Multiple Cardiovascular Involvement in a Case of Relapsing Polychondritis

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Abstract
Relapsing polychondritis is a multisystem disease characterized by recurrent inflammation of the cartilaginous tissue. Cardiovascular manifestations of relapsing polychondritis are rare but are the second most common cause of death in these patients. We report a case of relapsing polychondritis who underwent aortic valve replacement uneventfully but presented six months later with myocardial infarction due to bilateral coronary ostial stenosis.

INTRODUCTION
Relapsing polychondritis is an uncommon, multisystem, presumably autoimmune disease. It is characterized by recurrent inflammation of the cartilaginous tissues in various parts of the body, commonly involving the cartilage of the ears, nose, joints, ribs and throat. Not commonly, it affects the cartilage of eye, heart and blood vessels. Cardiovascular complications are rare but have been associated with adverse prognosis. Complications include aortitis, vasculitis of large and medium sized arteries with aneurysm formation, valvulitis, pericarditis and conduction disturbances. We report an interesting case of relapsing polychondritis with multiple cardiovascular involvement and a rarely reported ostial stenosis of both the coronary arteries.

CASE REPORT
A twenty-six year old lady, a known case of relapsing polychondritis, presented to us in August 2004 with history of breathlessness, lethargy, decreased appetite and weight loss.

In August 2002 she developed pain and swelling over the nose. She consulted an ENT surgeon and underwent surgery for drop out of mid nasal septum in August 2002. She had body ache and polyarthritis, underwent an extensive work-up for connective tissue disorder, rheumatoid arthritis and vasculitis including, ANA, rheumatoid factor, ANCA by the rheumatologist and was diagnosed to have relapsing polychondritis based on the clinical picture. She was started on Tab. Prednisolone 40 mg once daily and Tab. Dapsone 50 mg once daily. She stopped the medications on her own after one year.

In August 2004, she presented to us with NYHA Class III breathlessness. She had clinical evidence of severe aortic regurgitation - peripheral signs of AR, hyperdynamic apex and early diastolic murmur. Electrocardiogram showed evidence of left ventricular hypertrophy with volume overload pattern. Chest radiograph showed cardiomegaly (CT ratio 65%) and a dilated ascending aorta. Transthoracic echocardiography confirmed the presence of severe aortic regurgitation (Grade III) and the other valves were normal. Aortic annulus measured 24mm, aortic root measured 36mm and sinotubular junction measured 32 mm in size. Left ventricle was dilated and ejection fraction was 61%. Subsequently she underwent a routine aortic valve replacement with 25mm Omnicarbon valve in August 2004. The intraoperative findings noted were thick walled and inflammed ascending aorta, dilated aortic annulus and small coronary ostia. Intraoperatively the aortic root was dilated. Histopathologic examination (Figs. 1, 2), showed a myxomatous aortic valve. The specimen from the aortic valve showed fibrous tissue with a mixed inflammatory exudate of neutrophils, eosinophils, lymphocytes and macrophages with areas of necrosis. The aortic wall was not biopsied. Pericardium showed features of chronic pericarditis. The patient was not restarted on steroids because at that time the ESR which was 75mm / 120mm preoperatively, had decreased to 30mm / 70mm in the postoperative period. Secondly, steroids would delay wound healing. Good healing is required in operation of aortic valve surgery because the high systemic pressure can cause dehiscence leading to
She was on regular follow up with us postoperatively with no cardiac symptoms. However, she developed painful red nodules in both ears which was treated as auricular chondritis 2 months post surgery and was restarted on Tab. Prednisolone. In January 2005, she presented to us with acute onset chest pain. ECG revealed non ST elevation anterior wall myocardial infarction. Echocardiography showed mild left ventricular dysfunction with regional wall motion abnormality in the mid and distal anterior segments and normally functioning aortic prosthesis with valve gradients, maximum 10 mmHg and mean 6 mmHg. Coronary angiogram revealed ostial left main disease (Fig. 3) with ostial RCA disease (Fig. 4). Emergency CABG was planned but she had sudden hemodynamic collapse and, sustained a fatal cardiac arrest shortly thereafter.

**DISCUSSION**

Relapsing polychondritis is a rare multi-system disorder with no specific diagnostic test and numerous possible symptoms. Hence McAdam’s criteria is useful to diagnose this condition.

- Recurrent chondritis of both auricles
- Chondritis of nasal cartilages
- Chondritis of the respiratory tract involving laryngeal or tracheal cartilages
- Non-erosive inflammatory polyarthritis
- Inflammation of ocular structures
- Cochlear or vestibular damage

The diagnosis of relapsing polychondritis is made if,

- Atleast 3 of McAdam’s criteria are present. No histological confirmation required.

- One or more of McAdam’s criteria are present. Histologic confirmation is required.
- Chondritis in 2 or more separate anatomic locations with response to steroids and/or dapsone.

No controlled trials of therapy for RP have been published. The goal of treatment is to abate current symptoms and to preserve the integrity of cartilaginous structures.

The mainstay of treatment is systemic corticosteroids. Prednisone (20-60 mg/d) is administered in the acute phase and is tapered to 5-25 mg/day for maintenance. Severe flares may require 80-100 mg/day. Most patients require a low daily dose of prednisone for maintenance; however, rare patients can be treated successfully by intermittent administration of high doses during flares of the condition. McAdam et al found that continuous prednisone decreased the severity, frequency, and
duration of relapses. Other medications reported to control symptoms and, perhaps, progression of the disease include dapsone (25-200 mg/d), azathioprine, methotrexate (7.5-22.5 mg/wk), cyclophosphamide, and cyclosporin A. Methotrexate has been administered beginning at 7.5 mg/week, increasing up to 22.5 mg/week in conjunction with steroid administration and has been found to decrease patients’ corticosteroid requirements significantly while controlling symptoms. Oral administration of nonsteroidal anti-inflammatory drugs (NSAIDs) has not been effective.6

Cardiovascular manifestations have been described in 11% to 56% of patients. They are the second most common cause of death and include aortitis, vasculitis of large and medium-sized arteries, atrioventricular conduction disturbances, aortic regurgitation, and pericarditis.3 Serious cardiovascular complications appear in about 25% of patients with relapsing polychondritis, the most frequent being aortic or mitral regurgitation (11.1%) and aortic aneurysms (6.1%). Aortic regurgitation is the most common cardiac manifestation of relapsing polychondritis and occurs in approximately 10% of patients.4

We report this case, first to highlight the cardiovascular complications that can occur with this relapsing polychondritis and to address a few management issues. The cardiovascular complications in this patient were aortic regurgitation, bilateral coronary artery ostial involvement and pericarditis.5 Coronary angiography as a routine presurgical evaluation is done in all patients aged more than 40 years, but in patients with relapsing polychondritis it is advisable to do coronary angiography prior to surgery regardless of the age at presentation. During surgery it is be advisable to avoid ostial cardioplegia and in case of doubt regarding the coronary ostial involvement it would be prudent to do a Bentall’s procedure (aortic root replacement with valve replacement).

Cardiac surgery with cardiopulmonary bypass is associated with whole body inflammation and can result in reactivation of relapsing polychondritis. Should this mandate use of steroids prior to surgery followed by continuation of steroids in the post operative period? In case of coronary ostial involvement, whether to do coronary artery transfer using button technique and risk inflammation stenosis in the transferred button or give individual grafts to coronary arteries is open to debate.

REFERENCES