We are grateful to Professor ES Watkins for allowing us to report this case.

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Matters arising

Exteroceptive reflexes abnormalities in stiff-man syndrome

Sir: We read with interest the electrophysiological studies of a case of stiff-man syndrome by Meink et al. The authors analysed the properties of exteroceptive reflexes circuitry by means of stimulation of a limb mixed-nerve trunk. They showed that exteroceptive reflexes are pathologically enhanced in stiff-man syndrome, in so far as they lack habituation, show lowered threshold and simultaneous co-contraction of antagonistic muscles. In a previously reported case of stiff-man syndrome we observed comparable abnormalities of exteroceptive reflexes. By means of stimulation of both mixed and purely sensory nerve trunks we demonstrated that exteroceptive reflexes lacked habituation phenomena and that they induced co-contraction of antagonistic muscles. Furthermore, exteroceptive reflexes were elicited only by their specific stimuli and only at the specific site of stimulation, without any enlargement of their receptive field. In contrast, we observed that monosynaptic reflex activity behaved substantially normally. Thus Meink et al.'s data confirm our previous findings in showing that isolated abnormally enhanced exteroceptive reflexes activity underlines the major electrophysiological abnormalities in stiff-man syndrome. As regards the involvement of the central nervous system in stiff-man syndrome, we recall that the prevalence of generalised epilepsy in this syndrome is about 10%, that is, much higher than among the general population. Furthermore abnormal muscular activity in stiff-man syndrome completely disappears during sleep, a behaviour typical of centrally originating motor disorders. These findings lend indirect support to the hypothesis that the loss of inhibitory control of exteroceptive activities reflects a more diffuse central derangement. The absence of any pathological correlates in the few cases necropsied, further shows that stiff-man syndrome is a functional rather than a structural disturbance.

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References


Meink replies:

The fact that exteroceptive stimuli induce irradiating spasms in the stiff-man syndrome has been described earlier than 1950, suggested by Martinelli and Montagna (for example "C"). The contribution of Martinelli et al. to this particular problem consists of three sentences, namely the statement that "polysynaptic flexion and exten- sion reflexes . . . were normal" whereas "non-nociceptive reflexes . . . behaved abnormally, lacking habituation phenomena and exhibiting co-contraction ... (p. 459), and the suggestion of "abnormal functions, even if not specific . . ., of the polysynaptic system" (p. 461).

Besides having difficulties in understanding the exact meaning of their somewhat sibylline statement, I think that we did not confirm Martinelli et al.'s (or others) previous results, but substantiated their observations by means of a systematic analysis.

In my opinion, the author's calculation of prevalence rates for generalised epilepsy in the stiff-man syndrome is seriously affected by at least two factors: firstly, treatment of the stiff-man syndrome is usually performed with longterm administration of benzodiazepines at high dosages resulting in a high risk for epileptic seizures during withdrawal. Secondly, the term "stiff-man syndrome" most probably comprises a wide variety of disorders which have in common muscular stiffness and spasms, but are poorly understood with regard to their individual aetiology and pathogenesis. The author's hypothesis of