CASE REPORTS

Isolated Gonococcal Pulmonary Valve Endocarditis: Diagnosis by Echocardiography

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SUMMARY The perplexing clinical course of a 23-year-old black male with isolated gonococcal pulmonary valvular endocarditis is presented. M-mode echocardiography provided the first clue to the presence of pulmonary valvular vegetations and the proper diagnosis. Since Neisseria gonorrhea appears to have a particular affinity for the pulmonary valve, the presence of isolated pulmonary valvular endocarditis should raise the strong possibility that Neisseria gonorrhea is the offending organism. This case report of pulmonary valvular vegetations detected by echocardiography strongly emphasizes that all four cardiac valves must be visualized in order to rule out the presence of echocardiographically detectable valvular vegetations.

ECHOCARDIOGRAPHY has been useful in diagnosing valvular endocarditis. Most reports are limited to the echocardiographic findings of the mitral and aortic valves in bacterial endocarditis, although there have been a few descriptions of the echocardiographic manifestations of right-sided bacterial endocarditis. This report demonstrates the role of echocardiography in detecting pulmonary valvular vegetations in a patient with a confusing clinical picture.

Case Report

A 23-year-old black male presented at the outpatient clinic at the University of Nebraska Medical Center on October 19, 1976 with headaches of 1 month's duration. In 1975 he was treated for urethral discharge, presumably gonorrheal, with resolution of symptoms. Evaluation in the outpatient clinic revealed a well-developed, somewhat obese black male who was afebrile. We heard a grade I/VI decrescendo diastolic blowing murmur along the left sternal border and diagnosed mild aortic valve insufficiency.

In November 1976, the patient was re-evaluated because of a persistent headache. He was afebrile and the diastolic murmur was unchanged. The ECG showed a nonspecific intraventricular conduction delay and borderline voltage criteria for left ventricular hypertrophy. A chest x-ray demonstrated mild cardiomegaly without specific chamber enlargement. An echocardiogram was normal. Multiple blood cultures were negative and the VDRL was nonreactive. The erythrocyte sedimentation rate was 40 mm/hr, and a routine urinalysis was within normal limits.

In December 1976, the patient was admitted to the University of Nebraska Medical Center with a 2-day history of a nonpuritic, maculopapular, petechial rash appearing first on the legs, then on the arms and the remainder of the body. He also suffered migratory arthralgias of the knees, elbows and shoulders. Prehospital medications included acetaminophen and aspirin for headache. The hospital course was marked by daily fevers as high as 39°C. Significant laboratory data included hemoglobin 10.2 g/100 ml and a white blood cell count of 6200 × 10^4 with left shift and normal platelets. An ASO Titer was 166 Todd units; RA factor, antinuclear antibody, Coomb's test, throat and multiple blood cultures were all negative. Routine urinalysis was within normal limits, and a skin biopsy showed perivascular polymorphonuclear infiltrates with negative immunofluorescent staining. We tentatively diagnosed Henoch-Schoenlein purpura and discharged him on propoxyphene and aspirin, with some symptomatic improvement.

After discharge the patient had daily fevers as high as 38.4°C or more and was readmitted in January 1977 for evaluation. The physical examination findings were unchanged from the previous admission except for the disappearance of the previously noted rash. A chest x-ray was unchanged from previous films. M-mode echocardiography suggested the presence of a pulmonary valve vegetation. The blood urea nitrogen (BUN) was 44 mg/100 ml, serum creatinine 2.2 g/100 ml, creatinine clearance 59 ml/min, serum albumin 3.1 mg/100 ml and the sedimentation rate 144 mm/hr. Urinalysis revealed 20-30 red and white blood cells per high-power field. His 24-hour urine-protein excretion was 7.4 g, mostly albumin. Anal, urethral and throat cultures were negative for gonococcus.

Renal biopsy provided evidence of an immune complex glomerulonephritis. Hypercellular glomeruli with occasional crescents were present. Electron
microscopy showed small, dense deposits under the mesangial basement membrane with slight mesangial proliferation. Immunofluorescent staining was negative for IgC, but positive for IgA, IgM and β1-C globulin. During hospitalization, microscopic hematuria persisted and the 24-hour urine protein excretion was 18.5 g. He was treated with prednisone 80 mg/day and discharged free of rash and fever.

He did well for 6 weeks but was readmitted because of recurrent fever, chills, dizziness and muscle cramps. A physical examination revealed a febrile black male in no distress without a rash and with the previously described diastolic murmur along the left sternal border. The hemoglobin was 6.0 g/100 ml and the white blood cell count 19,500 × 10³ with a left shift and 23,000 platelets. BUN was 68 mg/100 ml, 24-hour urine-protein excretion was 11.6 g. The serum complement was slightly decreased and repeat blood cultures (total of 12) were negative. A bone marrow biopsy showed increased iron stores and granulocytic hyperplasia. Bone marrow cultures were negative for bacteria and fungi. Gallium, liver and spleen scans were normal. Chest x-ray showed mild cardiomegaly. The hospital course was marked by daily fevers as high as 42°C. The patient was transfused with packed cells and started on cyclophosphamide 200 mg/day by mouth. Proteinuria decreased to 4 g/24 hours and serum platelets increased. Discharge medications included cyclophosphamide 200 mg and prednisone 40 mg/day.

The patient was quickly readmitted because of fever as high as 42°C and with a hemoglobin of 6.8 g/100 ml. The diastolic murmur had increased to grade III/VI intensity, with inspiratory increase in intensity and a right ventricular lift. Repeat M-mode echocardiography was diagnostic for pulmonary valve vegetations (fig. 1). An excess amount of echo-dense structures was present on the pulmonary valve in both systole and diastole, and the pulmonary valve had a coarse systolic flutter. The aortic, mitral and tricuspid valves were negative for vegetations by echocardiography (fig. 2).

At cardiac catheterization the pulmonary artery pressure was elevated to 43/10 mm Hg, with a mean pressure of 16 mm Hg. There was no end-diastolic gradient across the pulmonary valve. Left ventricular end-diastolic pressure was normal at 9 mm Hg. Left ventricular angiography revealed a normally contracting left ventricle of normal size with no mitral insufficiency. An aortic root injection demonstrated good filling of the aortic root with no aortic insufficiency. A pulmonic arteriogram was done in the left lateral position and demonstrated 4+ pulmonic insufficiency. Two long, thick pedunculated lesions

**Figure 1.** M-mode echocardiogram of pulmonic valve. Vegetations are most easily seen during diastole (arrows).
were seen to be moving freely and appeared to be attached to the pulmonary valve (fig. 3). In systole these lesions extended into the pulmonary artery, which was somewhat enlarged. The right ventricle was well visualized because of the severe pulmonic insufficiency and appeared to be enlarged. No tricuspid regurgitation was noted. Six blood cultures taken from the main pulmonary were negative.

On February 28, 1977 the patient was brought to the operating room and his chest was opened by median sternotomy. The right ventricle and right atrium were markedly distended and very active. The patient was placed on total cardiopulmonary bypass and the pulmonary artery was opened just superior to the pulmonary valve. It was immediately evident that the valve was destroyed. Reddish, friable material was clinging to the remnants of the pulmonary valve. In order to excise the valve completely, a right ventriculotomy was made and connected with the pulmonary arteriotomy site. The valve was excised completely, the base of the valve was curetted with a small bone curette, and cultures were sent to the laboratory. The junction of the right ventricle and the pulmonary artery was oversewn with two bolstered sutures. A right atriotomy was performed to visualize the tricuspid valve, but no abnormalities of this structure were noted. All cultures taken of the blood and vegetations grew Neisseria gonorrhea.

After surgery the patient was started on cephalosporin 4 g four times per day. Three days later, when the results of the operative cultures were known, the antibiotic was changed to penicillin, 24 million units/day for 28 days by continuous infusion. The patient's renal function improved — BUN dropped from a high of 108 mg/100 ml to 16 mg/100 ml. The discharge chest x-ray showed a decrease in cardiac size.

At follow-up 6 months later, the patient noticed some musculoskeletal pain related to his sternal incision, but had no symptoms of left- or right-heart failure and remained free of fever. We heard a grade II/VI pulmonic insufficiency murmur at the upper left sternal border accompanied by a grade III/VI systolic ejection murmur. His sedimentation rate remained normal at 6 mm/hr.

Discussion

Several aspects of this case deserve mention: 1) isolated pulmonary valve endocarditis and vegetations; 2) Neisseria gonorrhea as the etiologic agent; 3) persistently negative blood cultures; 4) absence of pulmonary emboli; and 5) the clinical confusion over a definitive diagnosis until echocardiography provided the clue.

Right-sided endocarditis — particularly pulmonary

FIGURE 2. M-mode echocardiogram of normal aortic (AO), mitral (MV), and tricuspid (TV) valves. LA = left atrium.
valvular endocarditis — is a distinctly uncommon entity.\(^8\) Hamburger et al.\(^9\) cultured all four cardiac valves in dogs given injections of staphylococci with and without associated aortic insufficiency. In only two cases did pulmonary valves culture a significant number of organisms, and in no case did pathology indicate findings suggestive of early endocarditis. The incidence of positive cultures was much higher in the tricuspid area than in the pulmonic area. In several large reviews of infective endocarditis in narcotics addicts, where the etiology was felt to be similar to that in Hamburger’s experiment in dogs, heavy tricuspid valvular involvement was noted.\(^7\)–\(^10\) In only two of the 127 cumulative cases was pulmonic valvular involvement described. In each instance it was associated with involvement of another valve or valves.

In the pediatric age group, where pulmonary valvular abnormalities are more common, Johnson et al.\(^11\) reviewed their 40-year experience with bacterial endocarditis. They reported 149 cases retrospectively, most of whom were under 25 years of age, and found only two cases of pulmonary valvular involvement, both associated with severe valvular obstruction. Only one of these two patients had an isolated pulmonary valvular infection.

Right-sided endocarditis is felt to be less common than left-sided endocarditis for three reasons: 1) the lower right-sided intracardiac pressure,\(^12\) 2) a lower incidence of right-heart valve abnormalities, and 3) reduced blood oxygen content. Right-heart infections are usually from more virulent organisms such as staphylococcus, streptococcus, pneumococcus, and gonococcus.\(^13\)–\(^15\)

In the classic paper by Thayer\(^16\) on gonococcal endocarditis, a high frequency of right-heart involvement was noted. In fact, the pulmonary valve was second only to the aortic valve in the frequency of involvement. Thayer pointed out that the gonococcus had a predilection for infectivity in normal valves when compared with other virulent organisms, and felt that “involvement of the pulmonary orifice has been rather common.” In his series, the pulmonary valve was involved in 25% of the cases of gonococcal endocarditis; and in all 80 cases which had been reported in the literature, pulmonary valvular involvement was 12.5%. Clearly, this differs from recent data on the incidence of pulmonary valvular involvement. The diagnosis of valvular involvement was made in each case by autopsy examination.

Stone\(^17\) reviewed 112 cases of gonococcal endocarditis and found pulmonary valvular involvement in 18 cases (16%). Clearly, gonococcal involvement of the pulmonary valve was common in the pre-antibiotic era. With the advent of penicillin, the incidence appears to have dropped precipitously, and gonococcal endocarditis is now rare. The implication is that the advent of penicillin has changed the presentation of endocarditis and dramatically reduced gonococcal involvement of the pulmonary valve.

All the earlier data obtained on gonococcal en-

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**FIGURE 3.** A pulmonary arteriogram demonstrating pulmonic regurgitation and two large pulmonary valve vegetations (arrows).
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docarditis were based on autopsy material. Now, because many patients do not reach a postmortem examination, the clinical assessment of pulmonary valvular involvement with endocarditis can be exceedingly difficult. This is exemplified by our case presentation.

In spite of a high index of suspicion for bacterial endocarditis, blood cultures were consistently negative. Twenty-six blood cultures were taken throughout his multiple hospitalizations, all of which failed to grow the offending organism. Even with immunosuppression induced by cyclophosphamide and prednisone, cultures were negative. Six blood cultures taken from the pulmonary artery distal to the lesion at cardiac catheterization were negative. Since the patient gave a history of gonococcal urethritis, all cultures were specifically evaluated for the presence of gonococcus. In fact, multiple cultures of the urethra, rectal and oropharyngeal areas failed to reveal the presence of the organism. It was not until the vegetations obtained at surgery were cultured that gonococcus was proved to be the offending agent. It is possible that the so-called bacterial-free state of chronic endocarditis was present later on in the patient’s illness, but many of the cultures were taken early in the course.18

Although frequently present in right-heart endocarditis, pulmonary emboli were absent in this patient. At no time did evidence of pulmonary complications appear in the patient’s clinical course. In fact, a ventilation-perfusion scan failed to reveal any significant defects. Baker,19 in his clinical study of bacterial infection confined to the right heart, reviewed 15 cases of right-sided endocarditis chiefly involving the pulmonary valve. Pulmonary embolic phenomena were noted in 82%, and systemic embolization occurred in 19%. The unexpectedly high incidence of systemic embolization was felt to be due to the high incidence of combined right- and left-heart valvular involvement. Only 4% of the patients in their series had vegetations confined to the right heart.

Since the original description by Dillon20 of the echocardiographic findings in bacterial endocarditis, many articles concerning the diagnostic usefulness of echocardiography in bacterial endocarditis have been published. Most reports concern the findings of infective lesions on the aortic and mitral valves, and relatively little is found on the tricuspid valve.21-26

Only one case of pulmonary valvular involvement has been detected by echocardiography. Kramer et al.4 described a young heroin addict with Pseudomonas septicemia, complicated by pulmonary valve vegetations and destruction, as well as recurrent pulmonary emboli. The vegetations were confirmed at autopsy.

Although the pulmonary valve echo in our patient was consistent with the presence of vegetations, we could not exclude a diagnosis of pulmonary valve tumor, especially in view of the negative blood cultures.27

In summary, pulmonary valvular endocarditis is distinctly uncommon and difficult to diagnosis. Echocardiography can be helpful in demonstrating the presence of pulmonary valvular vegetations. Isolated pulmonary valvular endocarditis should suggest the possibility that Neisseria gonorrhoea is the offending organism. With the recent echocardiographic demonstration of isolated pulmonary valvular endocarditis, it seems reasonable that all four cardiac valves must be visualized to rule out the presence of valvular vegetations.

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The Conduction System in Hypoplasia of the Aortic Tract Complex

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SUMMARY This is a study of the course of the conduction system in two cases of hypoplasia of the aortic tract complex, one with mitral stenosis and the other with mitral atresia. In both there was a posterior atrioventricular (AV) node which formed the AV bundle. In case 1 the bundle was short and bifurcated early. The branching bundle gave off a large left bundle branch (LBB), many Mahaim fibers to the septum and a small right bundle branch (RBB). In case 2 the AV node was divided into two parts which formed two posterior bundles which joined together to form a short branching bundle. Instead of a LBB there were profuse Mahaim fibers passing from the branching bundle to the depths of the ventricular septum. The RBB was larger. The abnormalities seen in the conduction system particularly in the LBB are discussed from the embryologic standpoint.

THE CONDUCTION SYSTEM in hypoplasia of the aortic tract complex\(^1\)\(^,\)\(^2\) has, to our knowledge, not been described. We studied the conduction system in this complex to ascertain what happens to the left bundle branch when the left ventricle is small, thick-walled and accompanied by fibroelastosis, as in this complex with aortic atresia and mitral stenosis, or markedly atrophic, as in this complex with aortic and mitral atresia.

Case 1

This was a 4-day-old infant. We took no ECG, but there was no clinical evidence of atrioventricular block. The anatomic diagnosis was hypoplasia of the aortic tract complex with aortic atresia and mitral stenosis (fig. 1) with hypertrophy and enlargement of the right atrium and ventricle, a small left atrium with thick wall, a small left ventricle, thick wall and fibroelastosis, an atrial septal defect, fossa ovalis type, and a widely patent ductus arteriosus.

Histologic Examination

Conduction System

Methods. The sinoatrial (SA) node and its approaches were serially sectioned as previously described,\(^3\) and all sections were retained. The posterior and anterior walls of the atria were then removed and the remainder of the heart was serially sectioned as previously described,\(^4\) and all sections were retained. Alternate sections were stained with hematoxylin and eosin and Weigert-van Gieson stains. In this manner 5343 sections were examined. The findings were compared with those found in normal newborn hearts.\(^5\)\(^,\)\(^6\)

Findings

SA node, and its approaches, atrial preferential pathways and approaches to the atrioventricular (AV) node. These were in normal position but markedly infiltrated with lymphoid cells.

AV node. This structure was in normal position.
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