Scimitar Syndrome

Maria Consuelo Dolores Lapak-Tumaneng, MD; Eden D. Latosa, MD; Ana Maria A. Reyes, MD; Jaime S. Nuevo, MD

Background --- Scimitar syndrome, which presents with an anomalous right pulmonary vein that drains part or the entire right lung into the inferior vena cava (IVC) as its hallmark, is a rare association of congenital cardiopulmonary anomalies with an incidence of approximately 1 to 3 per 100,000 live births worldwide. It commonly presents with symptoms of respiratory distress and has an over-all mortality rate of 45%.

Case --- We present a case of a two month old male infant who presented with difficulty of breathing, for which he was mechanically ventilated. Chest radiograph showed dextroposed heart. Further work-ups, which included echocardiography, high resolution CT scan and hemodynamic studies, demonstrated the presence of an anomalous right pulmonary vein that drains into the inferior vena cava. Patient underwent ligation of the feeding vessels, which he tolerated well.

Conclusion --- This case illustrates the value of recognizing scimitar syndrome as one of the differentials in children who presents with respiratory distress. Although it is rare, timely surgical intervention improves the outcome of these patients. ***Phil Heart Center J 2008; 14(1):80-83.***

Key Words: Scimitar syndrome, Anomalous Pulmonary Venous Return

First described in 1836, Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies. The hallmark of Scimitar syndrome is an anomalous right pulmonary vein that drains part or the entire right lung into the inferior vena cava (IVC). Associated anomalies are variable and include hypoplasia of the right lung, dextroposition of the heart, hypoplasia of the right pulmonary artery (RPA), and anomalous systemic arterial supply from the aorta to the right lung. The term scimitar syndrome was coined because of the radiographic appearance of the anomalous vein, which appears as a tubular opacity paralleling the right cardiac border resembling a curved Turkish sword or scimitar. This is the so-called scimitar sign.1 Infants with this syndrome become markedly symptomatic after birth. They present with respiratory distress, cyanosis, failure to thrive, or cardiac failure. The patients receive a variety of medical and surgical treatments with an overall mortality of 45%.

Case

Our patient is a 2 months old male infant who was admitted at our center for dyspnea. He was born term to a 25 year old G3P3 mother with via caesarean section due to fetal distress after an uneventful pregnancy. Upon birth, the patient was acyanotic, with good cry, and good activity.

At one month of age, he was admitted at a local hospital for cough and was managed as a case of pneumonia. He was mechanically ventilated at the third hospital day due to respiratory distress.

During this confinement, he was referred to a pediatric cardiologist due to finding of dextroposed heart on chest radiograph and dextroposed heart with patent foramen ovale and moderate tricuspid regurgitation on 2-D Echo. He was transferred at our center for further evaluation and management.

His physical examination on admission showed presence of crackles on bibasal lung field, as well as presence of Grade 2/6 systolic ejection murmur at right upper sternal border.

He was admitted at the Pediatric ICU (PICU) and was hooked to a mechanical ventilator. CBC showed leukocytosis with predominance of segmenters, anemia, and thrombocytosis. Chest x-ray showed haziness in the left upper and right mid to lower lung field with ipsilateral deviation of the mediastinal structures. The left lung was hyperaerated and hypervascular, suggesting pneumonia and atelectasis, and pleural effusion, right. Piperacillin-Tazobactam was started. Salbutamol and Budesonide nebulizations as well as N- acetylcysteine IV were given. Electrocardiogram (ECG) showed right axis deviation and right ventricular hypertrophy. The P waves in Leads I, II and aVF are

Accepted paper for PHC 16th Annual research Paper Competition 2008

Correspondence to Maria Consuelo Dolores Lapak-Tumaneng , M.D. Department of Pediatric Cardiovascular Medicine. Philippine Heart Center, East Avenue, Quezon City, Philippines 1100 Available at **http://www.phc.gov.ph/journal/publication** ISSN 0018-9034

upright, hence, we say that the heart is dextroposed. Two dimensional echocardiography showed mesocardia, AV and VA concordance, intact interatrial and interventricular septa, tricuspid regurgitation moderate, PR mild, turbulent hypoplastic right pulmonary artery with PAP 75mmHg at the left upper pulmonic vein, right atrial and ventricular enlargement, with dilated main pulmonary artery. There was persistence of the atelectasis of the right lower lung despite treatment of antibiotics. High resolution CT- scan revealed consolidation on the left upper and right lower lobe with bilateral pleural thickening. A small right pulmonary artery is small compared to the left, and the pulmonary veins in the right lower lobe drain into the inferior vena cava. This time an anomalous pulmonary venous return was highly considered. Hemodynamic study showed O2 saturations of 86-88% in the IVC, 72% in the SVC and high RA, and 85% in the low RA with peripheral saturation of 98%. There was a significant O2 saturation step-up of 13% from the SVC to the RA indicative of left to right shunting at the atrial level. Oxygen saturation at the LLPV was normal at 98% which was carried over to the LA, aorta and to the peripheral artery. Pressure studies showed normal SVC and RA pressures. The RV systolic pressure was elevated at 45 mmHg. The systolic PA pressure was likewise elevated at 45mmHg indicative of RV and pulmonary arterial hypertension. Selective angiography of the MPA showed opacification of a hypoplastic RPA supplying the right upper lobe with subsequent drainage to the LA. The aortogram showed smooth opacification of the abdominal aorta feeding a large collateral that supplies a portion of the abdominal area as well as supplying the middle and lower lobes of the right lung. On levophase, this collateral drains into a vein that eventually drains into the IVC and the RA.

This constellation of findings points to a very rare anomalous pulmonary venous connection to the inferior vena cava, otherwise known as the Scimitar Syndrome.

The patient was then referred to TCVS. Ligation of the feeding vessels found in the thoracic cavity was performed through a R thoracotomy approach. During ligation of the feeding vessels, there was no change in the color of the involved tissue. No complication was noted intra- and post- operatively. The patient was discharged improved on the 60th hospital day.

Discussion

Anomalies of the pulmonary veins are uncommon and vary widely in their anatomic spectrum and clinical presentation, course, and outcome. A classification of pulmonary venous anomalies based on embryologic principles introduces a unifying concept to the considerations of these anatomically and physiologically diverse conditions. Scimitar syndrome is a rare association of congenital cardiopulmonary anomalies consisting of a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia, dextroposition of the heart, and anomalous systemic arterial supply to the right lung.[1]

Embryology

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 initial communication between the pulmonary portion of the splanchnic plexus and the cardinal and umbilicovitelline systems are, for the most part, obliterated. The pulmonary vascular bed then drains via four individual major pulmonary veins into the common pulmonary vein, which in turn empties into the left atrium. The common pulmonary vein is a transient anatomic structure. By a process of differential growth, it becomes incorporated into the left atrium, resulting in the ultimate anatomic arrangement wherein the four individual 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. Any one of these collateral channels may persist and enlarge. If only the right or left portion of the common pulmonary vein becomes atretic, persistence of the pulmonary venous- systemic venous connections of that side provides the etiologic basis for PAPVR. The most common type is to the right superior vena cava (SVC) and the second most common is to the right atrium. Though very rare, all the right pulmonary veins or occasionally the veins draining the right middle and right lower lobes enter the IVC either just above or below the diaphragm. The normal pulmonary venous pattern of the right lung is altered. This very rare malformation is termed the scimitar syndrome.

Incidence

This rare anomaly has an incidence of approximately 1 to 3 per 100,000 live births worldwide; the true incidence may be higher because many patients are asymptomatic.2 In the Philippines, in the past 10 years, three cases had been reported. The first patient, an eight year old female, was diagnosed at the Philippine General Hospital, an 8 year old female; the other is a 25 year old female at the St. Luke's Medical Center, and the third is our patient.

It has been reported most widely in adults and older children and is usually found during work-ups for dyspnea, fatigue, recurrent respiratory infections, or as an incidental finding on a routine chest radiograph. This adult form of scimitar syndrome usually is not associated with pulmonary hypertension and typically has mild symptoms and a benign prognosis. A second, infantile group of patients become symptomatic soon after birth or within the first 60 days of life. This is where our patient is classified. Their course is often complicated by severe pulmonary hypertension and cardiac failure. Pulmonary hypertension is secondary to a combination of conditions, including arterial blood supply from the descending aorta, stenosis of the anomalously connecting right pulmonary veins, pulmonary parenchymal abnormalities, and associated obstructive anomalies of the left heart and aorta.

Clinical Manifestations

Scimitar syndrome can have variable clinical manifestations. 3-5 Dupuis and associates stated in their study published in the American journal of Cardiology that the clinical presentations range from severe congestive heart failure in infancy to mild symptoms in childhood. They present with dyspnea, respiratory distress, and cyanosis. In a study done by Ching – Chia Wang et al published in the European Journal of Pediatrics, patients in the infantile form of Scimitar syndrome had severe respiratory distress that required ventilator support.

Classic findings on physical examination include a shift in heart sounds to the right and a systolic murmur. Auscultation of the lungs is usually normal, although breath sounds may be diminished on the right.

Pulmonary hypertension is often recognized as the cause of the severe symptoms and poor outcome of scimitar syndrome during infancy. In 1976, Dupuis and his colleagues reported to the Association of European Pediatric Cardiologists that severe pulmonary hypertension was a constant feature in symptomatic infants with scimitar syndrome. In a case report by Haworth et al published in the British Heart Foundation, all four patients with scimitar syndrome who manifested with failure to thrive, cardiac failure and cyanosis at 1-5 month of age, on cardiac catheterization studies indicated pulmonary hypertension, the systolic pulmonary and the systemic arterial pressures being similar.10 In a report published by Gao et al8 in the Journal of American College of Cardiology in 1993 on Scimitar syndrome in infancy, it has been stated that multiple factors may be responsible for the pulmonary hypertension. The size of the left-to-right shunt by the systemic artery collaterals may determine the severity of symptoms in infants. Studies have emphasized that the pulmonary hypertension may be caused by multiple factors including large left-to-right shunting by the anomalous pulmonary vein or other intracardiac lesions such as an atrial septal defect (ASD) or ventricular septal defect (VSD). Pulmonary venous stenosis is a well known cause of pulmonary hypertension in infants with total anomalous pulmonary venous connection and has been recognized as the source of pulmonary hypertension in some patients with scimitar syndrome. Pulmonary overcirculation can interrupt the normal postnatal regression of pulmonary artery muscularity, resulting in persistent pulmonary hypertension of the newborn. It may be difficult to determine which of these many factors are responsible for the pulmonary hypertension in an individual patient, so each factor should be considered and investigated. Unrelenting pulmonary hypertension can cause irreversible damage to the pulmonary vascular bed and lead to eventual right heart failure.

Diagnostic Modalities

The electrocardiographic (ECG) findings are comparable to those seen in uncomplicated ASD. The rR' pattern and the rSR' pattern are most commonly seen; peaked P and right ventricular hypertrophy of the systolic overload pattern occur in older patients exhibiting pulmonary hypertension.

Echocardiography may delineate both the scimitar vein as well as any systemic arterial supply to the right lung. The subcostal window is most useful in this condition. The presence of mesocardia or dextrocardia in 70% of patients and a smaller caliber right pulmonary artery compared to the left pulmonary artery are useful clues.

Cardiac catheterization and angiography are probably the most useful procedures for confirming the diagnosis and clarifying the exact anatomy and degree of pulmonary hypertension. The anomalous right pulmonary vein can be seen draining all or part of the right lung into the IVC. Oxygen saturations in the IVC and RA may be increased. The RPA is almost always hypoplastic, atretic, or otherwise abnormal in those infants with pulmonary hypertension. All these were seen in our patient.

An aortogram should also be performed to visualize the presence or absence of an anomalous systemic artery entering the right lower lobe.

The most striking radiographic feature in each of these patients was the dextroposition of the heart along with varying degrees of opacity of the right hemithorax. A small, opaque hemithorax generally implies volume loss. Mediastinal shift toward the opacified hemithorax with compensatory hyperinflation of the normal lung can be seen with atelectasis or pulmonary agenesis. Pulmonary hypoplasia can also cause a unilateral small lung with the heart shifted toward the affected side. The term dextroposition, rather than dextrocardia, is used when the heart is shifted into the right chest but the cardiac chambers maintain a normal relationship and the cardiac apex still points to the left. This dextroposition of the heart is secondary to right lung hypoplasia. In scimitar syndrome, the right cardiac border is often poorly defined, and the marked dextroposition of the heart along with the variable opacity in the right hemithorax frequently obscures the classic scimitar sign.

Management

Infants with scimitar syndrome who are symptomatic at an early age probably have pulmonary hypertension or congestive heart failure and require intervention to prevent irreversible pulmonary vascular disease or death. According to Huddleston et al in their report published in the Annals of Thoracic Surgery in 1999 entitled Scimitar syndrome presenting in infancy, in a compilation of 3 large series of patients, patients received a variety of medical and surgical treatments with an overall mortality of 45%.[9]

Medical management of the cardiac failure may require intubation and ventilatory support, sedation, digoxin or inotropic support, and diuretic therapy while the diagnostic workup is in progress and definitive therapy is being considered. Inhaled nitric oxide may be effective in stabilizing patients with pulmonary hypertension. Despite maximal medical support, patients with pulmonary hypertension will require surgical intervention to reduce the left-to-right shunting and relieve any pulmonary venous obstruction.

There are 3 primary surgical procedures used to treat infants with scimitar syndrome. These include occlusion of the systemic arterial collaterals, right pneumonectomy, and complete repair. Early reports focused on surgical ligation or catheter embolization of the aortopulmonary collaterals in an effort to reduce the left-to-right shunting. Right-sided pneumonectomy generally is reserved for those patients with pulmonary infarction, RPA atresia, and those patients in whom the scimitar vein cannot be transferred up to the right atrial wall for reimplantation. When possible, pneumonectomy is avoided as the right lung continues to contribute to gas exchange and the volume of the right lung may increase during the first 7 to 10 years of life. Early pneumonectomy can also lead to chronic respiratory insufficiency and severe scoliosis.

Transplanting the anomalous pulmonary vein carries a significant degree of surgical risk in both adults and infants. There are a number of reports of thrombosis of the anastomosis or occlusion of the baffle, which can lead to infarction of the right lung. Stenosis at the pulmonary venous anastomosis site is also common.

The postoperative course is often characterized by episodes of acute pulmonary hypertension. Acidosis, hypoxemia, hypercarbia, and hypothermia should all be avoided because they can contribute to the pulmonary hypertension. Mechanical ventilation with sedation and inotropic support are usually necessary. These episodes are often exacerbated during ventilator weaning, so the patient should be weaned slowly. Inhaled nitric oxide may also be useful in managing postoperative pulmonary hypertension. Lung transplantation may be an option for those patients with persistent pulmonary hypertension after complete repair because mortality remains very high in those cases.

Reference

1. Moss and Adams' Heart Disease in Infants, Children, and Adolescents 6th ed. 35: 736-751
2. Mordue Barbara C., A Case Series of Five Infants with Scimitar Syndrome. Adv Neonatal Care 3(3):121-132, 2003. W.B. Saunders
3. Ching-Chia Wang et al (2007). Scimitar syndrome: Incidence, Treatment, and Prognosis. Eur J Pediatr.
4. Takeda S, Imachi T, Arimitsu K, Minami M, Hayakawa M, Two cases of Scimitar variant. Chest 1994; 105; 292-293
5. Dupuis C, Charaf LA, Breviere GM, Abou P (1993). Infantile form of the Scimitar syndrome with pulmonary hypertension. Am J of Cardiology 71:1326-1330.
6. Sehgal A, Loughran-Fowlds A, Scimitar Syndrome. Indian J Pediatr 2005; 72: 249-251 (http://www.ijppediatricsindia.org/text.asp?2005/72/3/249/15134
7. Espinola-Zavaleta, Javita-Chavez, Munoz-Castellanos, Zamora- Gonzalez, Clinical and Echocardiographic Characteristics of Scimitar Syndrome. Rev Esp Cardiol 2006; 59: 284-288
8. Gao YA, Burrows PE, Benson LN, Rabinovitch M, freedom RM (1993) Scimitar syndrome in infancy. J Am Coll Cardiol 22:873-882
9. Huddleston CB, Exil V, Canter CE, Mendeloff EN (1999) Scimitar syndrome presenting in Infancy. Ann Thorac Surg 67:154-159
10. Haworth Sheila, Saue Ursula, Buhlmeyer Konrad (1983) Pulmonary Hypertension in Scimitar Syndrome in Infancy. Br Heart F; 50:182-9