IDIOPATHIC PAROXYSMAL CHOREOATHETOSIS – REPORT OF 2 CASES AND REVIEW OF LITERATURE

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SUMMARY

2 cases of Idiopathic Paroxysmal Choreoathetosis with late onset and non familial were described. Examinations and investigations were normal and both responded to phenobarbitone.

INTRODUCTION

Idiopathic Paroxysmal Choreoathetosis is an uncommon disorder. The symptoms are very distressing, but the condition is very easily treated and therefore it is very important to recognise it.

CASE 1

H.B.O. is a 24-years-old right-handed married Malay clerk. She was first seen in March 1982 complaining of recurrent attacks of spasm involving left side of limbs, body and face since early 1979. The attacks came 4 to 5 times per day and each episode lasted 5 to 10 seconds. Each attack started with current-like sensation in the left foot ascending to the trunk lasting a fraction of a second. This was followed by progressive plantar flexion and inversion of left foot with flexed toes and partial flexion at the knee. In the upper limb there was partial flexion of fingers at carpometacarpal joints with extension at interphangeal joints, adducted thumb and progressive supination of forearm. The elbow was partially flexed and the arm was abducted. The trunk was slightly flexed to the left and there was spasm of facial muscle and orbicularis oculi on the involved side. There was no loss of speech or consciousness, and she was able to

Fig. 1 Posture of upper limb in a mild attack.

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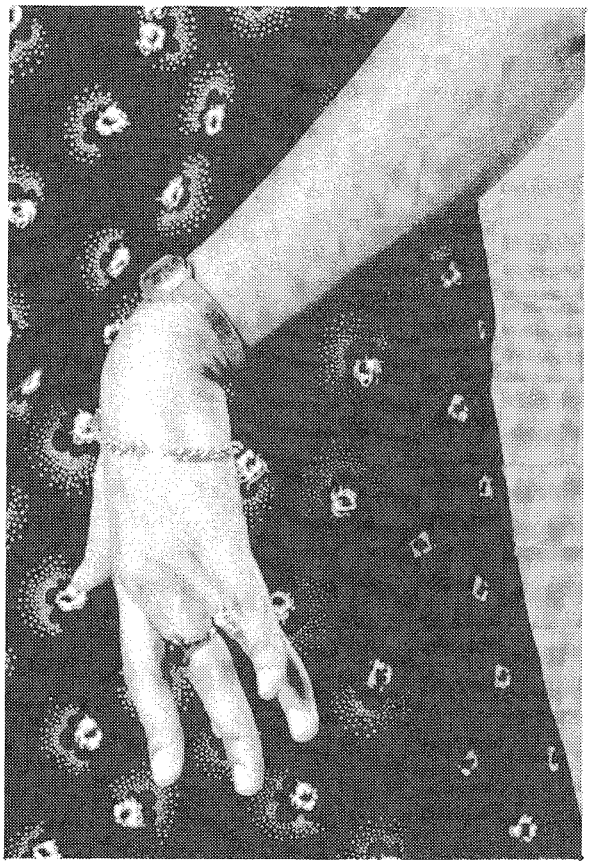

Fig. 2 Close up of hand in an attack.

Fig. 3 Patient straining to abort an attack.

resume her activities immediately after that. The attacks were precipitated by a startle, when called unexpectedly or when trying to stand up from sitting position in certain situations such as when catching a bus.

There were times when she managed to abort attacks by stifling her body and pressing on the left foot.

She was the seventh child in a family of 11 siblings and there was nobody in the family with similar illness or neurological or psychiatric illnessess.

On examination she appeared normal. BP 120/80 mmHg, PR 76/min. No Kayser Fleisher ring noted. Systemic and neurological examination were normal. Blood counts were normal and L.E. cell was negative. Serum caeruloplasmin and 24 hour urine copper were normal. Skull X-Rays, EEG, radioisotope brain scan and CAT scan of brain were normal. She was treated with phenobarbitone 30 mg three times per day with complete control of symptoms.

CASE 2

L.E.L., a 21-year-old right-handed Chinese engineering student was first seen in Neurology Clinic in August 1982. He complained of frequent spasms of one side of body and limbs for the past 8 years. The attacks came 20 to 30 times per day and each episode lasted 4 - 6 seconds. In the first 3 years, only the left side was involved, but subsequently, either side could be involved.

Each attack was preceded by momentary current like sensation in one big toe, followed by progressive inversion of foot, flexion of toes and flexion at the knee. There was associated lateral flexion of trunk to the affected side. The arm was adducted and elbow extended with the forearm in hypersupination moving into progressive pronation with extended fingers, adducted thumb and flexion at the wrist. There was also associated spasm of facial muscles and orbicularis oculi on the involved side. Sometimes he was able to abort an attack by pressing hard on the foot at the onset of symptoms.

The patient had poliomyelitis in early childhood which resulted in mild wasting of right leg and mild equinovarus deformity of right foot. He was the eldest in family of 3 siblings. A younger sister has schizophrenia but there was no similar illness or other neurological disease in the family.

On examination he looked well. BP 130/80 mmHg, PR 80/min. No Kayser Fleischer ring. There was mild wasting of muscles of right leg with partial equinovarus deformity of right foot. Otherwise, the rest of the examinations were normal. Blood counts and ESR were normal. Serum calcium, inorganic phosphate, skull X-Rays, EEG and radioisotope brain scan were all normal. Treatment with phenobarbitone 30 mg three times per day relieved him of his symptoms.

Review of Literature

In 1930, Wilson [1] first described a boy, who suffered from unilateral paroxysmal involuntary movements from the age of 5 years. The attacks were precipitated by a startle or a jar. Neurological examinations were normal and he called the condition as Reflex Epilepsy.

Mount and Reback [2] in 1940 first used the label Familial Paroxysmal Choreoathetosis to describe a condition of paroxysmal choreoathetosis in a young man with onset in infancy. There were two grades of attacks lasting about 2 hours. The attacks were precipitated by alcohol, fatigue and concentration. Physical examination was normal except for the presence of atypical Kayser Fleischer ring. Lishman [3] in 1962 described 7 cases of "seizures induced by movements". The age of onset ranged from 3 to 20 years with negative family history. The choreoathetoid attacks were more brief in duration and were precipitated by movements especially when performed suddenly. Clinical examinations were normal and patients improved with phenytoin, phenobarbitone and primidone either alone or in combination.

The case described by Rosen [4] in 1964 had a secondary cause. The child was delivered by forceps because of transverse arrest and was cyanosed for several hours post delivery. Subsequently he was noted to have right monoparesis and hemiatrophy of skull. In 1966 Stevens [5] described 4 cases of similar illness with age of onset ranging from 1 to 33 years. In all the cases, the attacks could be induced by hyperventilation. Clinical examinations were normal and in one of the patients that died, an autopsy showed slightly asymmetry of substantia migra.

Hereditary non Progressive chorea of early onset as described by Haerev [6] is certainly different from paroxysmal choreoathetosis. The attacks were non writhing and choreoform which occurred throughout the day with little variation but exacerbated by mental efforts and stress.

The cases described by Hudgins and Kendall, [7] Kertesz [8] and Jung [11] were quite similar to Lishman's [3] case in their clinical characteristics and response to anticonvulsants. One of Kertesz cases was autopsied, and the abnormalities noted were diffused vascular congestion and melanin pigments in macrophages in the locus ceruleus which suggested slight loss of neurons in this nucleus.

Richard and Barnetts' [9] cases bore resemblence to the case of Mount and Reback. In 1969 Kato [10] described paroxysmal choreoathetosis in a Japanese patient, with a positive family history. The features were quite similar to the Chinese cases described by Jung and Chen except that in Kato's patient there was also ballismic component.

DISCUSSION

Paroxysmal choreoathetosis is a clinical syndrome with varying degrees of severity. In the majority of patients the inheritance was antosomal dominant but there were also sporadic cases.

The possibility of underlying central nervous system pathology must be kept in mind and reasonable investigation done. [4] In the idiopathic type there are 2 subgroups, the one with onset in infancy, [2,9] and later onset [3,7,8,10] with very important differences.

In the infancy onset type, inheritance was antosomal dominant; the attacks could last few hours; not precipitated by initiation of movements; not associated with sensory aura and anticonvulsants has uncertain results.

In the later onset type, the inheritance could be antosomal dominant or sporadic. The attacks were usually precipitated by initiation of movements, preceded by sensory aura and the choreathetoid movements lasted few seconds to minutes only. Sometimes the patients were able to abort a full blown attack by certain manoeuvres and anticonvulsants has definite value in treatment.

The differences are significant enough such that it is not wise to attach eponynms to the condition. A descriptive label is more appropriate (until more is

known about the illness) that is Idiopathic Paroxysmal Choreoathetosis with onset in infancy or late onset.

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