# Paravalvular Insufficiency In Multisystemic Hypersensitivity Vasculitis

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**Citation**

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**Abstract**

We present a case of a 52 year-old woman suffering from Hypersensitivity vasculitis with multisystemic involvement of skin, blood, lungs, and kidneys. The patient developed acutely paravalvular insufficiency of her prosthetic mitral valve. This is the first case, to our knowledge, who presented with paravalvular insufficiency of an artificial valve as a consequence of hypersensitivity vasculitis.

## INTRODUCTION

Hypersensitivity Vasculitides (HV) are a heterogeneous group of necrotizing vasculitides of the small vessels. The disease affects equally men and women. Usually, it appears between 16 and 65 years of age [1,2]. Causative factors and/or coexisting diseases, associated with HV, are autoimmune and collagen diseases, bacterial and viral infections, AIDS, malignancy, hematological diseases, and drugs [3,4]. Prognosis depends on the severity of the coexisting disease and renal involvement. Most patients survive for years. The manifestations of the disease are:

Mitral valve replacement is associated with a paravalvular leak in up to 12.5% of patients and depends on the type of artificial valve that is used, the age and the center where the surgery was performed. The mean time of occurrence of this complication is 119 days (1 day to 23 years). Aggressive surgical correction of the problem is the treatment of choice [7].

We report a patient suffering from hypersensitivity vasculitis with multisystemic involvement and acute onset of paravalvular insufficiency in a previously well functioning artificial mitral valve, two months after the diagnosis of the HV. This is the first report, to our knowledge, in which hypersensitivity vasculitis is implicated as cause of paravalvular insufficiency in a mechanical valve.

## CASE REPORT

A 52 year-old woman was admitted in our department because of fever up to 38°C, fatigue, anemia, leukocytosis and purpuric lesions on the lower extremities. Past history included, left hemiparesis, total thyreoidectomy due to malignant fibrous histiocytoma, and total hysterectomy due to Krukenberg tumor. She had surgery for mitral valve replacement twice, four years ago. Chronic atrial fibrillation was present. Laboratory findings were also characteristic. Positive Coombs haemolytic anemia, reticulocytosis, neutrophilic leucocytosis, erythrocyte sedimentation rate at 112 mm. Renal insufficiency with Clearance 54,3ml/min, microscopic hematouria and trace of albumin in the urine sediment. Rheumatoid factor, antinuclear, anti-double-strain, anti-neutrophilic (C and P), antibodies and antibodies for hepatitis and HIV were all negative. Complement factors C3, C4 were within normal limits. Bone marrow aspiration was compatible with hemolytic anemia. Bone marrow biopsy showed reactive marrow as after toxic effect. Skin biopsy revealed leykocytoclastic vasculitis with diffuse polymorphonuclear infiltration. High resolution computed tomography (CT) of the lungs revealed bilateral coalescent bronchopulmonary opacities, giving a ground glass appearance and bilateral pleural effusions. Bronchoscopy and ultrasound examination of the abdomen were negative.

Cardiac echocardiogram revealed no signs of endocarditis and a normal functioning valve. Therapy with prednisolone 60mg/24h and cyclophosphamide 125mg/24h was instituted. Fifteen days later she showed remarkable symptomatic improvement and her renal function returned to normal (clearance: 92,3ml/min), Coombs test became negative, E.S.R fell to 51mm and lung CT showed remarkable resolution of the parenchymal opacities and pleural

effusions. The patient was discharged from the hospital in good condition on prednisolone and cyclophosphamide and was followed up in the outpatient department. Two months after discharge, the patient was admitted again to the hospital because of leucopenia, which was attributed to yclophosphamide, and she was given granulocyte-colony stimulating factor, with good response. Heart auscultation, at this time, revealed increased intensity of her mitral valve systolic murmur. A transesophageal heart ultrasound was performed and severe degree of paravalvular insufficiency of the prosthetic mitral valve was diagnosed. She was transferred to another hospital for a heart surgery, where she died.

## DISCUSSION

The American College of Rheumatology (ACR) has established criteria for H.V diagnosis. The presence of three or more criteria is required for the diagnosis and the reported sensitivity and specificity is 71% and 83,9% respectively [9] (Table 1). In our case the diagnosis was based on three out of five criteria, namely, rash, positive skin biopsy and age.

Work up for collagen vascular disease, Granulomatosis Wegener, Behcet syndrome, infection, Hepatitis, HIV and history for medications, was negative. Due to the past history of thyroid and ovarian cancer we investigated her for relapse, with negative results.

In H.V. the skin involvement is common and usually precedes the other clinical manifestations [1]. Leukocytoclastic vasculitis is a term that is used to describe the histopathologic picture of necrotizing inflammation in most of the diseases of this group. In our case there was simultaneous involvement of many systems: with manifestations from blood (Coombs positive hemolytic anemia), lungs (parenchymal involvement, pleural effusions), kidneys (renal insufficiency), skin (leukocytoclastic vasculitis) and heart (paravalvular insufficiency). With the exception of her heart disease all other manifestations had been improved with prednisone and cyclofosfamide.

Cases of valvulopathy due to vasculitis have been reported in Henoch-Schonlein purpura, pruritic vasculitis and necrotizing vasculitis related with C-anca. In all the above cases, the involved valve was the native aortic valve and usually aortitis coexisted. In our case, the paravalvular insufficiency of a prosthetic mitral valve developed acutely two months after the clinical event of the vasculitis while the patient was on the appropriate treatment. This could be due to direct involvement of the myocardium, as it happens in Henoch Schonlein purpura. [9,10]

### Figure 1

Table 1: American College of Rheumatology (1990): Criteria for the diagnosis of hypersensitivity vasculitis.

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| 1. | Age above 16 years at disease onset |
| 2. | History of medication taking at the onset of the disease that may have act as a precipitating factor |
| 3. | Presence of palpable purpura |
| 4. | Presence of maculopapular rash |
| 5. | Biopsy demonstrating granulocytes around an arteriole or venule |

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