, also together with TLM, GS, SM, VM, NM, AM, AS, PP, ZE took history, and performed physical examination. NM, SK performed echocardiography, FRL interpreted the all CT scan images. All authors reviewed and contributed to the final version of this case report. All authors read and approved the final manuscript.

Ethical Approval and Consent to Participate

Ethical clearance was granted from the directorate of research of the Jakaya Kikwete Cardiac Institute.

Consent for Publication

Written informed consent was obtained from patient's legal guradians for publication of this case report and any accompanying images.

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