SEXUAL PRECOCITY IN A BOY AFTER MEASLES ENCEPHALOMYELITIS

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The clinical impression that measles encephalomyelitis is becoming increasingly prevalent is confirmed by recent studies (Jacobsson and Holmgren, 1949). It is all the more necessary, therefore, to be alert to the sequelae of this baffling condition, sequelae whose rarity is probably exaggerated because of unfamiliarity with their mode of presentation. Thus, in the case to be described of a boy with sexual precocity, which seems quite clearly in retrospect to have been due to measles encephalomyelitis, the only diagnoses which were first considered were those of endocrine tumours.

Probably not more than one child out of 10 dies in the acute stages of measles encephalomyelitis; unfortunately, of those who survive more than half show definite evidence of permanent damage to the central nervous system (Ford, 1944). The commoner residual defects include spastic paralysis, cerebellar ataxy, mental deficiency, personality changes and epilepsy. Among the rarest to have been described is sexual precocity. This interesting sequel is mentioned in all surveys of the subject, though detailed original accounts are hard to discover. Three have been found (Boenheim, 1927, one case; Ford and Guild, 1937, two cases) all referring to girls whose ages (7, 9 and 10 years respectively) bring them rather close to the lower limits of normal puberty. The present report concerns a boy in whom sexual precocity developed soon after the age of 2 years. No other case has apparently been reported in a boy, nor has any case in either sex been described in this country.

Case Report

The patient is the second son in an apparently healthy family. He is said to have been normal at birth (birth weight 6 lb. 3 oz.) after an uncomplicated pregnancy. Development during the first two years of life was uneventful, and there was no delay in passing any of the usual milestones.

At the age of 2 years he had measles, but was ill for no less than 10 weeks. He was, according to the mother, 'sleepy all the time, and it was too much trouble to look at anything or do anything'. During this illness he was incontinent of urine and faeces, though previously these had been controlled. At night 'he slept on his hands and knees, and kept banging his head'.

After the illness he seemed, the mother says, 'different'. During the next year it was noticed that the genitalia were enlarging. He developed violent screaming attacks, which seemed gradually to be replaced by epileptiform seizures; for these he has since been treated medically. He grew rapidly in height and weight, and Fig. 1 shows his precocious general and genital development at the age of 4 years.

At 6 years he was unusually hairy and began to suffer from widespread acne vulgaris. A radiograph of the skull at this time showed no abnormality. At 7 years his voice 'broke'. He began to shave at 11 years, but, according to his mother, should have done so several years earlier.

He continued to grow rapidly, and at 8 years was taller than his brother who is 4 years older; growth in height has slowed down almost completely since the age of 12, and the brother is now much taller than the patient. Some figures of height and weight are fortunately available from school records. They show the patient at 5 years to have been 52½ in. in height and 85 lb. in weight, figures which would be expected at twice his age. At 10½ years he weighed 118 lb. and was 59½ in. tall. In the subsequent two years he has gained 15 lb. in weight but only ½ in. in height.

At the age of 12 years he was referred for an opinion because of uncontrollable 'giggling attacks', which had been coming on for some years. They occurred

FIG. 1.—Patient at 4 years, to show marked precocity of general and genital development.

at school, especially during prayers, and he tended to yawn afterwards and to feel rather tired. Very occasionally typical major epileptic manifestations had occurred despite continued treatment with anticonvulsants.

Physical Examination. He was at 12 years of age a big, plump, rather simple lad, with red cheeks and a frequent smile (Fig. 2). The skin was hairy and greasy, with active acne vulgaris on the face and trunk, and many scars of old acneiform eruptions. Striae atrophicae were present on the flanks and lower abdomen. The blood pressure was 110/80 mm. Hg. A functional systolic cardiac murmur was audible. The optic fundi and visual fields appeared normal. No abnormalities were found in the central nervous system or other systems.

Mental and Psychological State. Though the patient considered his ability to be average at school, this opinion was not shared by his schoolmaster who considered him rather backward. His I.Q. was unexpectedly low at 51. It had been assessed at 80 when he was 5 years old, and again at 10 years, though at 11 and 12 years of age it was estimated at 56.

There was no history of sexual activity; he is not particularly attracted to girls, and prefers the company of adults to children of his own age. His ideas as to vocation are variable, and at present he wishes to become either an actor or a B.B.C. announcer.

Investigations at the Age of 12 Years. Blood counts and blood sugar levels were normal. Routine urine examination revealed no abnormalities; the estimation of 17-ketosteroids (calculated as androsterone) gave a figure of 5.6 mg. in 24 hours, which is within the normal range. A radiograph of the skull was normal, but radiographs of long bones showed a moderate degree of

FIG. 2.—Patient at 12 years.

chronological advancement. An electroencephalogram showed a pathological tracing, with a wave form reminiscent of the wave-and-spike discharge of petit mal and a moderate potential theta discharge.

Comment

The description of the acute illness, particularly the features of protracted somnolence and loss of sphincter control, leaves little room for doubt, even in retrospect, that this was a case of measles encephalomyelitis. The abrupt changes in developmental pattern, which had previously appeared normal, date back to the time of the illness; to this unfortunate episode the mental retardation, cerebral dysrhythmia and sexual precocity must therefore be attributed.

The diagnosis in cases of sexual precocity is frequently tinged with out-dated ideas, and the consequent and often unnecessary tumour-hunt may do considerable harm. Jolly's (1951) recent survey synthesizes and simplifies the available information, and is a delightful diagnostic guide. The present case has no exact counterpart among Jolly's 66 cases, but clearly falls into his cerebral group with 'true puberty'. The single feature of enlarged testicles, with the clear-cut history, provides all the data necessary for a complete diagnosis.

I am indebted to Dr. F. L. Golla for information regarding the early history and for the first photograph, to Dr. A. Guiridham for helpful advice concerning the mental and psychological development, and to Dr. B. A. Astley Weston for providing details from the patient's school records.

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Vascular Abnormalities Associated with Pseudoxanthoma Elasticum

H. H. Wolff, J. F. Stokes and B. E. Schlesinger write as follows:

In our article (*Archives of Disease in Childhood*, 27, 82) we included an electron microphotograph of a skin biopsy taken from a patient with pseudoxanthoma elasticum. In the legend to this photograph and in the text the fibres shown on electron microscopy were referred to as elastic fibres. These fibres should have been described as normal collagen fibres. No elastic fibres could be detected on electron microscopy of the

material removed at biopsy. Similar discrepancies between the appearances obtained on light and electron microscopy have also been described in other cases of the same disorder (Tunbridge, *et al.*, 1952) in which elastic fibres were seen under the light microscope but not on examination by electron microscopy.

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