CORRESPONDENCE

Hypocomplementemic Urticarial Vasculitis Syndrome: An Interdisciplinary Challenge

by Prof. Dr. med. Wolfgang Grotz, Prof. Dr. med. Hideo A. Baba, Dr. med. Jan U. Becker, Dr. med. Martin W. Baumgärtel in volume 46/2009

Recurrent Episodes of Sudden Acute Sensorineural Hearing Loss

We wish to point out that in otological terms, Cogan's syndrome presents primarily as recurrent, sometimes bilateral, episodes of sudden acute sensorineural hearing loss. These episodes, which may improve initially after intravenous administration of steroids, may result in progressive hearing loss and permanent surdity. In such a scenario, a cochlear implant is the only option to restore a patient's hearing. We also ask that current terminology be used regarding surdity ("loss of hearing" rather than "deafness" [Box 2], Muckle-Wells syndrome).

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Question Mark Over Hypocomplementemia

Unfortunately it remains unclear whether the intention was to describe an independent pathology (hypocomplementemic urticarial vasculitis syndrome) or whether the scenario was one of immunovasculitis (leukocytoclastic vasculitis) with a particularly pronounced reduction of complement factors. It would be interesting to know whether hypocomplementemia exists in such patients before disease onset or for a long period of time after symptoms have disappeared. Further, it is well known that diverse medications are the most common cause of immunovasculitis, which is always accomxpanied by a reduction in complement factors. No mention was made in the case report about drug or topical treatment preceding (or during) the illness.

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In Reply:

We thank Vent and Hüttenbrink for their comments from an otorhinolaryngological perspective. Their letter underlines the interdisciplinary challenge posed by treating patients with this type of systemic vasculitis. The questions asked by Barz provide us with an opportunity to point out again that HUVS is undoubtedly an independent disease entity. The hypocomplementemia is not specific. Leukocytoclastic vasculitis—not identical with immunovasculitis—is always accompanied by complement deficiency and often by hypocomplementemia. The main symptom that is typical for this systemic vasculitis of the small vasculature is chronic urticarial vasculitis characterized by the immunological marker of the C1q antibody and accompanying complement deficiency. According to the classification criteria for HUVS as set out by Schartz (2), Lupus erythematodes and cryoglobulinemia have to be excluded because for both disorders, similar clinical and immunological constellations have been observed.

Whether hypocomplementemia may exist before onset of HUVS is not known, but is not to be expected either, because in patients in remission, the measured complement concentrations are normal (authors' own observations). Of course, a multitude of medications may trigger vasculitis (3). Whether medications can trigger HUVS could not be confirmed on the basis of our case patient. As we mentioned in our case report, therapeutic attempts using topical triamcinolone or oral prednisolone remained unsuccessful, as was stopping the ACE inhibitor that the patient had taken for many years. Only immunosuppressive treatment with mycophenolate led to long term remission.

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Conflict of interest statement
The authors of all letters declare that no conflict of interest exists according to the guidelines of the International Committee of Medical Journal Editors.