Summary of the literature concerning intracranial fusiform aneurysms

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Mills' syndrome: ascending (or descending) progressive hemiplegia: a hemiplegic form of primary lateral sclerosis?

We describe two patients with slowly progressive hemiplegia. These two cases bring to mind a rare clinical syndrome described in 1900 by Mills—namely, ascending (or, less often, descending) progressive hemiplegia. Mills'[1] claimed that this disorder was a new form of degenerative disease characterised by progressive deterioration of the corticospinal pyramidal pathways. Despite its age, the concept of Mills' syndrome is still controversial. Indeed, a number of pathological factors can cause such clinical findings, and the cases that remain isolated and can be considered as primary are rare.

Case 1, a 49 year old right handed woman with no previous personal or family medical history, complained in 1975 of motor deficiency on the right side of the body. The initial symptoms were weakness of the right foot and leg, which slowly progressed to the thigh. The patient was admitted to hospital at the age of 55. Physical examination showed right Babinski's and Hoffmann's signs. Tendon reflexes were very pronounced on the right side. The motor deficiency was strictly limited to the right lower limbs. There was no sensory loss. An EEG, CSF examination (cytology, EMG, brain CT, and contrast myelography) were normal. Re-examination in 1987 showed that the disability had increased: there was a pyramidal gait and a distal motor deficit of the right arm associated with a moderate hypertonia. Facial mobility was normal. No sensory deficit was noted. Routine laboratory tests were normal. Serological tests for syphilis were negative. Examination of CSF showed a slight increase in protein content (0-60 g/l) without pleocytosis; immunological tests did not disclose intrathecal synthesis of immunoglobulins or oligoclonal IgG bands. Visual, auditory, and somatosensory evoked potentials were normal. EMG was performed again without results. Routine laboratory tests were normal. Serological tests for syphilis were negative. Examination of CSF showed a slight increase in protein content (0-60 g/l) without pleocytosis; immunological tests did not disclose intrathecal synthesis of immunoglobulins or oligoclonal IgG bands. Visual, auditory, and somatosensory evoked potentials were normal. EMG was performed again without results. Routine laboratory tests were normal.

immune system, detection of inflammation, and serology testing for syphilis were normal. An EEG and visual, auditory, and somatosensory evoked potentials were normal. Brain and spinal cord MRI was normal. Analysis of CSF gave a protein content of 0·1 g/l with no pleocytosis and normal immunological findings. Three EMGs performed at two-year intervals (1985, 1987, and 1989) showed no evidence of denervation. A muscle biopsy performed in 1985 was normal. In June 1993, after 17 years of evolution, the patient’s condition was stationary.

Mills described eight cases of a very slow advancing form of hemiplegia beginning usually in the extremity of a lower limb, then spreading up to the homolateral upper limb. 1, 2 Five more cases were published between 1927 and 1951. 3, 4 In those 13 and in our two cases pyramidal signs were always seen on the side of hemiplegia and often bilaterally (seven of 15 cases). A moderate amyotrophy without fasciculations is common (six of 15). The palsy can involve the face (five of 15). Sensory disturbances are usually absent but in a few cases (three of 15) hypaesthesias have been noted. The manifestations very gradually worsen. Progression is more often ascending (13 of 15) than descending (two of 15). Involvement of the contralateral side of the body has been reported in advanced stages (five of 15). A family history of the syndrome was not noted in any of the cases. The clinical picture presented by our two patients is identical to that described by Mills. 1, 2 After an 18 and 17 year duration of development all manifestations were still unilateral except for increased tendon reflexes in case 2.

Mills’ syndrome is supposedly due to primary degeneration of the corticospinal pyramidal pathways. Pathological examination has been performed in only one case: the results showed non-specific, staged, irregular lesions of the pyramidal tracts predominating on one side at the level of the spinal cord and in the brainstem. The motor cortex was not involved; Betz cells were normal.

The scarcity of case reports and the availability of only one case confirmed by necropsy raise doubt as to the authenticity of this syndrome. Moreover, many reports date back to times when diagnostic methods did not allow reliable differential diagnosis. In this regard it should be said that ascending hemiplegia with Mills’ syndrome-like features could be due to a variety of causes—for example, brain tumours, abnormalities of the cervical spine or cervico-occipital junction, small lacunar infarctions, pontine lesions, multiple sclerosis, or early stage amyotrophic lateral sclerosis. In our two cases the very slow progression of symptoms and the absence of any peripheral motor neuron involvement are inconsistent with amyotrophic lateral sclerosis. Similarly brain MRI, CSF examination, and evoked potentials in both cases ruled out multiple sclerosis. Finally, compression of the cervical spinal cord was not found.

Mills’ syndrome could be considered as a variant of primary lateral sclerosis, which is characterised by a pure upper motor neuron syndrome related to a bilateral degeneration of the corticospinal pyramidal pathways. Several cases of primary lateral sclerosis have been documented after elimination of all other causes of spastic syndromes. 5–10 Pringle et al 10 proposed clinical and laboratory diagnostic criteria for primary lateral sclerosis: all of these are present in our two cases except for negative Lyme disease and HTLV-1 serologies which have not been tested. But Pringle et al 10 specified that these tests have to be done in endemic areas only and the south east of France is not such an area. Thus we propose that Mills’ syndrome could be considered as the hemiplegic form of primary lateral sclerosis. We hope that this report will incite others to publish similar findings. Many more cases documented with current diagnostic technology will allow the confirmation or otherwise of the nosological authenticity of Mills’ syndrome.

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Figure 1 T2 weighted axial MRI at the level of thepons showing an area of increased signal in the ventral aspect of thepons extending across the midline to the left.

Figure 2 T1 weighted midline sagittal MRI showing a low signal area in the ventral pons.
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