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A case of Sjögren's syndrome leading to mitral and aortic valve replacement

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A 62-year-old woman presented with dyspnea on exertion, and an echocardiogram demonstrated an ejection fraction of 55% to 60%, severe mitral regurgitation, and moderate aortic regurgitation. The patient had rheumatic disease as a child, and her valve failure was presumed to be due to rheumatic disease sequelae. She previously had an axillary lymph node biopsy for adenopathy at an outside hospital and was thought to have Castleman's disease. At the University of Iowa Heart and Vascular Center, she underwent aortic and mitral valve replacement through a full sternotomy and bicaval cannulation. The aortic valve was replaced with a 21-mm Magna ThermaFix Edwards bovine pericardial valve (Edward Lifesciences, Irvine, Calif). The mitral valve was replaced with a 29-mm St Jude Epic porcine valve (St Jude Medical Inc, St Paul, Minn). Intraoperatively, the pericardium was found to be inflamed and adherent to the aorta and right ventricle. The native aortic valve was trileaflet, and both valves were rubbery in consistency with significant nodules present. The leaflets were friable. No calcification was found on the mitral annulus. The mitral valve was not repairable.

The gross pathology showed tan white nodules, 0.3 cm in the greatest dimensions, with spiculated appendages on the surface of the valves. The microscopic pathology demonstrated fibroelastic tissue with focal palisading and noncaseating granulomatous inflammation with central necrosis (Figure 1). Postoperative recovery was uneventful. The patient underwent multiple serologic tests that suggested a diagnosis of Sjögren's syndrome (Table 1), which was

confirmed with a positive Schirmer's test and a labial salivary gland biopsy.

DISCUSSION

Sjögren's syndrome was first identified in 1933 by Dr Henrik Sjögren and is a chronic autoimmune disorder with lymphocytic infiltration of the exocrine glands. Sjögren's syndrome manifests with *sicca* symptoms, including xerophthalmia and xerostomia. This disease primarily affects women, and most cases are diagnosed during the middle-age years. Sjögren's syndrome occurring without any other manifestation of other autoimmune diseases has been labeled as primary Sjögren's syndrome. Secondary Sjögren's

FIGURE 1. Mitral valve leaflet pathology demonstrating palisading and granulomatous inflammation, 200 $\times$.

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TABLE 1. Serologic testing results

Test	Result	Normal range
C-reactive protein	4.6 (H)	0–20 mm/h
Erythrocyte sedimentation rate	55 (H)	<0.5 mg/dL
Rheumatoid factor	38 (H)	<14 IU/mL
SSA antibody	127 (H)	<19 units
SSB antibody	182 (H)	<19 units
Anti-nuclear antibody titer	>1:2560 (H)	<1:40
Double-stranded DNA antibody	423 (H)	0–200 IU/mL
Anti-RNP antibody	59 (H)	0–19 units
IgG, serum	2337 (H)	694–1618 mg/dL
ANCA screen	Negative	
Antistreptolysin O	<55	0–330 IU/mL
Anti-cyclic citrullinated peptide	4	0–19 units
C3 complement	151	90–180 mg/dL
C4 complement	18	16–47 mg/dL
Anti-Smith antibody	5	0–19 units
IgM, serum	89	60–263 mg/dL
IgA, serum	335	68–378 mg/dL
Angiotensin-1-converting enzyme	16	8–52 U/L

RNP, Ribonucleoprotein; SSA, Sjögren's syndrome A; SSB, Sjögren's syndrome B; ANCA, anti-neutrophil cytoplasmic antibody; Ig, immunoglobulin. High (H) is denoted in the result column, as is the normal reference range at the University of Iowa Heart and Vascular Center.

syndrome occurs when associated with another autoimmune connective tissue disease, such as rheumatoid arthritis, systemic lupus erythematosus, or scleroderma. Primary and secondary Sjögren's syndromes occur with approximately equal frequency.¹ In addition to the salivary and lacrimal glands, Sjögren's syndrome often affects other organ systems, including the gastrointestinal tract, skin, lungs, vasculature, kidneys, bladder, and vagina.²

Mitral valve replacement for Sjögren's syndrome has not been previously reported. Previous studies have reported echographic findings of valvular regurgitation and pericardial effusion in patients with primary Sjögren's syndrome.³ There has been 1 previous case report of a patient with Sjögren's syndrome associated with aortic valve regurgitation who underwent aortic valve replacement for infective endocarditis.⁴ This is the first report of double-valve replacement in connection with Sjögren's syndrome. A previous report documented a patient with similar characteristics to our patient, including age, gender, medical history, and clinical symptoms on presentation. The patient was found to have mitral and aortic valve stenosis. Further workup showed

the patient to be rheumatoid factor positive, antinuclear antibody positive, and Sjögren syndrome A positive. The patient underwent a Schirmer's test and a salivary gland biopsy to confirm a diagnosis of Sjögren's syndrome. The patient refused surgical treatment and was treated with prednisolone therapy. Her symptoms reportedly improved, although the valve lesions remained.⁵ To our knowledge, there has been no previous documentation of cardiac valvular failure requiring double-valve replacement in connection with primary Sjögren's syndrome. In addition, this is the initial report of insufficiency in Sjögren's syndrome being treated with valve replacement.

CONCLUSIONS

Pathology indicated a nonrheumatic autoimmune connective tissue disease for which the differential diagnosis includes Sjögren's syndrome, systemic lupus erythematosus, mixed connective tissue disorder, and undifferentiated connective tissue disorder. This type of pathology is seen in connection with rheumatoid arthritis, which is a form of a connective tissue disorder, but the patient does not have any clinical symptoms of rheumatoid arthritis. Laboratory evidence supports a diagnosis of a connective tissue disorder. The patient was found to have high antinuclear antibody, Sjögren syndrome A and B, and anti-ribonucleoprotein titers, suggesting a diagnosis of Sjögren's syndrome or systemic lupus erythematosus. Sequelae of childhood rheumatic fevers cause valvular dysfunction, but the typical calcifications, valve thickening and fusion of the commissures typically seen in conjunction with rheumatic fever, were not observed. Sarcoidosis is a possible diagnosis based on bihilar lymphadenopathy and the presence of noncaseating granulomas on the patient's native valves, although serologic tests did not support this diagnosis. Infectious causes are other possible causes of valvular dysfunction, although no infectious organisms were identified.

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