

- 37 **Skworc KH**, Windl O, Schulz-Schaeffer WJ, et al. Familial Creutzfeldt-Jakob disease with a novel 120-bp insertion in the prion protein gene. *Ann Neurol* 1999;**46**:693-700.
- 38 **Van Gool WA**, Hensels GW, Hoogerwaard EM, et al. Hypokinesia and presenile dementia in a Dutch family with a novel insertion in the prion protein gene. *Brain* 1995;**118**:1565-71.
- 39 **Owen F**, Poulter M, Lofthouse R, et al. Insertion in prion protein gene in familial Creutzfeldt-Jakob disease. *Lancet* 1989;*i*:51-2.
- 40 **Dermaut B**, Cruts M, Backhovens H, et al. Familial Creutzfeldt-Jakob disease in a patient carrying both a presenilin 1 missense substitution and a prion protein gene insertion. *J Neurol* 2000;**247**:364-8.
- 41 **Priola SA**, Chesebro B. Abnormal properties of prion protein with insertional mutations in different cell types. *J Biol Chem* 1998;**273**:11980-5.
- 42 **Xiang F**, Almqvist EW, Huq M, et al. A Huntington disease-like neurodegenerative disorder maps to chromosome 20p. *Am J Hum Genet* 1998;**63**:1431-8.

NEUROLOGICAL PICTURE.....

doi: 10.1136/jnnp.2003.031393

Unusual differential diagnosis of leptomeningeal enhancement: moyamoya disease

We report on a 13 year old boy who presented with a focal left sided motoric seizure and a history of progressive headaches since 6 years of age. On neurological examination, motor function of the limbs, deep tendon reflexes, and sensory examination were normal, but there was slight left sided bradydisdiadochokinesis. Routine blood tests and anti-immune antibody screening as well as cerebrospinal fluid (CSF) analysis were normal.

Magnetic resonance imaging (MRI) showed multiple curvilinear areas of high signal intensity in leptomeninges (fig 1A) and diffuse enhancement along leptomeningeal surfaces (fig 1B). Digital subtraction angiography demonstrated severe stenoses of both distal internal carotid artery (ICA) segments (fig 1C) as well as extensive leptomeningeal collaterals (fig 1D).

Leptomeningeal enhancement on post-contrast T1 weighted MR images has been reported in association with meningitis, meningeal carcinomatosis, and angiitis. Common among these diseases is pathologic inflammation or carcinomatous infiltration of the leptomeninges, with or without involvement of the small leptomeningeal vessels.

In 1995 Ohta *et al*¹ reported diffuse leptomeningeal enhancement on post-contrast T1 weighted MRI in children with moyamoya disease and named these finding the "ivy-sign", because it resembles ivy creeping on stones. The ivy-sign can be found on fluid attenuation inversion recovery (FLAIR) images, as well as on contrast enhanced MRI.^{2,3}

The characteristic enhancement is caused by an engorged pial network via leptomeningeal anastomosis. The source of leptomeningeal high signal intensity on FLAIR images is not clear but it seems largely due to slow flowing vessels of the engorged pial network.² Thus, the mechanism of the leptomeningeal ivy-sign can be explained by an engorged pial network and possibly the congested meninges.

J Gaa, S Weidauer

Institute of Neuroradiology, Klinikum der Johann Wolfgang Goethe-Universität Frankfurt, Germany

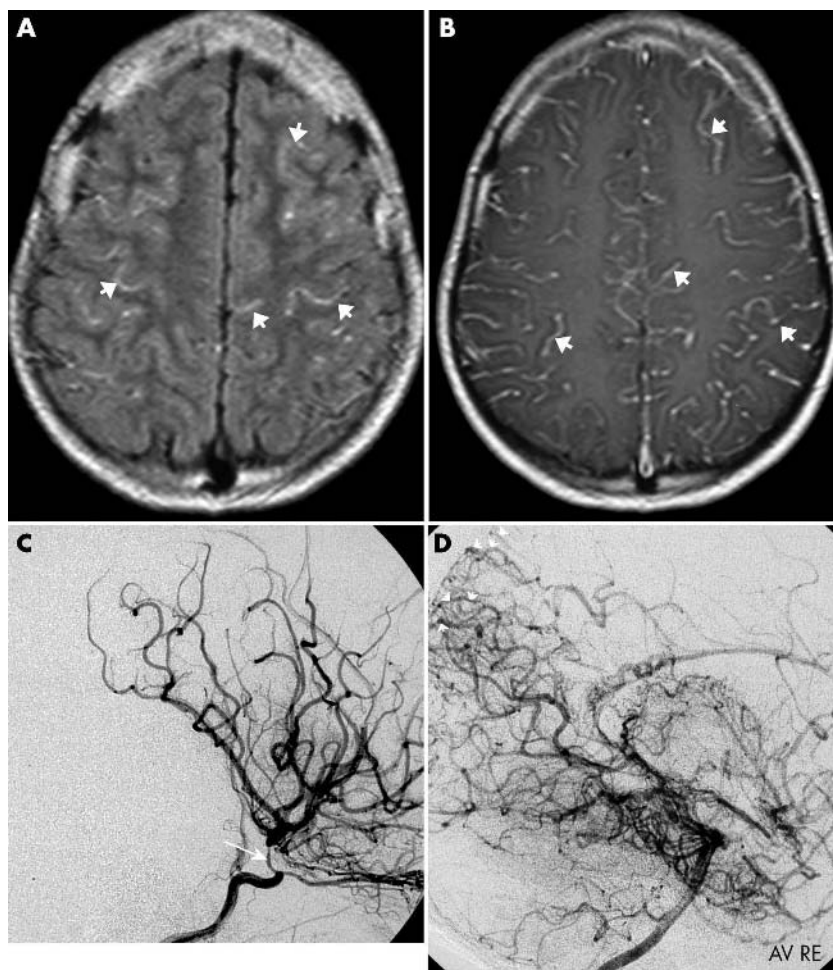


Figure 1 (A) Axial FLAIR MRI reveals multiple areas of high signal intensity (arrows) in leptomeninges. (B) Axial contrast enhanced T1 weighted image shows diffuse enhancement along leptomeningeal surfaces (arrows). (C) Lateral view of right carotid angiogram and (D) right vertebral angiogram demonstrate severe stenoses at the distal ICA (arrow) and wide terminal segments of the posterior and middle cerebral arteries due to extensive leptomeningeal collateral flow (arrowheads).

Correspondence to: Dr J Gaa, Department of Neuroradiology, University of Frankfurt, Schleusenweg 2-16, 60528 Frankfurt, Germany; jochen.gaa@kgu.de

References

- Ohta T, Tanaka H, Kuroiwa T. Diffuse leptomeningeal enhancement, "ivy sign", in magnetic resonance images of moyamoya

disease in childhood: case report. *Neurosurg* 1995;**37**:1009-12.

- Maeda M, Tsuchida C. "Ivy-sign" on fluid-attenuated inversion recovery images in childhood moyamoya disease. *American Journal of Neuroradiology* 1999;**20**:1836-38.
- Yoon HK, Shin HJ, Chang YW. "Ivy sign" in childhood moyamoya disease: depiction on FLAIR and contrast-enhanced T1-weighted MR images. *Radiology* 2002;**223**:384-89.