drome is usually accompanied by long tract signs. Although rare in vascular accidents, damage to the striatum with virtual sparing of other structures has been implicated in the development of hemidystonia and focal dystonia following mild head injuries, and torticollis has been described in association with an arteriovenous malformation close to this region.

Of particular interest in this patient was the focal nature of the dystonia in association with a discrete contralateral cerebral lesion. Denny-Brown, in his discussion of segmental dystonia described various abnormal hand postures which may follow a focal contralateral subthalamic lesion that damages the fields of Forel. In such cases the upper limb is rigid and initially the wrist is flexed and pronated with the hand open, but after some years the fingers may become tightly clenched and the elbow flexed. Whilst our patient exhibited rigidity at the left elbow and wrist, her abnormal posture was restricted to three fingers. In the absence of the CT scan findings, her hand posture might have been regarded as functional.

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References

Association of Klippel-Trenaunay-Weber syndrome with myotonic dystrophy

Sir: Myotonic dystrophy, an inherited disorder, is associated with cataract, frontal baldness, cardiac dysfunction, testicular atrophy, abnormal insulin secretion, Klinefelter syndrome, and Down’s syndrome, in addition to central and peripheral nervous system dysfunction. Cutaneous manifestations, other than balding, are rare. Few cases of pilomatrixoma (a benign calcifying epithelioma) have been reported in myotonic dystrophy. We observed an extensive systematized vascular naevus in a segmental distribution with hypertrophy affecting one upper limb, the characteristic features of Klippel-Trenaunay-Weber syndrome, in a patient with myotonic dystrophy. An exhaustive review of the literature has failed to disclose similar cases.

A 39-year-old male Muslim, an agriculturist, born of a consanguineous union was seen in December 1980, with complaints of weakness of lower limbs of 2 yr duration. He had most of the cardinal signs of myotonic dystrophy such as myotonia, frontal balding, testicular atrophy, posterior capsular cataract, wasting and weakness of temporalis, masseter, sternocleido-mastoid muscles of the forearm, hands and feet (fig). Electromyography showed well sustained spontaneous, high frequency discharges, myopathic pattern and nerve conduction studies showed normal motor and sensory conduction velocity in the upper and lower limbs. A cutaneous patch on the right side of the chest and medial aspect of the arm was noticed since birth. This was dull red in colour in the distribution of thoracic 1 and 2 segments (fig). Global hypertrophy of the right arm including the shoulder was present. No venous varicosities were seen nor was there any evidence of associated disorders like syndactyly, polydactyly, scoliosis, pulmonary hypertension, mental retardation, epilepsy or paraplegia.

Fig (A) The dystonic posture of the left hand is demonstrated. (B) CT scan shows a low attenuation area in the right lentiform nucleus as well as enlargement of the right Sylvian fissure. (Côt SB).
Matters arising

Evoked potential changes in clinically definite multiple sclerosis

Sir: Drs Walsh, Garrick, Cameron and McLeod described changes of evoked potentials in patients with clinically definite multiple sclerosis. They concluded that serial studies can be useful if the clinical diagnosis remains doubtful. Apart from this, we already established role of evoked potentials in the diagnosis of multiple sclerosis, frequent serial recording of evoked potentials might be useful also for quantitating the spontaneous course of the disease and the effect of therapy.

In successive measurements in normal subjects of pattern reversal visual evoked potentials (VEP) and short latency somatosensory evoked potentials (SEP) only small variations of the latencies of the evoked potentials were noted, which seems to allow detection with evoked potentials of changes within the visual and sensory systems of patients. In a serial study in our laboratory of 25 patients with multiple sclerosis it was demonstrated that across an interval of 3½ months the latencies of the VEP and SEP changed substantially in many of the patients. The changes of the latencies of the evoked potentials had a certain relation to clinical parameters of the visual and sensory systems; in some patients, however, the variation of the evoked potentials were unaccompanied by clinical change. This could indicate that evoked potentials might be more sensitive to variation in functions of the central nervous system than clinical examination.

In their investigation across an interval of 2–2½ years, Walsh, Garrick, Cameron and McLeod noted only exceptionally some reduction in latencies of the evoked potentials; the overall deterioration demonstrated in the electro-physiological measurements correlated with an increase in clinical disability of the group of patients over the period of study. In our study, across a much shorter interval, four patients showed a normalisation of an abnormal VEP; the SEP returned to normal in five cases. In other patients also significant decreases of the latencies of the evoked potentials were noted. Perhaps this improvement reflects a restored conduction in demyelinated fibres in the central nervous system, which might be an effect of the apposition of new layers of myelin, diminishing of oedema in plaques or changes in the internodal axonal membrane.

A further progression of the disease over a period of years might be the cause of the disappearance of the improvements which can be demonstrated after a short time interval. Frequent serial recording of evoked potentials, however, seems to be useful as a parameter of (short lasting) changes occurring in the central nervous system and might play some role in the evaluation of therapies in patients with multiple sclerosis.

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References