optic atrophy and a central scotoma in the right eye. Both received steroid therapy, detailed as ACTH for the female and unspecified for the male. Four months after presentation our patient had a residual arcuate scotoma in his left eye, but no optic atrophy. The two differences between our patient and those previously reported were that our patient had a coxsackie B5 infection as opposed to coxsackie B4 in the French patients; secondly our patient received high dose prednisolone therapy as opposed to ACTH. It may be that the differences in treatment influenced the final visual outcome.

At presentation the visual loss in the left eye was ascribed to optic nerve disease but the electrodiagnostic tests suggest this was more likely to be retinal in origin. The flash ERG showed no definite abnormality in the left eye whereas the PERG was grossly abnormal. It has recently been reported that if the PERG is abnormal in optic nerve disease the abnormality is likely to be confined to the N95 component. The left PERG showed a severe P50 component abnormality in keeping with macular dysfunction, demonstrating the ability of the PERG to determine whether a PVEP delay reflects optic nerve or more distal retinal dysfunction.


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Some historical observations on involuntary movements of the face

Part I

In June 1920, Pierre Marie and Gabrielle Levy described 49 patients with transient or permanent movement disorders related to epidemic encephalitis and possibly influenza seen since 1918. Their paper was called "Le Syndrome Excito-Moteur de L'Encephalite Epidemique". Four patients were described in detail and the movements were categorized as:

1 Choreic, local or generalised, of coarse or small amplitude.
2 Bradykinetic oscillations, coarse, slow, rhythmic movements particularly of the proximal limbs.
3 Myoclonus of either the trunk or the limbs.
4 Parkinsonism, with or without tremor.
5 Isolated tremor, rare and present almost exclusively in the face.
6 Localised facial movements, either tongue-face-masticatory or ocular, sometimes with concomitant facial pain similar to tic doloreux.

Included in the facial movements was trismus as well as difficulty in opening and closing the mouth and involuntary movements of the jaw in several directions. These movements were sometimes accompanied by pain in the face and irritation and grinding of the teeth and a repetitive sucking movement.

There was also muscle spasm around the eyes and mouth and excessive persistent salivation. Speech was sometimes explosive without a real dysarthria and the tongue was often fasciculating.

In the same year Marie and Levy reported three other cases of postencephalitic dystonias. Two of these had involuntary movements shown exclusively as lingual-facial-masticatory disorders. The third had the same movements plus tic doloreux.

A sensation of the throat being obstructed, paroxysmal or constant, was also reported. It was accompanied by disturbances of phonation, shortness of breath or dysphagia, all related to spasm of the muscles of the larynx and pharynx. Abnormal yawning was commonly associated with the development of Parkinsonism and hiccoughs with the myoclonic abnormalities. A sensation of suffocation with an increase in respiratory rate occurred and occasionally a prolonged inspiratory phase as if the patient was sobbing.

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References

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