

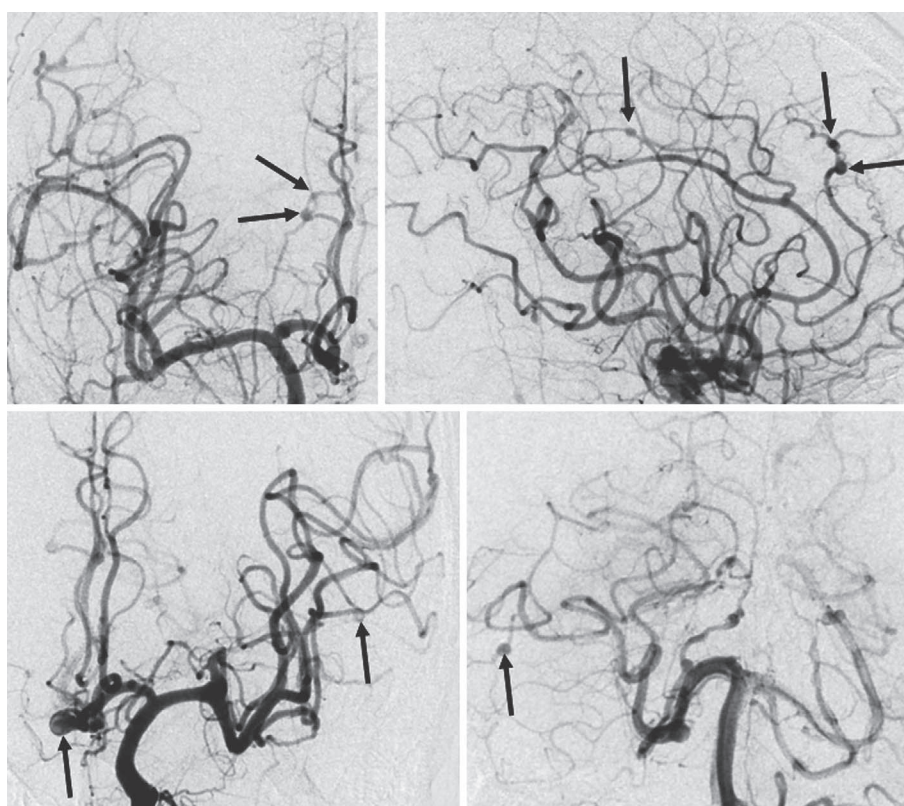
Multiple Intracranial Aneurysms Associated with Hyper-IgE Syndrome

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Key words: hyper-IgE syndrome, hypereosinophilia, intracranial aneurysm, multiple, angiography

(Intern Med 51: 515-516, 2012)

(DOI: 10.2169/internalmedicine.51.6939)



Picture 1.

A 61-year-old man consulted us due to the suspicion of the presence of cerebral aneurysms based on follow-up MRI examinations associated with previous ischemic strokes. He had a past history of hyper-IgE syndrome (HIES) (peak, 92,256 IU/mL), hypereosinophilia [(peak eosinophil count > 3,959/ μ L (normal, <700/ μ L)], and epidermodyplasia verruciformis of 19 years duration. Cerebral angiography showed multiple aneurysms (Picture 1, arrows). One of these aneurysms was then clipped at the left A1-A2 junction. HIES is a rare primary immunodeficiency disease characterized by

recurrent multisystem infections and increased serum levels of IgE (>2,000 IU/mL) (1, 2). Central nervous system abnormalities associated with this disease have been identified as lacunar infarctions, Chiari type 1 malformations, cerebritis, and aneurysms (1, 2). Eosinophilia is a prominent feature of HIES, and eosinophilic infiltrations have been implicated in the development of aneurysms (1). Therefore, the eosinophilia might have contributed to the development of multiple cerebral aneurysms in this case. Patients with HIES should therefore be recommended to undergo regular cere-

bral vascular investigations.

References

The authors state that they have no Conflict of Interest (COI).

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