Matters Arising

Progressive choreo-athetosis related to birth anoxia

Sir: "Static" encephalopathy causing delayed-onset dystonia after perinatal anoxia was reported by Burke *et al*,[1] who also stressed the differential diagnosis with dystonia musculorum deformans.

A patient we followed for over 30 years showed progressive abnormal movements with associated statural and mental retardation after birth anoxia. This girl was the product of an uneventful, term pregnancy. Family history was negative, including the parental consanguinity. Delivery was very complicated and prolonged, the baby being cyanotic and in danger of death (it was hurriedly baptised). Difficulties in sucking were noticed in the following days, but the child thrived thereafter. She started walking when aged 15 months and was considered normal until age 4 years, when slight involuntary jerks began in the right hand. They slowly worsened, forcing the child to use her left hand in writing and in feeding herself. By the age of 12 years, choreic movements were observed, at rest and during movement, in the right hand and also in the right leg, associated with hypotonia. Sydenham's chorea was suspected, but routine investigations were all negative and the patient was diagnosed as having "hemichorea". The abnormal movements slowly worsened, and, by the age of 18 years, they also spread to the lower left limb. Pneumoencephalography and right carotid angiography performed at the time were negative.

Finally, the choreic movements spread to involve the left arm and, when aged 27 years, also the neck. Neurological examination at 31 years of age showed left-sided central facial palsy, a trembling voice, irregular "tremor" distally in all limbs, worse on the right, present at rest and worsened by posture and movements, associated with slight dystonic features, diffuse muscular hypertrophy with hirsutism. Height was 145 cm (normal 157·716, SD 5·718) and weight 38 kg (normal 63·758, SD 9·911). Routine examinations, EEG, ECG, radiography, copper and caeruloplasmin, and slit-lamp examination were all negative. EMG and polygraphic recordings showed bursts at rest, irregularly rhythmic at 4-6 Hz, sometimes associated with isolated myoclonic jerks and with clonic or continuous activity lasting several seconds and clearly worsened by the voluntary movements, which also provoked abnormal spread of activity. EMG activity was also evoked on passive shortening of the muscle. CT scan, done twice, was normal. On repeated neuropsychological testings, (WAIS), IQ was 63 (verbal IQ 67, non-verbal IQ 64), with a mental deterioration of 20%. Hyperkinesias worsened just before and clearly improved during the menses. Therefore, we performed a thorough investigation of hypothalamic function (basal rates, spikes of gonadotropins, prolactin release, tests with RF, TRH and nomifensine, progesterone and basal temperature curves, in both the follicular and luteal phases) which, however, gave completely normal results.

Several drugs were tried from the age of 12 years: bromides, ACTH, amantadine, orphenadrine, levodopa, bromocriptine, fluphenazine, perfenazine, haloperidol, sulpiride, phenobarbitone, clonazepam, propanolol. They all had no effect. In spite of this, however, the patient is still self-sufficient, has no problems in walking and still works in a factory. She has a normal 9-year-old daughter.

We would like to stress the following points: in our patient abnormal movements developed 4 years after birth anoxia. They progressively worsened and spread to the four limbs, neck and voice very slowly in the course of the following 27 years. The abnormal movements were associated with mental and statural retardation, and the abnormal movements were very resistant to all therapeutical attempts. Evolving neurological deficits can thus follow a non-progressive cerebral lesion over a period of many years. They may not stabilise, and their clinical features may change during their evolution.[2] Progressively worsening abnormal movements and dystonia should therefore not be unquestionably taken as indicative of dystonia musculorum deformans.

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