

Normocomplementemic Urticarial Vasculitis Occurring at Early Postoperative Period After Mitral Valve Replacement: Case Report

Mitral Kapak Replasmanı Sonrası Erken Postoperatif Dönemde Gelişen Normokomplementemik Ürtikeryal Vaskülit

Mustafa Bahadır İNAN, MD,^a
 Çağdaş BARAN, MD,^a
 Burak AÇIKGÖZ, MD,^a
 Sadık ERYILMAZ, MD,^a
 Zeynep BAŞTÜZEL EYİLETEN, MD,^a
 Aylin HEPER, MD,^b
 Kemalettin UÇANOK, MD,^a
 Ümit ÖZYURDA, MD^a

Departments of ^aCardiovascular Surgery,
^bPathology, Ankara University Faculty of
 Medicine, Ankara

Geliş Tarihi/Received: 17.09.2008
 Kabul Tarihi/Accepted: 23.03.2009

Yazışma Adresi/Correspondence:
 Çağdaş BARAN, MD
 Ankara University Faculty of Medicine,
 Department of Cardiovascular Surgery,
 Heart Center, Ankara,
 TÜRKİYE/TURKEY
 cagdasbaran@gmail.com

ABSTRACT Hypocomplementemic urticarial vasculitis syndrome (HUVS) is a clinically rare disease with unknown etiology and incidence. In this case report, we present a 29-year-old male patient diagnosed with normocomplementemic urticarial vasculitis with mitral valvular involvement.

Key Words: Mitral valve; vasculitis, hypersensitivity

ÖZET Hipokomplementemik ürtikeryal vaskülit sendromu (HUVS) hastalığı, insidansı ve etiolojisi bilinmeyen ve klinik olarak nadir gözlenen bir hastalıktır. Biz, 29 yaşındaki erkek olgumuzda mitral kapak tutulumuyla birlikte olan normokomplementemik ürtikeryal vaskülit hastalığını sunduk.

Anahtar Kelimeler: Mitral kapak; vaskülit, hipersensitivite

Türkiye Klinikleri J Cardiovasc Sci 2009;21(2):282-4

Vasculitis is the group of heterogeneous diseases which progress with tissue ischemia and occlusion in variable ranges, characterized by pathological inflammation of the vessels. In the two thirds of the neutrophilic leukocytoclastic vasculitis, immunoglobulin and complement deposits can be seen at the vessel wall. Urticarial lesions with histological evidence of leukocytoclastic vasculitis are the characteristic lesion of the urticarial vasculitis.¹ Although HUVS is a clinical entity in a group of patients with urticarial vasculitis it's been shown that a clinicopathological correlation between hypocomplementemic and normocomplementemic urticarial vasculitis also exists.^{1,2} This is one of the rare cases reported since 1993 which underwent valvular replacement because of cardiac valvulopathy in association with HUVS.^{3,4}

In this paper, we report a case with HUVS who underwent mitral valve replacement.

CASE REPORT

A thirty-year-old male patient admitted to our clinic with the diagnosis of mitral stenosis. His chief complaint was dyspnea and he was NYHA Class 3 before the operation. He had a history of maculopapillary rash with arthralgia a year ago, but recovered in 5 days without any medication.

The echocardiography showed mitral stenosis with a valve area of 1.4 cm² in planimetric measurements, and 1.35 cm² in Doppler measurement. The echocardiography score was 9-11 with a pulmonary artery pressure (PAB) of 40 mmHg. Informed consent was obtained from the patient.

After preoperative preparation, he underwent mitral valve replacement with a size 27 Sorin (Sorin Biomedica Cardio, Italy) mechanical prosthesis. The posterior leaflet was preserved in the operation. The patient was carried to the ward in the 2nd postoperative day without any problem. He had intravenous heparine and per oral warfarin. Heparine was discontinued when INR levels reached 2,5.

In the second postoperative day, he had fever with severe pain localized to the distal upper extremities and joints with maculopapillary lesions in legs bilaterally (Figure 1). Prophylactic antibiotic therapy was suggested by the department of infectious diseases because of the suspicion of bacterial endocarditis. However, blood cultures were negative and the prosthesis was functioning normally without any vegetation in transeusophageal echocardiography (TEE).

Although early bacterial endocarditis is the main suspected diagnosis with inadequate clinical findings, the patient was also consulted to the immunology and dermatology departments.

The immunologic laboratory findings were within normal ranges. Following normal repetitive TEE results and normal complement levels [C3 1.450 g/L (0.9-2), C4 0.355 g/L (0.1-0.4) and C1q 0.14 g/L (0.1-0.16)] skin biopsy was performed. And he was diagnosed as leukocytoclastic vasculitis (Figure 2). There were no physical signs of arthritis and no deformities were observed in the joints. Also the ocular examination was also normal.



FIGURE 1: Bilateral maculopapillary lesions in lower extremities.

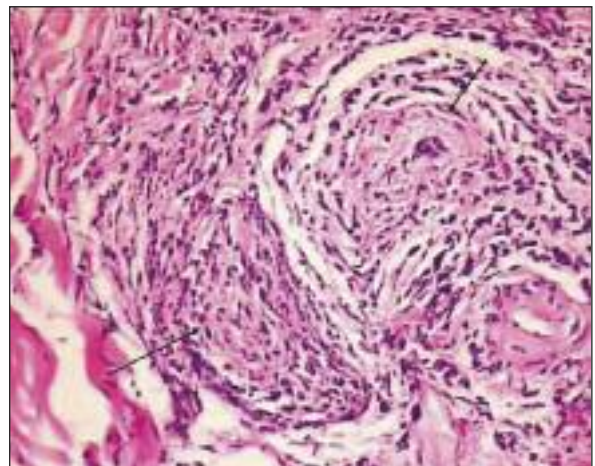


FIGURE 2: Haematoxylin and Eosin stain (x40). The obliterated vessels (arrows) which were infiltrated by leukocytes and associated with slight fibrinoid necrosis and nuclear fragments.

The symptoms immediately recovered after administration of antiinflammatory drugs and low-dose systemic corticosteroid treatment and the patient was discharged with similar medication. At the end of the first week after the diagnosis of vasculitis all the symptoms were revealed and the patient was totally free of the maculopapillary lesions and discharge with anticoagulant, anti-inflammatory medication and low-dose systemic corticosteroids.

DISCUSSION

HUVS is an uncommon disorder that may be seen in routine clinical practice, and characterized by

persistent urticarial lesions with histological evidence of leukocytoclastic vasculitis. It is an uncommon vasculitic disorder of unknown etiology, and its cause, incidence, and prevalence are unknown.⁵

Patients with HUVS should be evaluated for the presence and/or development of valvular heart disease.² However cardiac involvement can be seen in Behcet's disease, which is another type of vasculitis.⁶ Urticarial vasculitis is a disease which can also interfere with endocarditis in the early postoperative period. Valvular diseases can be as a consequence of vasculitis and the clinicopathological examinations must be done if suspected.

Although valvular involvement can be seen with urticarial vasculitis there are only five cases of HUVS and valvular disease in the literature.²

In our case, the patient was preoperatively diagnosed as rheumatic valve disease and had valve

replacement. Although his urticarial vasculitis symptoms (macula-papillary lesions, urticaria, etc.) occurred after the 2nd postoperative day he was asymptomatic before the operation. This was the main reason why we did not perform valvular biopsy. And another important questions is the correlation with the vasculitis and the valvular disease we believe that our patient had normocomplementic urticarial vasculitis with mitral valve involvement and the vasculitis is aggravated with cardiopulmonary bypass, which explains the onset of the vasculitis following the cardiac operation.

As a result we believe that physicians must keep in mind that valvular biopsy must be performed in case of presence of preoperative vasculitic symptoms (mild or severe) additional to valvular symptoms.

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