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Septic and aseptic valvular involvement in hypocomplementemic urticarial vasculitis syndrome

Scheggi V.; Olivotto I.; Alterini B.

Careggi University Hospital, Department of Cardiology, Florence, Italy

Background: Hypocomplementemic Urticarial Vasculitis Syndrome (HUVS) is a very rare disease involving small vessels and characterized by chronic urticarial vasculitis, arthralgia, arthritis, and activation of the classical complement pathway. To our knowledge, just other seven cases of HUVS with cardiac valvular involvement have been reported in the literature. Unlike previous cases, ours shows a broader antibody positivity, (but no anti-C1q antibodies) with an early and serious cardiovascular involvement.

Case summary: A 32 years old woman was brought to the emergency department because of sudden onset of right hemiplegia and global aphasia due to cerebral haemorrhage.

She suffered from a severe HUVS form with cardiac valvular involvement and she was cronicly treated with immunosuppressant drugs.

During previous years she had already undergone cardiac surgery twice for chronic aseptic inflammatory process involving aortic and mitral valves.

Right hemiplegia and global aphasia were caused by septic embolism to left cerebral artery complicated by cerebral haemorrhage.

The patient underwent neurosurgical intervention followed by antibiotic therapy.

This condition was secondary to *Listeria monocytogenes* aortic infective endocarditis in an immunosuppressed patient.

Transesophageal echocardiogram showed periaortic abscess with internal colliquation.

A further surgical intervention would have been necessary, but operative risk was judged to high and the patient died one year later.

Discussion: The association between HUVS, Jaccoud's arthropathy and cardiac valvular disease is rarely described in the literature. The presence of valvular involvement is a negative prognostic factor.

Unlike most cases, it is interesting to note that our patient was negative for anti-C1q antibodies and exhibited an extremely severe cardiac involvement.

The case was complicated by *L. monocytogenes* endocarditis, that is also a rare condition associated with a high mortality rate.

Abstract P1457 Figure. Abscess

