putamen and the anterior limb of the internal capsule. The latter localisation could be responsible for the aphemia. To our knowledge aphemia has not been described previously as a first symptom of multiple sclerosis.

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References

Limb girdle type muscular dystrophy associated with a Wolff-Parkinson-White syndrome

Sir: Cardiac involvement varies greatly with different types of muscular dystrophy. In general the so called dystrophic heart disease is common in Duchenne's muscular dystrophy (50–80%), infrequent in limb girdle type muscular dystrophy and occasional in all other types of muscular dystrophy. The most important clinical features of myocardiial disease in muscular dystrophies are tachycardias, arrhythmias, congestive heart failure and sudden death. Various publications emphasise the even more frequent appearance of an altered ECG. The Wolff-Parkinson-White syndrome has been described in patients with cardiomyopathy and in three cases with Duchenne's muscular dystrophy. To our knowledge the syndrome has not been reported in association with limb girdle type muscular dystrophy.

A 40 year old male patient was admitted because of weakness and atrophy of shoulder and pelvic girdle muscles. He had a history of 12 years with lumbosacral pain and slow deterioration of muscular function which incapacitated the patient in his daily activities. Examination revealed normal vital signs, a weight of 45 kg and a height of 168 cm. Cardiologic examination was normal. The patient had severe asymmetrical proximal muscular atrophy of both upper and lower limbs. Distal, facial and abdominal muscles were not affected, and there were no skeletal deformities. Biceps, triceps and knee reflexes were diminished. The patient had a lordotic gait, difficulty in climbing stairs, combing his hair and Gower's sign was positive. The rest of the neurological examination was normal.

Laboratory data revealed a CK of 500 U (normal up to 170 U) with a muscle fraction of 1.1%, LDH, SGOT and all other standard laboratory tests were normal. Chest and abdominal radiographs were normal. The resting ECG showed a sinus bradycardia of 49 beats per minute and an axis of 20°. The QRS complex measured 0.15 ms, the PR interval 0.1 ms and the PJ interval 0.26 ms. Positive delta waves were seen in leads I, aVL and V4-V6 and a negative delta wave was observed in V1 compatible with a type B Wolff-Parkinson-White syndrome (fig). The ECG also revealed sinus bradycardia which was reversed by exercise and thought to be physiological. An M-mode, two dimensional and dynamic echocardiogram did not reveal abnormalities and all standard measurements were normal. The EMG showed fibrillations of low amplitude and polyphasic motor unitdeltoid, biceps, quadriceps and anterior tibial muscles. Neuroconduction was normal. The muscular biopsy specimen showed muscle fibre necrosis, wide variations in fibre size, increased connective tissue and internal muscle fibre nuclei, being compatible with muscular dystrophy. We have obtained detailed information of 38 family members. One brother of the patient was found to have the same progressive muscular process but without cardiac involvement. He was 9 years older than our patient and his disease had progressed further. A muscle biopsy showed the same myopathic features.

Limb girdle type muscular dystrophy is an autosomal recessive disorder characterised by insidious onset in early adulthood and

Letters

Fig Electrocardiogram demonstrating a prolonged QRS complex shortened QT and PJ times and delta waves.
Letters

progression to incapacitation within 20 years. At least seven different types have been reported in literature, with subclassification of this disorder according to onset, inheritance pattern and major clinical features. Our patient was considered to have the autosomal recessive adult onset form of limb girdle type muscular dystrophy. The diagnosis was based on the clinical presentation, muscle biopsy, EMG, laboratory studies and the pedigree. Reported associated conditions of the Wolff-Parkinson-White syndrome such as AV blocks, bundle branch blocks, Ebstein anomaly or mitral valve prolapse were looked for, but were not found in our patient.

An abnormal ECG is very often the earliest evidence of myocardial involvement in muscular dystrophies and therefore an accurate parameter of dystrophic heart disease. Cardiac lesions in limb girdle type muscular dystrophy are rare. Rhythm and conduction disturbances are the most frequent of them, especially atrial tachycardias, atrial flutter, heart blocks, abnormal QRS configuration and abnormal AV conduction. Seldom found features include persistent atrial standstill, severe cardiomyopathy and hypertrophy. Cardiac involvement in muscular dystrophies is progressive and probably associated with the severity of skeletal muscle disease; it may be accompanied with elevation of some serum enzymes like CK-MB (these changes are absent with pure affection of the conduction system). Mechanisms of the rhythm disturbances are still unknown but various explanations have been proposed. Among them are fibrosis, fatty infiltration and sinal and AV nodal arteriopathies. Early detection of cardiac abnormalities may help to prevent sudden death in limb girdle type muscular dystrophy and should therefore be searched for, followed up and treated. When the Wolff-Parkinson-White syndrome is associated with haemodynamically significant tachyarrhythmias, treatment with a combination of procainamide and verapamil or quinidine and propranolol might be beneficial. If pharmacological treatment fails, pacemaker implantation or surgical ablation of the accessory pathway have been recommended.

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References


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Thymoma without myasthenia gravis. Electrophysiological study after thymectomy

Sir: The onset of myasthenia gravis after total extirpation of a thymoma has been reported in the literature. Symptoms and signs of myasthenia gravis have been observed 2 weeks to 6 years following total removal of thymoma. All the reported patients showed clinical signs and symptoms of fatigability. There appear to be no reports of neuromuscular transmission studies in thymectomized cases that are not followed by clinical myasthenia gravis.

A 46 year old woman with a 7 years history of weight loss of more than 20 kg required admission because of exertional dyspnoea, mild chest discomfort and subjective sensation of tiredness. Radiological examination showed a mass in the anterior mediastinum. Laboratory studies, neurological examination, and clinical test of fatigability were normal. An encapsulated tumour was totally removed. There were no signs of invasion into adjacent structures, or evidence of metastasis. Microscopically, the tumour was a thymoma of the mixed cell type. After thymectomy the patient was asymptomatic, and there was no clinical evidence of myasthenia gravis. Neurological examination was also normal at 3 weeks, 17 months and 2 years after thymectomy.

Repetitive nerve stimulation of the accessory nerve under resting conditions, and repetitive stimulation of the ulnar nerve before and after five minutes of ischaemia on stimulating at 3 Hz did not show significant changes in the amplitude of the compound muscle action potentials. How-

Fig. Increased jitter values in seven out of 12 (Top = Three weeks after thymectomy) and in seven out of 20 end-plates evaluated (Bottom = 17 months after thymectomy).

Jitter values were calculated manually from superimposed recordings and expressed in µs as mean consecutive differences of the interpolar interval. Normal upper limit for EDCC muscle in our laboratory is marked by vertical dotted lines. End-plates without (○) and with (●) impulse blocking.
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