[Cerebellar Ataxia]

GRAND ROUNDS
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[redacted] a five year old white American boy entered [redacted] on [redacted] 1958 with the chief complaint of recurrent attacks of "incoordination". The child was the product of a normal full term pregnancy and uncomplicated delivery. After a normal to early developmental pattern and several minor but febrile respiratory infections while living on an isolated ranch in New Mexico, the family and patient moved to Montana in [redacted] 1956. In [redacted], 1956 at the age of 3 years 9 months the patient had febrile (102 degrees) pharyngitis and tonsillitis and numerous "mosquito bites". About the fourth morning of this illness he awakened afebrile, unable to walk properly. Because of incoordination and frequent falling he was taken to the hospital, where a diagnosis of tonsillitis and chicken pox was made. In addition, incoordination of the hands, a weaving gait, and falling on walking were noted. The child appeared frightened while in the hospital and during the five day stay there he wouldn't or couldn't talk. He was not drawsy, stuporous, incoherent nor confused, nor was there any vomiting or fever. Appetite was said to have been poor. Although the child had only 12 to 15 skin lesions he followed a typical course of varicella and his 2½ year old sister developed typical varicella about two weeks later. Hospital work-up included two lumbar punctures, both normal.

At home he continued rapid improvement over a period of 3 to 4 months. His gait improved, although with fatigue staggering became worse, and after 4 months he could run at play entirely normally. His hand movements continued to be clumsy, however, and the family noted almost constant small purposeless movements which were sufficiently subtle to be hardly noticeable by people outside the family. He exhibited more difficulty in fine movements of the hands than the average child, and still cannot tie his shoe laces. He likes to draw, but the objects drawn are unrecognizable. At the age of 4 years he was able to dress himself and to play with his toys, even building blocks, rather well. Speech on return home from the hospital was better but consisted of two or three word phrases instead of sentences which were hard to understand. Improvement in his speech was rapid, however, and he returned to normal for his age in about a month, except that he talked more slowly (and continues to) than he did before. No stuttering or stammering was noted. There were no muscle jerks, no facial grimaces, no tongue lolling, or chewing, and no gross purposeless movements. At no time was there emotional disturbance, behavorial change, mood abnormality nor depression.

In [redacted] 1956 a relative could not tell that he had been ill, but the parents still noted the busy fingers, slow speech, and clumsy gait with more falls than would be normal. He was well and developed normally during the next year, during which he had several febrile minor respiratory infections without any exacerbation of his neurological situation.

In [redacted] 1957, after stopping kindergarten because, although cooperative he was not able to do the things with his hands that the other five year olds were doing, he developed a minor respiratory infection with questionable fever. When this infection was about well his gait became quite wobbly again, and he could not walk without assistance. This time there was no speech or other motor involvement. The gait abnormality receded in two or three days, but the mother is quite sure that this could not have been simple weakness from his mild respiratory infection.

In [redacted] 1957 the patient developed typical rubeola. On the third day of fever he awakened with rash and inability to walk. He had to be supported by firm shoulder support to walk to the bathroom, his hands would shake and jerk so that he couldn't feed himself, and he exhibited periods of gross physical tremors of the whole body, especially early in the morning when taken to the bathroom. With intention, tremors of the extremities were moderately severe and persistent, although they have subsided to a large measure now. Speech was slower but not as severely involved as before. For the last month or so he has exhibited facial muscle jerks, biting, and chewing of tongue. He could walk with hand support by [redacted] 1958 and has improved tremendously during the month prior to this admission, at which time he was away from his mother.

Physical examination revealed an alert, cooperative, most appealing youngster with temperature 99.6, pulse 115, respiratory rate 25, height 41¼ in., weight 38 3/4 lbs. and blood pressure 110/65. The positive physical findings were as follows:- There were almost constant choriform movements of the head and extremities. Speech was slow and slurred but could be made out. Pupils were somewhat dilated but reacted to light and accomodation. Fundiscopic examination was unremarkable. Cranial nerves were intact, and there appeared to be no gross motor or sensory deficit. The deep tendon reflexes were absent bilaterally, but no pathological reflexes could be elicited. The ataxia and tremor appeared to be worse with intention. The eyes exhibited nystagmoid like jerks but not true nystagmus.

Laboratory data:- Hemagram-hemoglobin 12.4 gms., packed cell volume 35%, white count 10,200 with 50 segs, 44 lymphs, 3 eosinophiles and 3 monocytes. Red blood cells in platelets appeared normal. Urine specific gravity 1022, reaction 7, albumin sugar and microscopic negative. Sedimentation rate 18 mm. in one hour (Wintrobe),macrohematocrit 35%. Mantoux 1:1000 negative. Histoplasmin test negative, Coccidioidin test positive. Spinal tap revealed limpid clear spinal fluid under normal pressure, cell count was 3, all lymphocytes. Sugar 60 mg%, total protein 16 mg%, colloidal gold test non-reactive. Pa and lateral x-rays of the chest revealed no roentgen evidence of abnormality. Skull series revealed no roentgen evidence of abnormality. Electroencephalogram:- The recording was interpreted as probably abnormal, fast awake, and with various changes during sleep, particularly with suggestive spike wave discharges more on the left side. The positive spike clusters were considered significant mainly in an asymmetry in which they were reduced on the left. These findings all suggested a disorder that is greater in the left cerebral hemisphere.

Course in the hospital:- The patient was afebrile during his six day stay in the hospital. He was seen by numerous consultants whose opinions may be summarized as follows. [redacted] neurologist, considered this recurrent ataxia of childhood secondary to viral involvement of cerebellum and cerebellar spinal tracts. He did not recommend air study because "little can be hoped to be accomplished." [redacted] neurologist, agreed with [redacted] diagnosis and felt that improvement should continue but that the child will probably have some permanent residua. [redacted] ophthalmologist, found normal visual acuity, no evidence of disturbance in motility, and normal fundi. He felt that definite nystagmus was not present. [redacted] neurologist, considered that the findings suggest a cerebellar syndrome primarily of the vermis and presumably the result of post infectious encephalitis. He suggested

corticosteroid treatment as an anti-allergic or at least anti-inflammatory measure and stated that although the acute phase was subsided, there is evidence in other cases that the inflammatory reaction persists for many months. The child showed a gradual improvement in all symptoms during his six day stay in the hospital, although at the time of discharge he still exhibited ataxia, some tremor and some choreiform movements as well as hesitancy of speech.

Bibliography

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