Fig. Shows naevus on the right side of the chest and hypertrophy of the shoulder and frontal baldness.

Klippel-Trenaunay-Weber syndrome or "hemangiectatic hypertrophy" is one of the phakomatoses. The cutaneous naevus is usually observed at birth with a tendency to grow with age. The naevus can have a variable size ranging from a pin head to an extensive lesion affecting one entire limb or rarely the whole body and is generally unilateral and has a segmental distribution. Hypertrophy may involve the bone, muscle, sub-cutaneous tissue and fat. When the process begins early in life it may result in lengthening of a limb; however, when commencing at a later stage an increase in girth alone may result. Venous varicosities are an important component of this syndrome. When not evident clinically they can be detected by phlebography. Very few neurological manifestations have been reported in association with Klippel-Trenaunay-Weber syndrome. Recently paraplegia and ischaemic infarct of the brain stem have been observed.\(^2\) The present report documents the association with myotonic dystrophy. It is difficult to postulate a common origin for both diseases, but it is an interesting association.

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References


Matters arising

Evoked potential changes in clinically definite multiple sclerosis

Sir: Drs Walsh, Garrick, Cameron and McLeod\(^1\) 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 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 somato-sensory evoked potentials (SEP) only small variations of the latencies of the evoked potentials were noted,\(^2\) 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.\(^4\) 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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