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Coexistent Congenital Aortic Defects, Aneurysm of Sinus of Valsalva, Atrial Septal Defect and Infective Endocarditis: A case report

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ABSTRACT. Coarctation of the Aorta is frequently associated with bicuspid aortic valve. This is a risk factor for infective endocarditis. Aneurysm of a sinus of Valsalva is a rare defect with a prevalence of 0.09%. They are associated in 10% of cases with a bicuspid aortic valve and less frequently with coarctation of the aorta and atrial septal defect. It is extremely rare the association of coarctation of the aorta with an atrial septal defect. This is one of the first cases reported in Puerto Rico of an adult patient with coarctation of the aorta in association with a bicuspid aortic valve, a ruptured aneurysm of a sinus of Valsalva and an atrial septal defect. The patient is a 22 year old male with coarctation of the aorta diagnosed since childhood who was admitted at the Cardiovascular Center of Puerto Rico with signs of heart failure due to infective endocarditis secondary to a teeth infection. Upon evaluation with transthoracic and transesophageal echos, he was found to have a coarctation at the aortic isthmus, aortic root dilatation, bicuspid aortic valve with vegetation, severe aortic and tricuspid regurgitation, aneurysm of the non coronary sinus of Valsalva with perforation to the right atrium, biatrial enlargement and a dilated right ventricle. Successful antibiotic treatment of endocarditis was achieved followed by surgical replacement of the aortic valve and ascending aorta with closure of the non coronary sinus of Valsalva was done. An secundum atrial septal defect was found and was also closed. Surgical correction of the coarctation of the aorta was postponed for a future time. The patient had a successful postsurgical recovery and was discharged home with anticoagulation treatment.

Key words: Coarctation of the aorta, Bicuspid aortic valve, Infective endocarditis, Aneurysm of non coronary sinus of Valsalva, Atrial septal defect.

Coarctation of the aorta is a relatively common type of congenital heart defect. Its overall incidence is approximately 5-8% of all congenital cardiac disease (1). It predominates in males with a sex ratio as high as 3:1 (2). It may occur as an isolated defect or in the presence of other intracardiac and/or extracardiac lesions (1). The lesion most frequently associated with coarctation of the aorta is the bicuspid aortic valve with up to 85% of all cases. The valve may be functionally normal, stenotic or incompetent (2). There is a relationship between the bicuspid aortic valve and the inherent fragility of the aortic root which predisposes to dilatation, aneurysm formation and possible eventual dissection (2). These aneurysm formations may involve both the ascending aorta as well as the sinuses of Valsalva (2). It has also being reported aneurysms distal to the coarctation, in the descending aorta. The bicuspid aortic valve is also important because of its susceptibility to infective endocarditis. During the course of infection it may become grossly incompetent. In patients with coarctation of the aorta a bicuspid aortic valve is the most common site of endocarditis. Bacterial endarteritis at the site of the coarctation occurs less frequently (2). Infection in these patients tend to occur between ages 10 years and 40 years.

As indicated previously, aneurysms involving the sinuses of Valsalva may occur in patients with coarctation of aorta, particularly if a bicuspid aortic valve is present.. An aneurysm of a sinus of Valsalva is a rare defect with a prevalence of 0.09% and account for 1% of the congenital anomalies of the heart. Sixty five to 85% originate in the right coronary sinus, 10-30% in the non coronary sinus and project into the right ventricle or right atrium, leaving

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less than 5% originating from the left coronary sinus (3). In 10% of cases of aneurysms of sinus of Valsalva they are associated with a bicuspid aortic valve and less frequently with coarctation of the aorta and atrial septal defect (3). They occur primarily in males with a sex ratio of 4:1. Ruptured aortic sinus aneurysms typically express themselves in young men after puberty but before age 30 years (4).

Atrial septal defects, as mentioned before, are not frequently associated to aneurysms of sinus of Valsalva. The most frequent type is in the ostium secundum location, which is commoner in females with a sex ratio of at least 2:1 (5). Although ostium secundum atrial septal defects are among the commonest congenital cardiac malformations in adults accounting for 30-40% of naturally surviving patients over age 40 years (5), it is extremely rare in association with coarctation of aorta. The two most common shunt lesions associated with coarctation of aorta are: patent ductus arteriosus and ventricular septal defect (2).

This report describes a patient with coarctation of the aorta associated with a bicuspid aortic valve, aortic root dilatation, non coronary sinus of Valsalva aneurysm and a ostium secundum atrial septal defect who suffered a bacterial endocarditis.

Case Report

History

The patient is a 22 year old male with coarctation of the aorta diagnosed since childhood. Two years prior to admission he developed an infective endocarditis secondary to a tooth infection which did not receive adequate dental follow up. His endocarditis was treated in the United States where he lived at that time. Three months later the patient started to present symptoms of congestive heart failure for which he was started in Digitalis, Furosemide, Spirinolactone and Enalapril. He continued follow up with his primary physician. The patient remained clinically stable and seven months prior to admission he moved with his family to Puerto Rico. Six months later he developed progressive orthopnea, dyspnea upon exertion, non productive cough, chills, anorexia and general malaise which worsened significantly one week prior to admission. In view of these persistent symptoms he was brought to the Puerto Rico Medical Center for further evaluation and due to his clinical picture was hospitalized at the Cardiovascular Center of Puerto Rico and the Caribbean. Past history reveals that he smoked cigarettes (1 pack/day) and drank alcohol (2-3 cans of beer/day) for 1 ½ years but denied any use of illicit drugs. Family history is non contributory.

Physical Examination

The patient was alert, active, oriented in person, place and time with a temperature of 39.2ÚC, blood pressure of 125/60, heart rate of 104/min and respiratory rate of 20/min. There was no conjunctival petechiae nor Roth's spots. Jugular vein distension present with noticeable v waves. There was a left parasternal heave and thrill, apical impulse at midclavicular line in the fifth intercostal space. He was tachycardic with a regular rhythm, a systolic ejection murmur IV/VI was present at left lower sternal border, a diastolic murmur III/VI was present at right upper sternal border and a third heart sound was present at left lower sternal border. Lungs clear to auscultation. Normal abdominal exam. Lower extremities with bilateral pitting edema, faint femoral pulses (+1), adequate radial pulses (+2), no cyanosis, no clubbing, no Janeway lesions, Osler's nodes nor splinter hemorrhages.

Laboratory Studies on Admission

Biochemical data within normal limits. Hematological data showed hypochromic microcytic anemia, adequate white blood cell count and differential. Electrocardiogram revealed normal sinus rhythm, right axis deviation, first degree AV block, biatrial enlargement, right bundle branch block and biventricular enlargement. Chest film showed moderate cardiomegaly, prominent pulmonary arteries and vascularity, no consolidations and clear pleural spaces.

Course in the Hospital

The patient was admitted in the Cardiology ward where blood cultures were taken and afterwards was started in intravenous Vancomycin and Gentamicin. Infectious diseases service was consulted and agreed to initial antibiotic therapy. Initial treatment also included oral Digitalis, Furosemide, Enalapril, Spirinolactone and Ferrous sulfate. A transthoracic echocardiogram was done which showed biatrial dilatation, right ventricular dilatation, slightly decreased left ventricular systolic function, severe aortic regurgitation, a possible bicuspid aortic valve with vegetation, an enlarged aortic root, severe tricuspid regurgitation with moderate to severe pulmonary hypertension, tricuspid valve vegetation, a possible non coronary sinus of Valsalva aneurysm with perforation to the right atrium and severe coarctation at the aortic isthmus with a maximum peak gradient of 77 mmHg (Figure 1). In view of these findings a transesophageal echocardiogram was done which revealed right atrial enlargement, right ventricular enlargement, bicuspid aortic valve (Figure 2) with a vegetation, severe aortic insufficiency, severe tricuspid regurgitation with vegetation, enlarged aortic root, non coronary sinus of Valsalva aneurysm bulging into the right atrium (Figure 3)

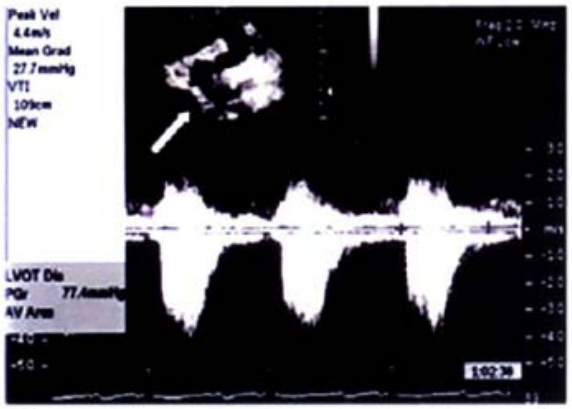

Figure 1. Suprasternal notch view during transthoracic echo showing coarctation of aorta flow gradient at the aortic isthmus (arrow).

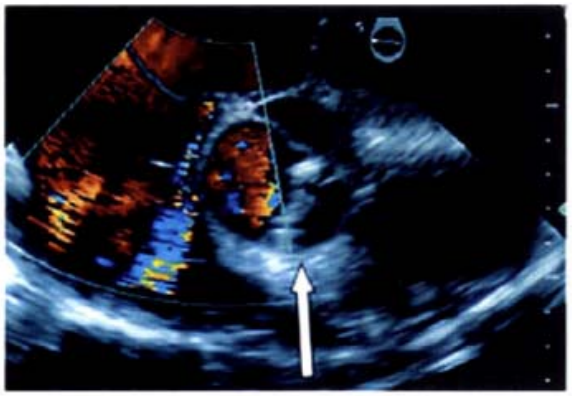

Figure 2. Transesophageal echo showing bicuspid aortic valve (arrow).

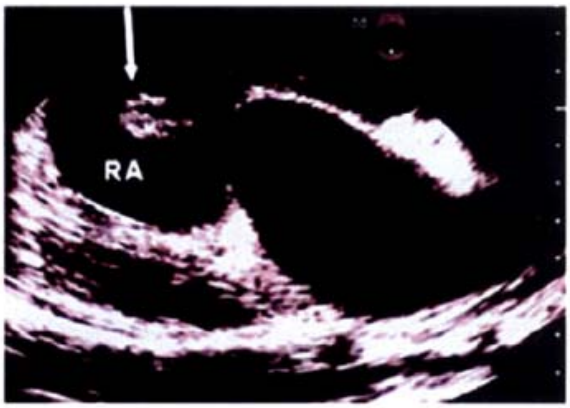

Figure 3. Transesophageal echo showing non coronary sinus of Valsalva aneurysm (arrow) projecting into the right atrium (RA).

and a negative bubble test for intracardiac shunts. On the fifth day of hospitalization the patient underwent dental evaluation which found two decayed teeth, one of them fractured. Both teeth were extracted. On the sixth day of hospitalization blood cultures results revealed the presence of Streptococcus mitis sensitive to current antibiotic treatment. The patient's symptoms promptly improved and remained hemodynamically stable throughout the course of antibiotic treatment. He was treated for 14 days with Gentamicin after which it was discontinued due to persistently increasing creatinine levels. Vancomycin was given for 25 days and afterwards treatment was discontinued due to still increasing creatinine levels and was continued instead with intravenous Linezolid for 17 additional days. After successfully completing 42 days of antibiotic therapy, cardiothoracic surgery evaluated the patient and on day 48 the patient underwent surgery. During the surgical procedure it was found a aneurysm

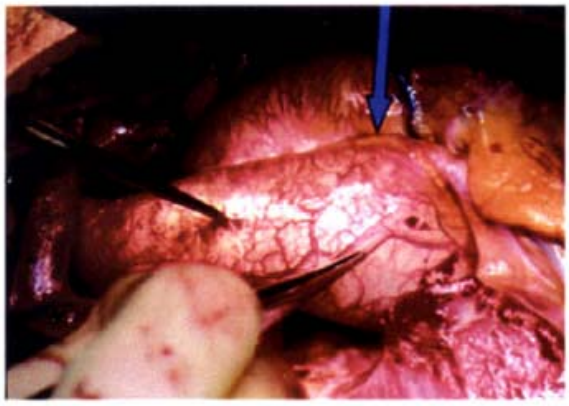

Figure 4. Aneurysm at sinotubular segment of aorta (arrow) visualized during surgery.

of the sinotubular segment of the aorta (Figure 4), a aneurysm of the non coronary sinus of Valsalva which ruptured into the right atrium, a bicuspid aortic valve with fenestrations in the non coronary cusp, a healed abscess in the right and non coronary cusp and subannular area and a 15 mm secundum atrial septal defect (Figure 5). No vegetations were found in the tricuspid valve. Replacement of the aortic valve and the ascending aorta was done with a 27 mm Carbomedics Conduit (valved) and direct closure of the secundum atrial septal defect was done as well as direct closure of the non coronary sinus of Valsalva after resection of the ruptured aneurysm. The procedure went uneventfully and the patient did well postoperatively without complications. A transthoracic echocardiogram on postoperative day 6 showed only mild aortic and tricuspid regurgitation with no significant changes in left ventricular systolic function. Anticoagulation treatment was started with Warfarin and was discharged home on postoperative day 10. Due to the

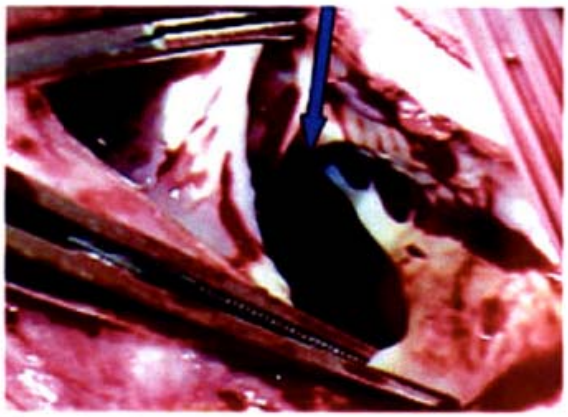

Figure 5. Fifteen millimeters secundum atrial septal defect (arrow) visualized from right atrium during surgery.

multiple procedures that had to be done during surgery, correction of the coarctation of the aorta was postponed for a future time.

Discussion

This 22 year old patient initially came to seek medical attention due to signs and symptoms compatible with an active infectious process, in this case an infective endocarditis, which due to his multiple cardiac defects contributed to the clinical picture of decompensated heart failure. The presence of a bicuspid aortic valve in addition to a very poor dental hygiene most likely were the determinant factors for the development of this infection. This was confirmed with blood cultures of Streptococcus mitis which is the most prevalent member of the normal flora of the upper respiratory tract and is a principal cause of endocarditis on abnormal heart valves (6).

Regarding the patient's physical findings upon admission it can be indicated that the left parasternal heave he presented is most likely due to the increased right ventricular impulse secondary to decompensated heart failure in the presence of coarctation of the aorta as well as due to an overloaded right ventricle secondary to the atrial septal defect. The latter could also explain the thrill that was detected in that same area. Large shunts with hyperkinetic right ventricular ejection into a dilated pulmonary trunk may cause a thrill (5). The presence of jugular vein distension and a third heart sound are signs of cardiac decompensation and of significant left ventricular dysfunction. He also presented with prominent v waves which are explained by the severe tricuspid regurgitation found in the echos which most likely is a result of right ventricular dilatation secondary to pulmonary hypertension (patient had a pulmonary systolic pressure of 65 mm Hg) caused by increased blood flow from the atrial septal defect. The systolic ejection murmur in the left lower sternal border is most likely due the previously mentioned significant tricuspid regurgitation. On the other hand, the diastolic murmur heard at the right upper sternal border is probably due to the severely regurgitant bicuspid aortic valve also diagnosed with the echos. The bilateral pitting edema in both legs is another sign of right sided heart failure. The decreased femoral pulses detected when compared with the radial pulses is characteristic of patients with coarctation of the aorta. Concerning the electrocardiographic findings in this case, the right bundle branch block and right axis deviation, are compatible with patients having secundum atrial septal defects (7). Also the patient presented a first degree atrioventricular block which can also be encountered in secundum atrial septal defects (5) as well as in patients with non coronary sinus of Valsalva aneurysms (4). The chest x ray revealed cardiomegaly and prominent pulmonary vasculature. The former is explained by the significantly dilated right ventricle and the latter by the prominent atrial shunt which eventually caused dilatation of the pulmonary vascular trunk.

The most significant aspect of this case is the fact that this patient had the extremely rare combination of multiple coexistent aortic abnormalities with an atrial septal defect. As indicated in the introduction, coarctation of the aorta may exist in the presence of other important intracardiac and/or extracardiac lesions. This is what is called a complex coarctation of the aorta. Among the several other lesions that may coexist with the coarctation are included two types of shunts, the patent ductus arteriosus and the ventricular septal defect. Upon reviewing the literature concerning this topic, no information was found regarding reported cases in which a shunt such as the atrial septal defect was found in association with coarctation of the aorta and even less in combination with aneurysms of the sinuses of Valsalva, aortic root dilatation and bicuspid aortic valve, all in the same adult patient. From the standpoint of adult cardiology, to the best of our knowledge, this is the first documented case of a patient with this particular combination of cardiovascular abnormalities in Puerto Rico.

Concerning the complex coarctation of the aorta present in this patient, the existence of a bicuspid aortic valve, a dilated aortic root and a aneurysm of the non coronary sinus of Valsalva protruding into the right atrium, supports the concept some authors have mentioned in previous works, that this might be due to congenital abnormalities in the aortic media (1). All these findings are most likely manifestations of a common underlying condition most

probably related to a generalized arteriopathy (1). This would explain why in these patients aneurysms with the possible complication of dissection may occur not only in the ascending aorta, but also distal to the coarctation, in the descending aorta and in the intracranial vasculature such as in the circle of Willis (berry aneurysms).

Finally, regarding the patient's management of these lesions, the bicuspid aortic valve had to be replaced in view of being severely regurgitant and since this was the site of one of the vegetations the patient had, which had turned into a healed abscess by the time of surgery. 13-22% of bicuspid aortic valves, develop regurgitation and 59-81% of the cases develop stenosis (1). Also 11-15% will acquire dilatation of the ascending aorta, like in this case, which may progress to an aneurysm (1). Due to this finding replacement of the ascending aorta was done with a valved Carbomedics conduit.

On the other hand, ruptured sinus of Valsalva aneurysms almost always must be surgically repaired. Surgical repair is highly successful with a ten year survival rate of 91-95% (3). If left unrepaired, chronic shunting and/or infection usually leads to death within 1-3 years (3). Ruptured aneurysms of the sinuses of Valsalva may contribute to cardiac chambers overload and the jet from the apex of a perforation may predispose to infective endocarditis (3).

In relation to the atrial septal defect, the overall life expectancy is decreased in patients in which the lesion is not repaired. Long-term exposure to chronic right heart volume loading can have deleterious effects such as atrial arrhythmia, irreversible pulmonary vascular disease, right heart failure and may also be a potential source of paradoxical embolus (7). The mainstay of therapy is closure of the defect by surgical or transcatheter techniques. Closure is recommended once diagnosed if there is evidence of a hemodynamically significant shunt or some other pathologic process. Although the indications for closure generally include symptoms, a significant left to right shunt (Qp:Qs > 1.7:1), pulmonary artery systolic pressure less than 70 mmHg or history of a cerebrovascular event, some feel that all atrial septal defects should be closed (7). Primary surgical closure has been the standard approach for many years. Depending on the defect size and location, it can be closed by primary suture, or if needed, by use of an autologous pericardial patch. Regarding transcatheter closure of the defect, it has become an attractive alternative to surgery despite the advances in minimally invasive surgical techniques. But this method of closure is suitable for patients with isolated secundum atrial septal defect (7). With the devices available today, defects with a resting diameter less than 30 mm may be considered.

Finally, concerning the coarctation of the aorta, surgery remains the therapy of choice particularly in neonates and infants (8). Three types of surgical repair have been used for its correction: resection of the stenosed segment with end-to-end anastomosis, use of a subclavian flap and patch aortoplasty. The best approach in terms of relief of obstruction and long-term outcome is the stenosed segment resection and end-to-end anastomosis with a 4% risk of recurrence and rarely late aneurysms formation. Survival rates of more than 90% at 10 years and 84% at 20 years have been reported (8). Early repair is usually aimed in order to minimize late mortality and morbidity. Increasing age increases intraoperative mortality rates as well as the association with coexistent lesions (1). The latter point was the main factor in deciding to postpone the correction of the coarctation after undergoing reparation of the other multiple cardiovascular defects present in our patient. Percutaneous balloon angioplasty is usually less effective than surgery and neonates and infants treated with this method experience high rates of recurrence (50-60%) and aneurysm formation (5-20%) (8). This modality is usually reserved for older patients and patients with recurrent coarctation. Although preliminary data suggest efficacy in native and recurrent coarctations, the role of stent implantation is still being defined (8). Indefinite follow-up is indicated after the diagnosis of coarctation of the aorta is established, especially after any type of mechanical repair. Key issues to be cognizant of include the progression of hypertension if it is present, onset or worsening of heart failure and development of coarctation recurrence in those being previously repaired. In older patients, hypertension commonly persists despite treatment by catheterization or surgery. As a result, medical treatment for adequate control of blood pressure and prevention of end-organ damage is an important goal. Also, like in this patient's case, a critical component of medical management that must be followed is prophylaxis for infective endocarditis and the orientation of adequate dental hygiene in order to avoid new infections.

Conclusion

Coarctation of the aorta is a relatively common congenital heart defect that may occur as an isolated defect or in association with other extracardiac and/or intracardiac lesions. Most commonly it is associated with bicuspid aortic valves and to a lesser degree to other aortic abnormalities such as aortic root dilatation and aneurysms of the sinuses of Valsalva. Even though coarctation of the aorta can be associated with shunts lesions such as patent ductus arteriosus and ventricular septal defects, it has rarely being reported to be associated with atrial septal

defects. This article presents a case report of a coarctation of the aorta with bicuspid aortic valve, aortic root dilatation and ruptured non coronary sinus of Valsalva aneurysm in association with a secundum atrial septal defect in a 22 year old male who developed bacterial endocarditis. The clinical picture, diagnostic findings and medical as well as surgical management have been summarized.

Resumen

La coartación de la aorta es un defecto congénito cardíaco relativamente común que puede ocurrir como un defecto aislado o asociado con otras lesiones extracardíacas y/o intracardíacas. Comúnmente es asociado con válvulas aórticas bicúspide y a menor grado a otras anormalidades aórticas tales como dilatación de la raíz aórtica y aneurismas de los senos de Valsalva. A pesar de que la coartación de la aorta se puede asociar con lesiones de comunicación como el ducto arterioso permeable y el defecto septal ventricular, rara vez se ha informado estar asociado con defectos septales atriales. En este artículo se discute un paciente con una coartación de la aorta con válvula aórtica bicúspide, dilatación de la raíz aórtica y ruptura de un aneurisma del seno no coronariano de Valsalva en asociación con un defecto septal atrial secundum en un varón de 22 años que desarrolló una endocarditis bacteriana. Se revisa el cuadro clínico, las pruebas diagnósticas y el manejo tanto médico como quirúrgico de estos pacientes.

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