Case Reports

Double Valve Replacement for Lupus Valvulitis

Report of a Case and Review of the Literature

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Hemodynamically significant lupus valvulitis, requiring valve replacement, is rare: 21 cases have been reported so far in the literature, and only 2 of these have involved double valve replacement. We describe an additional case of double valve replacement in a patient with systemic lupus erythematosus. The histopathologic and clinical features of this case suggest that valvular involvement resulted from both acute and chronic disease processes. Medical success in the treatment of systemic lupus erythematosus, especially that achieved through prolonged or high-dose steroid therapy, may cause chronic valvular disease to become a more common surgical problem. A review of the literature supports this contention. ***(Texas Heart Institute Journal 1990;17:56-60)***

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Between 30% and 50% of patients with systemic lupus erythematosus (SLE) are found to have heart valve involvement at autopsy.[1] As many as 40% of these cases feature multiple-valve involvement.[1] Hemodynamically significant lupus valvulitis (necessitating valve replacement) is rare, however.

We recently replaced the mitral and aortic valves in a 34-year-old man with lupus valvulitis. Simultaneous double valve replacement in such a patient has been reported only once before in the English literature.[2] (We are also aware of a French case.[3]) This article describes the clinical features of our case and presents a review of the pertinent literature.

Case Report

In May of 1979, after a 9-month period of polyarthralgia, pleurisy, pericarditis, and malar rash, the patient, a 27-year-old enlisted man, received a diagnosis of SLE. Serum antinuclear antibody screening was positive for SLE, with a titer of 1:120, and the patient had a grade 1/6 systolic murmur over the 6th left intercostal space. Clinical features of this murmur suggested mitral regurgitation. When the murmur increased 4 months later, echocardiography showed mild mitral regurgitation, and the patient was started on a regimen of steroids, which subsequently enabled fair control of the immediate SLE symptoms, including symptoms of cardiac involvement.

Over the next 7 years, however, the patient developed retinal changes and progressive proteinuria, and his joint involvement worsened. In November of 1986, a new heart murmur denoting aortic insufficiency was detected. This was believed to be hemodynamically insignificant, yet the patient began to experience increasing exercise intolerance, especially during physical training. This limitation progressed until he was unable to walk more than a block without shortness of breath.

In March of 1987, three months before the present admission, progressive proteinuria necessitated a percutaneous renal biopsy. The biopsy specimen showed focal segmental glomerulosclerosis with epithelial crescent formation and rare glomeruli. These findings were thought to be consistent with diffuse proliferative lupus nephritis, so the steroid dosage was increased. Auscultation revealed a grade 3/6 systolic murmur that radiated to the axilla, a grade 2/6 aortic outflow murmur, and a grade 3/6 diastolic murmur at the left sternal border. Echocardiography disclosed left ventricular hypertrophy, decreased contractility, moderate mitral regurgitation, and moderate-to-severe aortic regurgitation. Cardiac catheterization indicated 4+ mitral regurgitation, 3+ aortic insufficiency, left ventricular hypertrophy, left atrial enlargement, and nonocclusive coronary disease. The pulmonary

artery pressures were elevated, and there was no gradient across the aortic valve. A brief attempt at nonoperative management was unsuccessful.

Present Admission

In June of 1987, the patient, by then 34 years old, was admitted to Letterman Army Medical Center for the treatment of lupus valvulitis. He underwent aortic and mitral valve replacement, the aortic valve being replaced with a #23 Carpentier-Edwards bioprosthesis and the mitral valve with a #29 Carpentier-Edwards mitral bioprosthesis. At operation, diffuse pericarditis and pleuritis were noted. The aortic and mitral valves were sclerotic, and the leaflets were rolled and thickened, particularly on the mitral valve. The aortic valve had verrucous lesions and areas of inflammation. Microscopic examination revealed fibrinoid necrosis with chronic inflammatory changes of the aortic valve (Fig. 1) and fibrosis with focal calcification, particularly on the ventricular aspect of the mitral valve (Fig. 2). These changes were believed to be consistent with lupus valvulitis, in both its acute and chronic forms.

Two months postoperatively, a regurgitant aortic murmur was detected. Further evaluation revealed a perivalvular leak around the aortic prosthesis, which valve we then replaced with another #23 Carpentier-Edwards aortic bioprosthesis. The patient's recovery was uneventful, and during the 2½ years since reoperation, he has remained well. His SLE has been stabilized with continued steroid therapy, and his exercise tolerance has improved to the extent that he can go backpacking and hunting without significant limitation.

Literature Review

A review of the literature revealed that, to date, 22 patients have undergone valve replacement for lupus valvulitis (Table I).[2-20] Although only 3 of these patients (including ours) have undergone simultaneous replacement of multiple valves, 5 patients (23%) showed evidence of multiple-valve involvement. All but 6 of the 22 patients were women. In general, patients were young (average age, 36.2 years), had long-standing SLE (average duration of symptoms, 7.0 years), and were receiving postoperative steroids (in 19 cases). They were likely to have mitral valve involvement (12 cases), lupus nephritis (9 cases), or pleurisy and pericarditis (19 cases).

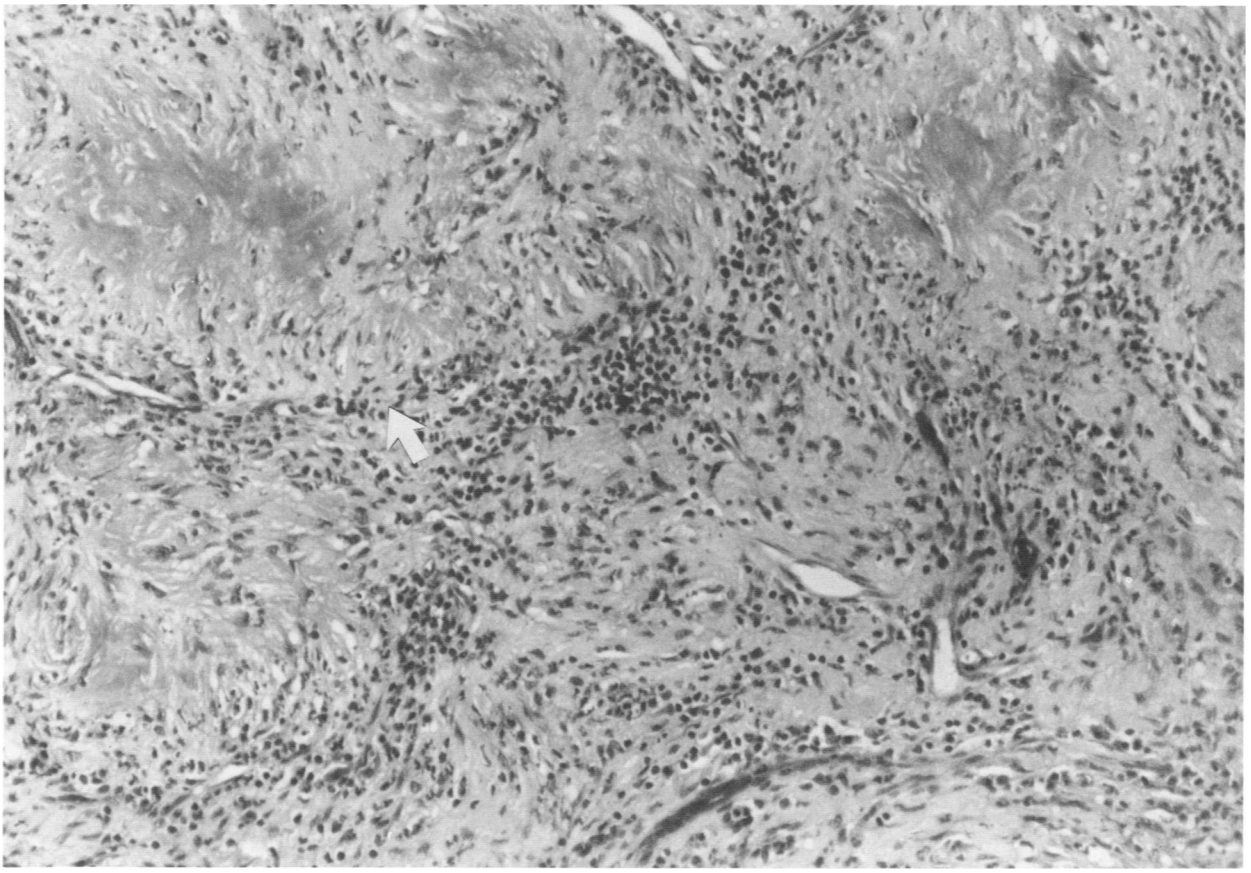

Fig. 1 *Microscopic view of acute aortic valvular changes. The pseudopallisading of lymphohistiocytic cells and the fibrinoid necrosis (arrow) suggest an acute valvular lesion. (orig. x100)*

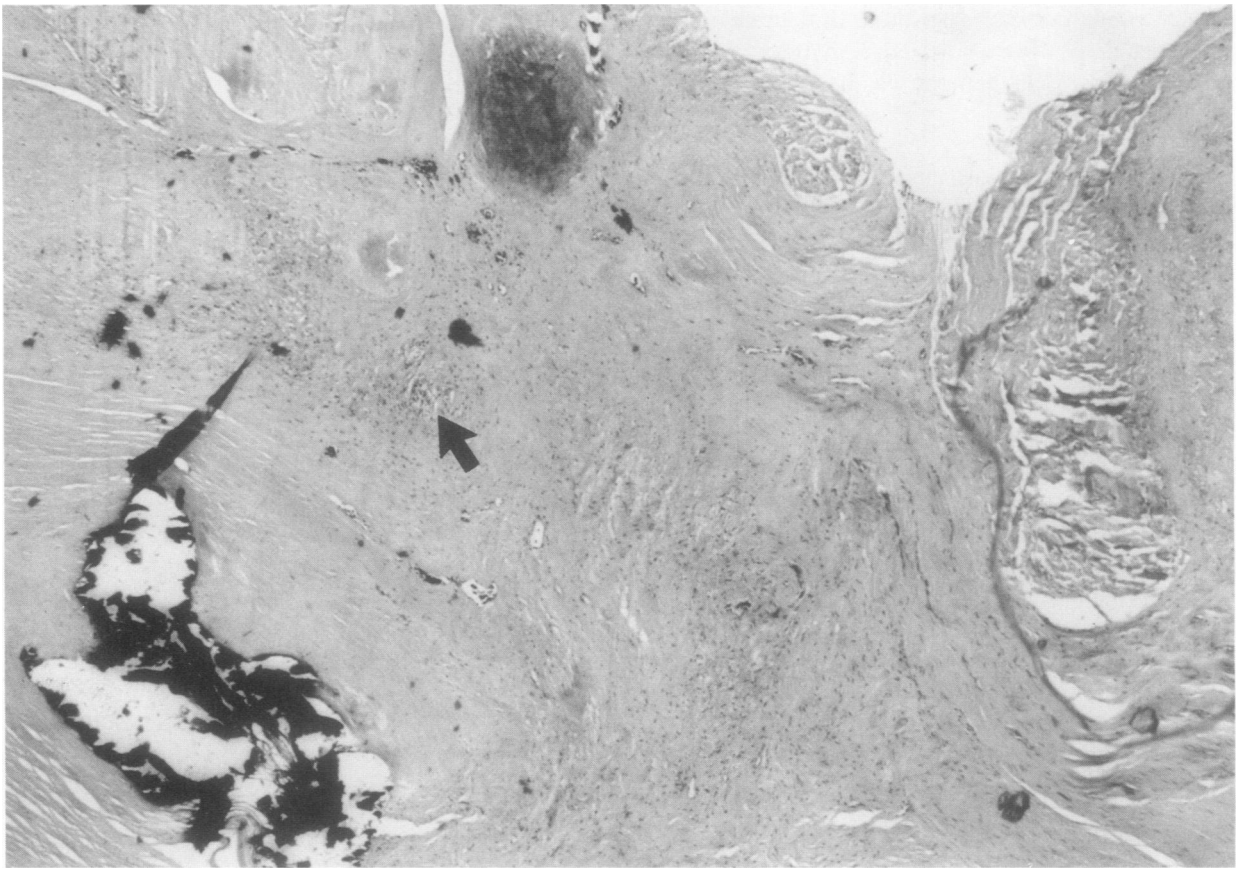

Fig. 2 *Microscopic view of the ventricular aspect of the mitral valve. The focal calcification and fibrosis with neovascularization (arrow) suggest chronic degenerative changes. (orig. x50)*

In the early years of the quarter-century under review, there were 2 operative deaths. Valve-related morbidity was common, occurring in 7 of the 22 patients. Two patients had perivalvular leaks requiring reoperation, 2 had central-nervous-system events suggesting postoperative embolism, 1 had thrombosis of a valve prosthesis, and 1 had early bioprosthetic valve degeneration.

Several authors[6,10,17] commented on the chronic fibrocalcific degeneration evident on histologic examination of the excised native valves. Only 9 of the 22 cases revealed acute changes consistent with Libman-Sacks valvulitis, such as the classic verrucous lesions associated with that disease process.[21] In 2 patients, bacterial endocarditis was superimposed on valves affected by lupus valvulitis.

Discussion

Lupus valvulitis necessitating replacement of multiple cardiac valves is extremely rare, having been reported in only 3 cases, including the present one.[2,3] Our case is unique in its presentation of histologic evidence that the hemodynamic compromise necessitating simultaneous double valve replacement was induced by a combination of chronic and acute valvular changes.

Choosing a valvular prosthesis for use in SLE patients is difficult. Clinical factors affecting this decision include the patient's age and the severity of other organ-system disease (particularly renal) associated with SLE. In the case presented here, for example, we chose bioprosthetic valves due to the risk of hemorrhagic complications associated with mechanical prostheses when Coumadin (sodium warfarin) is used—as would be the case if our patient's lupus nephritis were to cause sufficient renal damage to necessitate hemodialysis. Although one might argue that bioprosthetic valve degeneration would accelerate in a patient with renal failure—as a consequence of abnormal metabolism of calcium and phosphate—the risk of early bioprosthetic valve calcification drops in patients who are over 30 years of age. Our decision to use a bioprosthesis, grounded in these clinical considerations, was a difficult one to make; but the choice was reinforced by our knowledge of the patient's strong desire to maintain an active lifestyle that would include fishing, hunting, and camping.

TABLE I. Patients Requiring Valve Replacement for Lupus Valvulitis

Author	Year of Surgery	Age/Sex	Duration of SLE (yrs) /Steroids Used	Valve /Type	Complication
Bernhard[4]	1966	30/F	4/yes	Aortic/Gott	Operative death
Shulman[5]	1966	42/F	9/yes	Aortic/Starr	None
Oh[6]	1972	38/F	11/yes	Aortic/Starr	None
Yates[7]	1972	29/F	7/yes	Mitral/Starr	Operative death
Murray[8]	1973	43/F	2/yes	Mitral/Beall	None
Myerowitz[9]	1973	19/F	3/yes	Mitral/Hancock	Postop renal failure
Bulkley[10]	1974	18/F	4/yes	Mitral/(not stated)	(Not stated)
Dajee[11]	1975	47/M	1/no	Aortic/Björk	Late CNS event (embolus?)
Dajee[11]	1975	45/F	12/yes	Aortic/Hancock	Early valve degeneration
Mossard[3]	1976	59/M	10/yes	Aortic/Björk	None
				Mitral/Starr	None
Pritzker[12]	1976	36/F	10/yes	Aortic/Björk	Perivalvular leak
Vaughton[13]	1976	36/F	1/yes	Mitral/Starr	Valve thrombosis
Rawsthorne[14]	1978	51/M	21/yes	Aortic/Hancock	Congestive heart failure 9 mo postop requiring mitral valve replacement
Thandroyen[15]	1978	41/M	0/no	Aortic/Carpentier	None
Kinney[16]	1980	27/F	0/no	Mitral/Hancock	None
Laufer[17]	1980	9/F	3/yes	Tricuspid/Björk	None
Brennan[18]	1981	19/F	2/yes	Mitral/Björk	Late stroke (embolus?)
Dajee[11]	1981	34/F	20/yes	Mitral/Hancock	None
Rozman[19]	1981	47/M	6/yes	Mitral/Starr	Atrial dysrhythmias
Tornos[20]	1983	41/F	14/yes	Aortic/(not stated)	None
Moynihan[2]	1987	52/F	5/yes	Aortic/Ionescu	Thrombocytopenia
				Mitral/Carpentier	None
Ferraris	1987	34/M	9/yes	Aortic/Carpentier	Perivalvular leak
				Mitral/Carpentier	

CNS = central nervous system; SLE = systemic lupus erythematosus

In patients with hemodynamically significant lupus valvulitis, the reported intraoperative mortality and perioperative/postoperative morbidity rates have been relatively high. We postulate that several complicating factors are responsible. In addition to the complications posed by steroid therapy, which must be continued even though it slows or alters the healing process and can cause a postoperative hypercoagulable state (both in the immediate and long terms), the following SLE-associated diseases are often present with lupus valvulitis, complicating both the operative procedure and the recovery period: 1) lupus pericarditis, which can cause intraoperative technical difficulties; 2) SLE-associated coronary artery disease and hypertension, which can contribute to perioperative morbidity; and 3) SLE-associated renal disease, which can complicate perioperative fluid management and cause long-term morbidity. These and other factors appear to increase operative morbidity in these patients.

Examination of the clinical and pathologic features of our case, and of those reviewed in the literature, suggests that valvular involvement in SLE results both from active disease and from healed fibrotic lesions. In our case, pathologic examination showed acute inflammatory changes of the aortic valve (Fig. 1), as well as chronic fibrotic changes of the mitral valve (Fig. 2). It is reasonable to postulate that steroid therapy causes healing of some of the valvular lesions, with resultant fibrosis, and that recurrent disease can later produce a more acute, inflammatory lesion.

It was happenstance that, in our case, both types of lesions were evident on histologic examination of the excised valvular tissue. We recommend that clinicians confronted with SLE consider the postulate that, in this disease, all valvular lesions are not

necessarily the result of an acute flare-up but may be a chronic condition due to scarring. Reasonable extensions of this postulate are unverified assumptions that 1) some of the hemodynamic changes in lupus valvulitis are caused by chronic degenerative lesions rather than by acute inflammatory disease, and 2) medical success in prolonging patients' lives through long-term or high-dose steroid treatment will render more common cases of multiple-valve involvement that necessitate surgical intervention. As yet, however, there is no good evidence to indicate that prolonged steroid treatment of SLE symptoms is causing multiple-valve involvement to become more common.

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