"a more diffuse central derangement" in the stiff-man syndrome might reflect the diffuse understanding of the pathophysiology of this and related syndromes.

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Neurological findings in Miller-Fisher syndrome

Sir: I have read with interest the article "Neuropathological findings in Miller Fisher syndrome" by Phillips *et al*.¹ The syndrome of ophthalmoplegia, ataxia and areflexia is attracting new interest, and there seem to be purists who believe that all its features, including the "cerebellar" type ataxia, are due to some form of "unusual" dysfunction of the peripheral nerves.² The classical case, in the presence of the three cardinal features, shows remarkable absence of motor and sensory signs in the limbs.³ Evidence of brain stem involvement in Miller Fisher syndrome is based on pathological reports of abnormalities in the mid-brain and pons in three cases³⁻⁵ and the presence of CT scan evidence of brain stem lesions in several classical cases.^{3, 6, 7}

The relationship of Miller Fisher syndrome to the acute inflammatory polyneuropathy (Guillain-Barré syndrome) is not clear. It seems that Miller Fisher syndrome represents one of the ways in which the central nervous system responds in a "hypersensitive" or "allergic" manner to some external and possibly infective challenge.³ Thus it could represent one end of a spectrum where the pathological process lies entirely in the brainstem, while at the opposite end is the acute inflammatory polyneuropathy where the brunt of the disease falls on the peripheral nervous system. In between there might be cases in which both the central and the peripheral nervous systems are involved. This idea does not conflict

with the fact that cranial nerve dysfunction and even a complete ophthalmoplegia can be seen in the course of acute inflammatory polyneuropathy, where the pathological findings will be limited to the peripheral nervous system.

Phillips *et al*'s patient presented with total ophthalmoplegia, proximal limb weakness, profound sensory disturbances and neurophysiological evidence of involvement of the peripheral nerves. This does not conform to the classical description of Miller Fisher syndrome. Moreover, they were unable to demonstrate abnormalities in the third, fourth and sixth cranial nerves, partly because it was not possible to examine the distal portions of these nerves, but the proximal portions were normal. The crucial issue seems to be that the central nervous system was reported as normal. Thus, the pathological evidence presented only serves to explain the limb signs and stops well short of explaining the ophthalmoplegia.

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Phillips and Stewart reply

The classical description must be that of Fisher. The syndrome he described was one of ophthalmoplegia, ataxia and areflexia.¹ These are presumably Dr Al-Din's

"three cardinal features". All should be present for the diagnosis to be made and all were present in our patient. Two of Fisher's three patients had sensory disturbance and weakness of the limbs was questioned in one. Therefore, we do not think that the presence of these two features weakens our diagnosis in any way. Our patient's illness does conform to Fisher's description.

The article claimed to describe the classical case giving details of 18 patients, half of whom did not show all three of these cardinal signs. (Six with preserved reflexes and three without ophthalmoplegia).² If one changes a diagnostic criterion, then the "classical" case may also change.

Our post-mortem specimens demonstrated ample evidence of extensive demyelination in the peripheral nerves, so Miller Fisher syndrome cannot represent "one end of a spectrum where the pathological process lies entirely within the brain stem".

We agree the histologically normal brain stem and cerebellum were crucial findings. Whatever caused the ophthalmoplegia, it was not brain stem encephalitis and a peripheral lesion seems more likely than a central one.

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