CASE REPORTS

Multiple Systemic Emboli Complicating the Course of a Patient with an Atrial Septal Defect, an Atrial Septal Aneurysm and an Endocardial Right Atrial Pacemaker Lead

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ABSTRACT. We describe an adult patient with a large atrial septal defect, an atrial septal aneurysm and thrombus formation on a transvenous right atrial pacing lead. Because of right -to- left shunting through the atrial septal defect, she developed multiple systemic emboli to the spleen and left kidney (with infarcts), to the left leg, and probably to the brain as a cerebrovascular accident. The fundamental guiding principle of avoiding endocardial pacing leads in patients with congenital intracardiac communications, was violated in this patient, leading to serious dire consequences and complications. *Key words: Systemic emboli, Atrial septal aneurysm, Transvenous pacemaker lead.*

The general advice and counsel is that an endocardial pacemaker lead should be avoided in patients with intracardiac shunts or communications. Indeed, the presence of a right-to-left intracardiac shunt is stated to be an absolute contraindication for transvenous endocardial pacing because of the risk of paradoxical systemic emboli. Moreover, an intracardiac left -to- right shunt is regarded to be a relative contraindication for endocardial pacing, because of the possibility of incidental right-to-left shunting as shown by contrast studies and Doppler echocardiography.

Thus, this principle and guideline prevails because the potential exists for thrombus to cross the intracardiac communication and produce systemic embolic events and complications (1-6). We wish to report an adult patient with a large atrial septal defect (ASD) and an atrial septal aneurysm (ASA) in which this guiding principle was violated, resulting in serious systemic complications.

Case Report

This is a 65 year old woman, first seen at University of Puerto Rico Medical Center, Centro Medico, Rio Piedras, in 1983 at the age of 52 years. At the time she gave a history of a heart murmur since infancy, as well as multiple hospital admissions for episodes of easy fatigability, shortness of breath, and chest pain. She was on a medical regimen to treat congestive heart failure. Physical examination revealed a grade 2/6 systolic ejective murmur, with splitting of the second heart sound at the second left intercostal space. Serial electrocardiograms revealed right axis deviation, right bundle branch block and suggested right ventricular hypertrophy. There was also minimal first degree AV block (PR = 0.24 seconds). Although an echocardiogram was "normal" a clinical diagnoses of an ASD was entertained and a cardiac catheterization was scheduled. This revealed a step-up in oxygen saturation at the low right atrium, calculated shunt 1.0. However, the final official diagnoses after cardiac catheterization and electrophysiologic studies were: normal coronary arteries and left ventricular function, a patent foramen ovale (PFO), mild pulmonic stenosis, sick sinus syndrome, AV node dysfunction and infra-His block (although the data was compatible with an ASD). The patient required implantation of an epicardial DDD (Medtronic 7000A)* pacemaker, which was implanted with difficulty after dissecting through a fat pad. Examination by the surgeon revealed thick adipose infiltration (1.2 cm) of the ventricular surfaces and most of both atria.

Over subsequent years the patient remained with symptoms compatible with mild heart failure.

In 1985 and 1991, she received two new epicardial pacemakers secondary to malfunctioning and pacemaker

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end -of-life, respectively. In 1995 she received another new pacing system, this time an endocardial Medtronic Elite II 7086 DDDR,* with a 4504 atrial lead, and a 4024 ventricular lead. A chest X-ray was compatible with congestive heart failure. An echocardiogram revealed diastolic dysfunction of both ventricles, mild pulmonary stenosis, a thick right ventricular wall and possible effusion versus pericardial fat. In May 1997, at the age of 66 years, during a scheduled follow-up visit, the patient continued with symptoms of congestive heart failure. She related increasing shortness of breath, easy fatigability on exertion and general weakness. On examination there was a harsh grade 2/6 systolic murmur at the left sternal border. A transesophageal echocardiogram (TEE) was performed which revealed an atrial septal defect (secundum type) with a bi-directional shunt as well as some gradient across the pulmonary valve.

The finding of an ASD was confirmed by a cardiac catheterization which revealed normal coronary arteries, mild left ventricular dysfunction, and an ASD with a calculated Qp/Qs of 1.15. The patient was scheduled for surgical evaluation as an outpatient to electively repair the ASD. However, two weeks later the patient presented to the emergency room with sudden onset of severe right upper quadrant abdominal pain, nausea, vomiting, and diarrhea. An abdominal CT scan was compatible with left renal, as well as splenic infarcts. The patient was admitted and treated with full anticoagulation (IV Heparin) and IV antibiotics. However, five days later she developed a painful, pulseless left leg requiring an emergency arteriogram, which confirmed acute arterial embolization and led to an emergency femoral-popliteal embolectomy. On examination there was now a grade 3/6 harsh systolic murmur over the precordium, especially at the base. Following the embolectomy, she was transferred to the Cardiovascular Center of Puerto Rico and the Caribbean where a repeat TEE revealed an ASD (secundum type) with a predominant left to right shunt, as well as a very large thrombus lodged around the pacemaker lead in the right atrium (Figures 1 and 2).

Finally, on June 27, 1997, the patient underwent open heart surgery to repair the ASD. Intraoperative findings included dense hard adhesions, a 2.0 cm secundum ASD with right -to- left shunt by Bubble test, an endocardial lead with thrombus attached, and several cut epicardial leads. The surgery was complicated by a tear of the anterior aspect of the right ventricular wall which required circulatory arrest and a right ventriculotomy. The ASD was successfully closed, the epicardial leads were severed and the endocardial leads (including thrombus) were removed. A new epicardial pacemaker

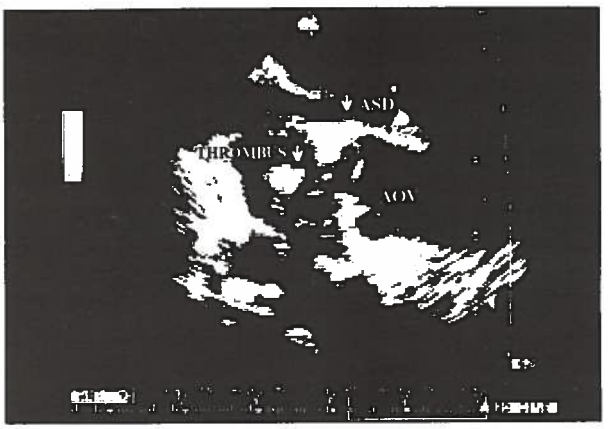

Figure 1. TEE showing thrombus and ASD.

was implanted (Intermedics 439-07 VVIR)†. Several days post operatively she suffered a stroke which suggested an embolus to the left middle cerebral artery. However, she recovered uneventfully and was successfully discharged to home. The patient has been followed regularly at clinics and her condition has improved.

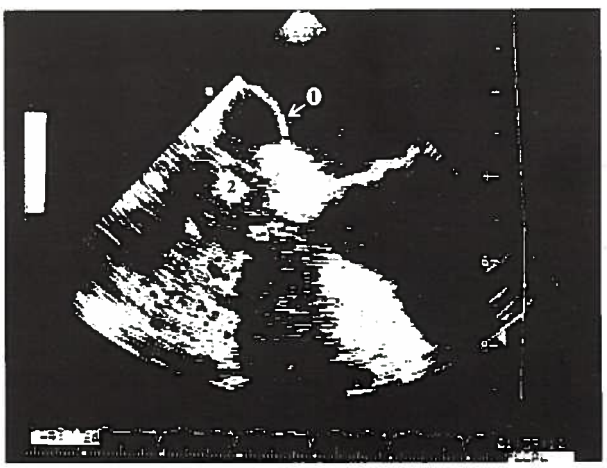

Figure 2. TEE showing: atrial septal aneurysm (1) and lead thrombus (2).

Discussion

The general counsel is that in the presence of an intracardiac congenital heart defect, an endocardial pacemaker lead should be avoided (1-5). This principle and guideline is appropriate and pertinent in this context because there exists the potential for small emboli to cross the defect, as well as right -to- left shunting, which may produce adverse events and complications in the arterial circulatory system. There exists the potential for right to left intracardiac shunting in the presence of a septal defect, even when paradoxic shunting would not be predicted on the basis of the peak pressure ratios between

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the two ventricles. It is well - known that a minute right -to- left shunt exists in the patient with an ASD (5,6).

Large right atrial pericatheter thrombus or clot is an infrequent to rare complication of permanent endocardial pacing. Right atrial thrombi tend to form on foreign bodies such as pacing leads, catheters and shunts. Silicone rubber (Silastic) pacing leads are more thrombogenic than polyurethane leads (7-13). Right atrial pericatheter thrombus formation may present as an incidental echocardiographic finding, or with various serious complications (7-13), as listed in Table 1.

Table 1. Complications of Pericatheter Right Atrial Thrombus

Silent or overt recurrent pulmonary emboli
Pyrexia of unknown origin
Sudden death, Syncope, Chest pain, Dyspnea
Hemodynamic impairments
• Severe persistent refractory congestive heart failure secondary to right atrial, tricuspid valve, and right ventricular inflow obstructions
• Right sided venous thrombosis
• Infection of the thrombus and endocarditis

Severe refractory congestive heart failure in spite of appropriate medical therapy in a patient with a permanent transvenous lead should call attention to the possibility of a right atrial thrombus (9). Clinically apparent pulmonary emboli as a complication of permanent transvenous pacing leads occurs in 0.2- 2%, usually less than 1%, of reported cases. But, clinically silent, asymptomatic pulmonary emboli may occur in a much higher incidence (9,11). Predisposing factors for the formation of right atrial clots on endocardial leads are the presence of congestive heart failure (75% of patients in one study), damage to the subclavian venous wall, immobilization and paroxysmal atrial fibrillation (9,11). Right atrial thrombi in this scenario carry a high mortality rate, up to 75%, especially when associated with clinically overt pulmonary emboli or right heart inlet obstruction (9,11). The diagnosis of right atrial thrombus formation on pacing leads can be confirmed by transthoracic 2-D echocardiography (TTE), and even better by transesophageal echocardiography. Angiography and venography may also be applicable (7-13).

Treatment for pericatheter right atrial thrombus consists of anticoagulation with heparin followed by chronic warfarin and dipyridamole therapy. Thrombolytics such as urokinase, streptokinase and prolonged infusions of recombinant tissue-type plasminogen activator (rTPA) have been utilized. Antiplatelet therapy should also be considered. Atriotomy and removal of the thrombus, lead, and hardware under cardiopulmonary bypass, followed by anticoagulation, may be indicated for those patients who fail anticoagulation or those warranting more aggressive therapy, as was applied in our patient (9,11,12,13).

Our patient was found to have an atrial septal aneurysm in association with the ASD. Autopsy studies suggest a 1% prevalence of ASA's. Transthoracic 2-D and transesophageal echocardiography yield a prevalence ranging from 0.08% to 10%. An association exists between ASA's and interatrial communications, either ASD or patent foramen ovale (PFO). In studies of patients with ASA's, 3% to 15% also have an ASD. By TTE, an ASD was found in 13% and 15% of cases; by TEE an additional ASD was reported in 9% and 12.5% of ASA patients (15-16, 18-19). Moreover, there is a high prevalence (most patients) of right -to- left interatrial shunting in patients with ASA's, either with an ASD or a PFO. With this and a Venturi effect, hypoxemia can exist. Thrombosis may occur on the aneurysm. Thus, patients with ASA's and right -to- left interatrial shunting are at risk for cardiogenic emboli. A number of recent studies have found an association between ASA's and cerebrovascular events of embolic origin, including transient ischemic attacks and cerebrovascular accidents/ strokes, as well as an association with other systemic embolic events. Indeed, cardiogenic emboli have been reported in 20% to 85% of these patients. These cardiogenic emboli may produce cerebrovascular events, as well as embolic events to the abdominal vessels and peripheral sites (often to the arteries of the lower extremities) (14-16,19). ASA's may be diagnosed by 2-D TTE, with contrast injection, and by TEE. But, even TEE may miss up to 47% of these aneurysms, even those that protrude more than 10 mm into the atrial cavity (14-19).

However, the general guiding principle of avoiding endocardial pacing in the above described patients may not always prevail or be correct. Cusimano and associates (20) in a recent abstract, concluded from their study of 148 adult patients with congenital defects and permanent pacemakers, (15 of these patients had both transvenous pacing and persistent intracardiac shunts) that permanent transvenous pacing may not necesarily be contraindicated in patients with persistent intracardiac shunts.

Nonetheless, in general, based on the overall reported unfavorable experience, it would be prudent to avoid the use of endocardial pacing in a patient with an endocardial communication and right -to- left shunting, or the

potential for right -to- left shunting. Most authors consider this to be a contraindication for an endocardial electrode. This is because of the already described great potential for systemic embolization. Also, newly placed electrodes are potential sources of small particulate matter with the risk of subsequent embolization until endothelialization occurs. This guiding principle must also be observed in patients with the potential for right -to- left shunting, even if their net intracardiac shunt is left -to- right. Children with ASD's and ventricular septal defects can show right -to- left shunting in the setting of elevated ventricular pressure, even with a net left -to- right shunt (1-6).

Conclusion

The risk of paradoxic embolism in patients with recognized or potential right -to- left shunting may limit the utilization of the preferable endocardial pacemaker lead. Therefore, the presence of an intracardiac shunt, in general, is a contraindication for permanent transvenous pacing (1-6). This fundamental principle was violated in our adult patient with a large ASD, in whom a transvenous endocardial pacing lead was placed. She developed a critical illness due to thrombus formation on the right atrial lead, which resulted in right -to- left paradoxical systemic embolic events to the spleen and kidney (with infarcts), as well as to her left lower extremity and probably to the brain.

Resumen

Describimos a una paciente adulta con un defecto grande del septo interauricular y un aneurisma del septum interauricular, con la formacion de un trombo en el cable del marcapasos transvenoso localizado en la auricula derecha. Debido a un cortocircuito de derecha a izquierda a traves del defecto del septum interauricular, esta paciente desarolló múltiples episodios embólicos a el bazo y el riñón izquierdo (con infartos), a la pierna izquierda, y probablemente al cerebro con un presunto accidente cerebrovascular. La guia principal y fundamental que establece como norma el evitar colocar cables de marcapasos en el endocardio de pacientes con comunicaciones intracardiacas congénitas fue violada en esta paciente, lo cual conllevó unas serias consecuencias y complicaciones graves.

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