Letters

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Paroxysmal presentation of motor neuron disease

Sir: A 40 year old lady complained of brief attacks of a tight feeling in her throat, slur-

ring of speech and difficulty in swallowing. No precipitating factors were known. Several such episodes were seen during a twenty minute consultation, each lasting for 15 to 20 seconds. In an attack she became severely dysarthric, indeed almost unintelligible. Her 12 month old child had recently been causing her considerable alarm by having breath holding attacks. There were no abnormalities on either general medical or neurological examination.

A definite diagnosis was not made and it was considered that the attacks might be "functional" in nature. On review 2 months later she had developed wasting of the small muscles of the hands, slight wasting of the tongue and had definite, mild, permanent dysarthria. Over the next few months she developed bulbar palsy and upper and lower motor neuron lesions in all four limbs. The now obvious diagnosis of motor neuron disease was made and electromyographic studies revealed denervation in all four limbs.

She deteriorated steadily and died within eighteen months at home. Permission for necropsy was not granted.

The author has not seen this presentation of motor neuron disease previously. Awareness of the possibility would have prevented the consideration of functional disorder which held sway for a brief period of two months before the fatal nature of the disorder became apparent. The patient was never aware of the suspicion of a functional diagnosis. Once more the danger of the diagnosis of hysteria is highlighted. Absence of a diagnosis should never be equated with absence of all possible physical disease.

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