A Pseudo-Cor Triatriatum in a 5-year old

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This is a case of the 5 year old female, asymptomatic during infancy, who presented with easy fatigability at 2 years of age. Work ups showed right ventricular hypertrophy on chest x-ray and ECG. Echocardiographic and cardiac catheterization results were compatible with Cor triatriatum sinister and partial anomalous pulmonary venous return with ASD secundum. However, intraoperatively, no fibromuscular membrane appreciated within left atrium was found. Patient underwent ASD patch closure with repair of partial anomalous pulmonary venous return. The left atrial membrane appreciated on two-dimensional echocardiography may represent the remnant of the common pulmonary vein, hence, it was not seen intraoperatively. Other possible diagnoses mistaken as Cor triatriatum sinister were also mentioned in the study. ***Phil Heart Center J 2008; 14(1):61-66.***

Key Words: ■ Cor triatriatum, congenital heart disease

Some movies are made for the viewers to think and analyze as the story unfolds. Moviegoers are hooked up with the story from the beginning to the end. In cardiac patients, no matter how definitive the pre-operative diagnoses are, intraoperative findings, sometimes, tell otherwise. Misdiagnoses arise when patient's clinical manifestations overlap among several differential diagnoses and that the intraoperative findings, therefore, becomes the gold standard in the diagnosis.

In this report, we presented a rare case of Congenital Heart Disease presenting as a left ventricular inflow obstruction. We discussed the embryology and anatomical variations of congenital lesions affecting the pulmonary veins and its incorporation to the true LA and presented the pitfalls in the diagnosis of the different anomalies that would present as in this particular case – obstruction to LV inflow.

Case

This is a case of a five year old female, born to a 27 year old G1P1 (1001) with unremarkable maternal history, delivered via CS secondary to cephalopelvic disproportion. There was no history of diaphoresis, recurrent respiratory tract infection, cyanosis and interrupted feeding infancy. At 14 months of age, patient had cough and colds. Routine chest x-ray revealed cardiomegaly, hence, she was referred to a Pediatric cardiologist. Likewise, electrocardiogram showed right ventricular hypertrophy. Initial 2D echo result revealed Congenital Heart Disease. Surgical management was advised. At 2 years of age, patient had episodes of easy fatigability while playing. Her parents were advised intervention, thus, she was admitted.

Pertinent physical examination on presentation, focusing on the cardiovascular system, showed the following findings: adynamic precordium, apex beat is at 5th ICS, LMCL, normal S1, fixed split S2, accentuated P2, Grade 3/6 pansystolic murmur at left lower sternal border radiating to the subxiphoid area, increasing with inspiration, and absence of cyanosis.

Work-ups done on our center showed presence of right ventricular hypertrophy, manifested on both chest x-ray (Fig.1) and ECG (Fig.2). Echocardiography study done revealed a membrane within the left atrium, superior to the left atrial appendage that separates into upper and lower chambers with the right upper pulmonary vein draining to the right atrium and interatrial communication between the membrane with left to right shunting, 0.5cm. With the following echocardiographic findings, the present working impression is cor triatriatum sinister, atrial septal defect with partial anomalous pulmonary venous return.

To further investigate the aberrant drainage of the right upper pulmonary vein, patient was subjected to a hemodynamic study 1 month prior to surgery. Oximetry studies showed normal saturation of inferior vena cava and superior vena cava. Mid right atrium has step up of 16%. There was a step down of 5% from right atrium to right ventricle probably due to the contribution of deoxygenated blood coming from coronary sinus. The increased saturation in the right atrium, right ventricle and pulmonary veins could be due to the

Accepted paper for PHC 16th Annual research Poster Competition 2008 and for 39th PHA Annual Convention held on May, 2008 at Edsa Shangrila Hotel, Philippines

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draining right pulmonary vein in the right atrium and the left to right shunting via the atrial septal defect. The left side of the heart has a normal saturation of 95% which was carried over to the peripheries. Pressures studies in the right side of the heart showed slightly elevated right ventricle systolic pressure of 40mmHg and right ventricle end diastolic pressure was normal at 7mmHg. MPA systolic pressure was slightly elevated at 44mmHg with mean pressure of 25mmHg which could be secondary to the increase flow coming from the anomalous draining right pulmonary vein and shunting via the ASD with QP: QS of 7:1. Pressure studies in the Left side of the heart revealed normal left ventricle pressure of 90/70 with left ventricular end diastolic pressure of 10mmHg and normal Aortic pressure of 90/60 with mean of 75mmHg. Right and Left pulmonary capillary wedge pressure were slightly elevated at 42mmHg with mean of 41 to 43mmHg which could be secondary to pulmonary venous obstruction. LPA cineangiography in LAO 30° Cranial 30° view showed opacification of right pulmonary artery both with good capillary blush and subsequent opacification of left atrium and a membrane was noted before the entrance of blood in the left ventricle. Right upper pulmonary vein cineangiography revealed the right atrium and coarse trabeculations of right ventricle and main pulmonary artery. Impression based on hemodynamic studies was Congenital heart disease, Cor triatriatum with partial anomalous pulmonary veins draining to right atrium; atrial septal defect, secundum, No Patent Ductus Arteriosus, Coarctation of aorta, Slightly elevated pulmonary artery pressure secondary to pulmonary venous obstruction.

Both the 2D echo and cardiac catheterization results reported this as a case of cor triatriatum, atrial septal defect with partial anomalous pulmonary venous return. Thus, surgical correction was recommended.

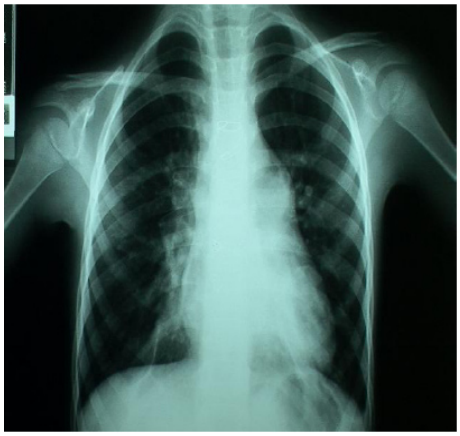

Figure 1. Chest x-ray revealed increased vascularity with right ventricular hypertrophy.

Intraoperatively, however, no left atrial membrane was noted. Patient underwent atrial septal defect closure and was discharged improved on the 10th hospital day. However, she was readmitted after 4 weeks due to pallor and easy fatigability and was managed as a case of Post Pericardiostomy Syndrome.

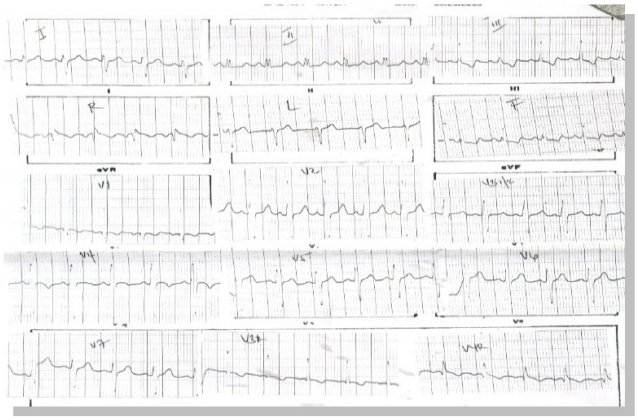

Figure 2. Electrocardiographic study showed sinus tachycardia, right axis deviation with right ventricular hypertrophy.

Discussion

Given these salient features, focusing on the acyanotic heart disease with increased pulmonary blood flow, Ventricular septal defect, patent ductus arteriosus and endocardial cushion defect were ruled out because they are characterized by left ventricular hypertrophy which was not found in the patient. Right ventricular hypertrophy are seen in cases of Atrial Septal Defect, Partial Anomalous Pulmonary venous return, Pulmonary Vascular Obstructive Disease and cor triatriatum sinister with atrial septal defect. (Fig 3). Pulmonary vascular obstructive disease was ruled out since patient did not have of chronic congestive symptoms and did not manifest the usual pruning of the pulmonary vessels on chest films.

Based on history, physical examination, chest xray and electrocardiographic findings, the present working impression is a Congenital Heart disease, with a left to the right shunt at the atrial level. Any or all of the differential diagnoses listed above may represent the patient's condition, thus echocardiography study was requested which revealed a membrane within the left atrium, superior to the left atrial appendage that separates into upper and lower chambers with the right upper pulmonary vein draining to the right atrium and interatrial communication between the membrane with left to right shunting, 0.5cm. With the following echocardiographic findings, the present working impression is cor triatriatum sinister, atrial septal defect with partial anomalous pulmonary venous return.

Oximetry studies were also consistent with Congenital heart disease, Cor triatriatum with partial anomalous pulmonary veins draining to right atrium; atrial septal defect, secundum, No Patent Ductus Arteriosus, Coarctation of aorta, Slightly elevated pulmonary artery pressure secondary to pulmonary venous obstruction. Cor triatriatum, described as a division or partitioning, of one of the atrial chambers, is a rare malformation, and when left untreated, may result into severe symptoms, even death. However, once appropriate surgical intervention was rendered, the life expectancy may be normal.[1]

To understand the pathophysiology of the disease, familiarization of the embryogenesis of the pulmonary veins as well as the incorporation to the atrium is a must. In the human embryo, the primordial of the lungs, larynx and tracheobronchial tree are derived by a division of the foregut. In their early stages of development, the lungs are enmeshed by the vascular plexus of the foregut, the splanchnic plexus. As pulmonary differentiation progresses, part of the splanchnic plexus forms the pulmonary vascular bed. At this stage there is no direct connection to the heart. Instead, the pulmonary vascular bed shares the routes of drainage of the splanchnic plexus. Subsequently the intraparenchymal pulmonary veins connect with the left atrium by establishing a connection with the common pulmonary vein, which evaginates from the left atrium. When the direct connection to the heart is established, the init communication between the pulmonary portion of splanchnic plexus and the cardinal and umbilicovite ine systems are, for the most part, obliterated. The p monary vascular bed then drains via four individ major pulmonary veins into the common pulmona vein, which in turn empties into the left atrium. T common pulmonary vein is a transient anatomic str ture. By a process of differential growth, it becon incorporated into the left atrium, resulting in the u mate anatomic arrangement wherein the four indiv ual pulmonary veins connect separately and directly to the left atrium. If the common pulmonary vein fails to develop or becomes atretic early in its development, collateral channels for pulmonary venous drainage are available in the form of primitive connections between the splanchnic plexus and the cardinal or umbilicovitelline systems of veins.

Van praagh diligently documented the embryogenesis of pulmonary veins and left atrium. In horizontal plane sections, at 27 days post ovulation, common pulmonary vein is a small, midline outgrowth from the dorsal wall of the still undivided primitive atrium. (Fig 4). It passes to the left of a prominent mass of sinus venosus tissue.

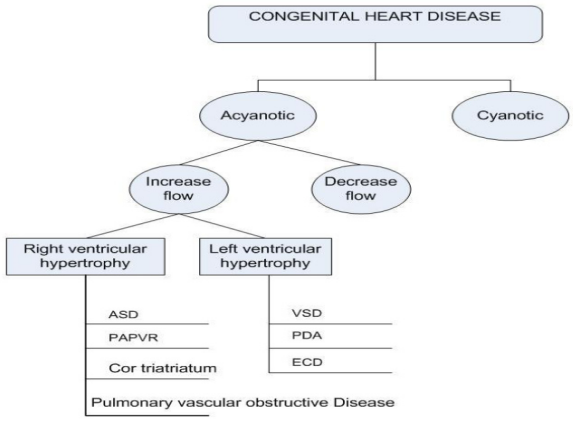

Figure 3. Algorithmic approach on the differential diagnoses for this case

Table 1. Comparative table of clinical manifestations and findings between the patient and the differential diagnoses

Symptoms	Patient	ASD	PAPVR	Cor Triatriatum
Easy fatigability	+	+	+	+
PE				
Fixed split S2	+	+		
CXR				
Increase flow	+	+		
RVH	+	+		
ECG				
RAD	+	+		
RVH	+	+		

Figure 4. Embryogenesis of pulmonary vein

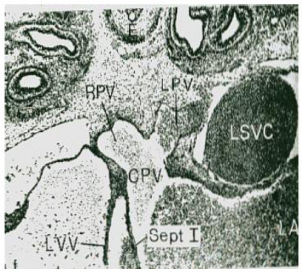

Figure 5. Separation of RPV and LPV

At 35 days post ovulation, incorporation of the common pulmonary vein into the left atrium commenced as well as the branching of the left and right pulmonary veins. (Fig. 5). Further branch incorporation progressed slowly and was still incomplete at 63 days post ovulation, as seen with the oldest embryo studied. In frontal plane sections, at about 33 days post ovulation, the pulmonary vein leaves the right, lower posterior corner of the left atrium passing beneath a relatively large mass of sinus venosus tissue and over the left horn of the sinus venosus, or the future coronary sinus. It runs rightward, inferiorly and dorsally then lies

beneath the plane of the septum primum, continuing dorsally in the midline. The mass of sinus venosus is the origin of the septum primum in the left and the left venous valve in the right and the common pulmonary vein normally grows out into the cardiac mesocardium through a narrow channel between the right and left sinus horns. After understanding the embryogenesis, three concepts of the morphogenesis of cor triatriatum were advocated. First, the malseptation hypothesis, describing the continuity of the pulmonary vein to the right of septum primum, instead of the left, which was initially thought to be normal development. Secondly, the malincorporation hypothesis, wherein the disturbance of the normal growth of the left atrium and that the subdividing diaphragm occurs in the posterior wall of the primitive left atrium. Third, the entrapment hypothesis, which suggests that the common pulmonary vein fails to incorporate normally into the left atrium, resulting in cor triatriatum because the pulmonary veins becomes entrapped by the relatively large mass of right horn sinus venosus tissue beneath which the vein runs early in its development. The left atrial ostium of the common pulmonary vein appears to get roofed in by the elastic tissue from the right sinus horn thereby preventing normal incorporation.[9] The atrial chambers have their similarities and differences. Both atria have a venous component, an appendage and a vestibule. Among the three, it is the structure of the appendage that distinct the morphological right and left atriums particularly the anatomy of its junctions with the venous component.[3] It is usual during fetal life for the right atrium to have sheet like valves to direct the richly oxygenated venous return from the inferior vena cava passing through the foramen ovale into the left atrium towards the developing brain. These valves may persists postnatally, commonly known now as Eustachian and Thebesian valves, which are anatomic substrate for division of the morphologic right atrium, giving rise to cor triatriatum dexter.[8]

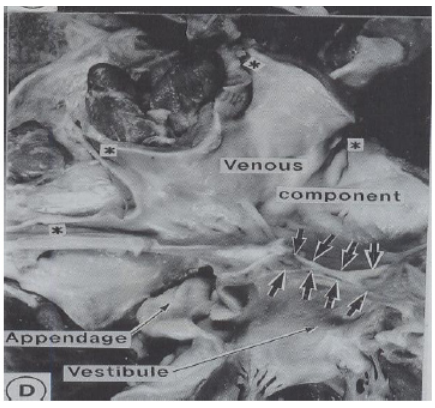

Figure 6. Fibromuscular shelf in LA

However, the left atrium has no counterpart valves in normal fetal development. But a literature had documented a fibromuscular partition between the venous component and the remaining left atrial segments. It usually extends obliquely across the cavity of the left atrium.[3] And this fibromuscular shelf (Fig. 6) is described as cor triatriatum sinister in echocardiography.

Clinical manifestations in cor triatriatum is directly proportional to the tightness of the communication between the proximal and distal chambers. In cases of tight stenosis, infants or children present with difficulty of breathing and frequent respiratory infections. Patients may remain asymptomatic until adulthood if the opening is less restrictive, but may present with dyspnea on exertion or frank hemoptysis. Cyanosis may be present in cases of right to left shunt through foramen ovale or atrial septal defect to the distal compartment of the divided left atrium. Partial anomalous venous connection, when present, relieves the cyanosis and may alleviate the degree of obstructive symptoms. The clinical signs are dominated by evidence of pulmonary venous congestion and pulmonary hypertension. Infants with restrictive opening may present with tachypnea, tachycardia, subcostal retractions, pallor, diaphoresis and interrupted feedings with good oxygen saturations. Physical examination of the heart revealed right ventricular heave, pulmonary component of the second heart second is almost always accentuated. No murmurs may be heard, although apical diastolic murmur mimicking mitral stenosis. A soft, blowing systolic murmur may represent secondary tricuspid regurgitation in severe disease. Once congestive heart failure develops, patients may manifest with pulmonary crackles and hepatomegaly.

Diagnostic modalities utilize in cor triatriatum include chest radiography, electrocardiography, echocardiography and cardiac catheterization. Chest radiograph typical of cor triatriatum presents with cardiomegaly, cephalization and left atrial enlargement. Right ventricular hypertrophy may be observed in the presence of chronic pulmonary congestion. Prominent aortic knob reflects pulmonary arterial hypertension. Right ventricular hypertrophy with an axis between 120 and 140 degrees are typical of electrocardiography of cor triatriatum patients. In echocardiography, the parasternal long axis view allows direct visualization of the obstructive partition, shows the sites of interatrial defects and recognition of any associated malformations, if present. The hemodynamic findings of cor triatriatum include an increase pulmonary wedge pressure in the presence of normal pressure tracings in the distal chamber. In selective pulmonary injection, there

is a prolonged opacification of the proximal chamber on follow through. The distal chamber is usually seen to contract vigorously during atrial systole, while the proximal chamber tends to contract poorly.

However, intraoperative findings in our case reported the following: two ASD secundum with intervening membrane, both the right upper and right lower pulmonary veins drained into the right atrium and no membrane was appreciated on the left atrium. This is just a case of atrial septal defect with partial anomalous pulmonary venous return. Patient underwent ASD patch closure with repair of PAPVR.

Two possible explanations may be deduced to possibly explain the discrepancy in the results between the 2D echo and intraoperative findings. A study described by Manning et al described the presence of the remnant of the common pulmonary vein mistaken as a left atrial mass.[6] Sadiq, on the other hand, reported a medially displaced and infolded left atrial appendage together with infolded left atrial wall which might give a false appearance of a left atrial membrane.[7] Other possible presentations of left atrial membrane appreciated on echocardiography were also reported. Benatar and company reported an infant with such a malformation associated with marked enlargement of the coronary sinus, which produced partial supramitral obstruction and consequently impairment to the left-ventricular inflow.[5] The patient presented with cardiac failure in infancy and features mimicking cor triatriatum. A case of a total anomalous pulmonary venous connection and restrictive foramen ovale, with left sided deviation of the atrial septum mimicking left sided cor triatriatum was reported by Atik et al.[4] Echolucency of the adenopathy due to sarcoidosis represented same echocardiographic findings as cor triatriatum was also reported by Walsh et al.[10] However, CT angiography revealed massive retrocardiac lymphadenopathy and normal cardiac anatomy hence cor triatriatum was ruled out.

There are other pitfalls in the diagnosis of cor triatriatum in 2d echocardiography and the most common is the inability to document the presence of partial pulmonary venous return in cor triatriatum which accounts in 15-25% of cor triatriatum cases. A study done by Wolf in 1986 has described a case of a 19 month old female whose right partial anomalous pulmonary venous return was not diagnosed until the post operative period.[11]

Dr Alwi et al describes supravalvar ring mimicking cor triatriatum, although this seldom occurs in isolation, commonly associated with congenital stenosis.[2] In cor triatriatum, continuous wave Doppler interrogation of the turbulent jet showed alternate interrogation of

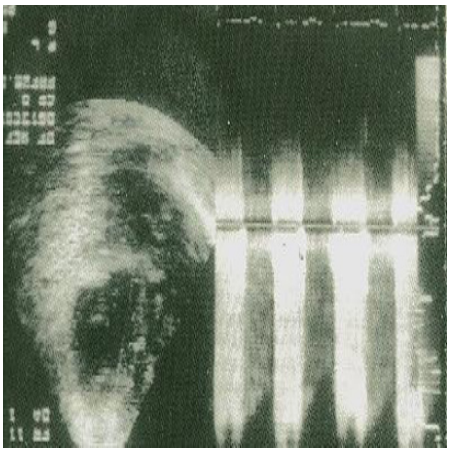

Figure 7. Continuous Doppler signals across the constriction

interrogation of the turbulent jet showed alternate bands of high intensity systolic signals in a positive direction producing a vertically striped appearance (fig 6).

In summary, we presented a case of a 5 year old female with episodes of easy fatigability since two years of age, right ventricular hypertrophy by x-ray and electrocardiography, with 2D echo and cardiac catheterization results consistent of cor triatriatum with partial anomalous pulmonary venous return. Intraoperatively, however, no left atrial membrane was noted. Patient underwent atrial septal defect closure, discharged improved on the 10th hospital day. The purpose of this presentation is serve as an eye opener that discrepancies in the diagnosis preoperatively and post operative may indeed happen and the team should always be prepared to face the management difficulties at is unfolds.

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