REPORT OF A CASE OF MYASTHENIA GRAVIS PSEUDO-PARALYTICA WITH NEGATIVE PATHOLOGICAL FINDINGS[1]

By J. Arthur Booth, M.D.

of New York

consulting neurologist to the manhattan eye, ear and throat; french and perth amboy hospitals

Since the original contributions of Erb, Jolly, and Oppenheim, concerning the symptom complex of myasthenia gravis pseudo-paralytica, the study of this mysterious disease has been stimulated by the investigations of numerous observers on the functions of the ductless glands. Moreover, as a result of this study much that was formerly obscure in other peculiar disorders of the nervous system has been made more clear, and also certain theories more worthy of support. It is now interesting to note in connection with the disease under consideration that there are at the present time a number of reports, showing involvement of the thymus gland or pituitary body, with lymphorrhages in different organs and lymphocytic infiltration in the muscles. The most recent and important of these being that of Mandlebaum and Celler, who found all the above changes to a marked degree. The parathyroids are also receiving their share of attention, and Chvostek in a recent review presents evidence in favor of the assumption that the symptom complex of the disease is best explained as the result of defective functioning on the part of these glands. Yet with all the advantages derived from improved methods in the preparation of specimens and histological study, cases of the disease are still recorded in which no lesions are found to satisfactorily account for the symptoms. Believing such contributions are not only of interest, but also of importance, the writer takes this opportunity of offering for your consideration the following history:

[1] Read at the thirty-fourth annual meeting of the American Neurological Association, May 20, 21 and 22, 1908.

The patient, Ralph A——, a lad of 11 years, was seen with Dr. William H. Haskin at the Manhattan Eye, Ear and Throat Hospital on October 16, 1907.

Family History.—The father and mother are both living, the former a drunkard. The mother has two sisters and one brother, all insane. She has had seven children and five miscarriages. Three children died during infancy from diarrheal disorders. Of the four living children our patient is the youngest. An examination of the others does not show any form of nervous disorder.

The mother states that he was healthy during infancy and childhood, with the exception of a mild attack of measles when four years old.

In his seventh year he had an acute attack of otitis media, followed by a purulent discharge from the left ear; this received treatment and soon ceased. In April, 1906, he was operated on for adenoids and from this dates the onset of his present illness. Within a few days following the operation he became easily tired, was listless and complained of a general muscular weakness, which would disappear after lying down for a time and then return on exertion or after any excitement. He was bright mentally, standing well in his class at school, but his attendance was interrupted by frequent periods of extreme lassitude and weakness, during which he would remain at home, lying down most of the time. In April, 1907, he complained of his eyelids feeling heavy and these finally drooped; then there rapidly developed a progressive weakness of the facial muscles and a difficulty in raising the arms and going upstairs. He also talked in a peculiar voice and had difficulty in chewing and swallowing his food. Any shock or excitement seemed to increase this disability, and at such times he would become almost powerless. Recently his neck muscles have become weak.

Examination.—The patient is poorly nourished, but not emaciated. The mentality is good and he answers questions intelligently, though in rather a feeble voice with a nasal tone. The expression of the face immediately attracts attention, being flat and expressionless, accompanied by a bilateral ptosis, and he says this drooping of the eyelids is worse at night than in the morning. The lips are not held together and he is not able to pucker them or whistle.

Muscular Status.—Bilateral ptosis and diplegia facialis. The muscles of the arms, shoulder girdle and thighs are flabby, but there is no degenerative atrophy or fibrillary twitchings. The calf muscles seem firm and hard compared to other muscles of the body; the right calf measuring 9½ inches and the left 9¾ inches. All voluntary movements are performed slowly and are soon followed by exhaustion. Lying down on the floor and requested to get up, he does so at the first attempt quickly, but

after one or two repetitions, he is unable to accomplish this without artificial aid. This same condition of induced fatigue is present in the speech and swallowing mechanism. The grasp of the hands is weak, only five of the dynamometer in each hand. The gait is slow, but there is no dragging of the feet or any evidence of spasticity. The pupils are equal and react to light and accommodation. The fundus is normal. Reflexes: All reflexes present and normal. The knee jerk response varied, being sometimes present and then again absent, especially after six to eight taps.

Sensation.—There is a slight loss of tactile sensation over the inner side of right arm and right face. No analgesia or loss of temperature or muscular sense. With the exception of a slight defect of hearing in left ear, all the special senses are normal.

Electrical Reactions.—To galvanism all muscles react normally and to faradism there is a contraction and finally exhaustion of the response in the sterno-mastoids, trapezii, deltoids and extensor communis digitorum of each side.

Diagnosis.—In the absence of hereditary disease of similar kind in the family history, the sudden onset following shock and the presence of marked bulbar symptoms, with the results of the electrical reactions, we can eliminate the pseudo-hypertrophic or Landouzy Dejerine type of myopathy. On the other hand, taking into consideration the variability of the symptoms, increased by emotion and excitement, together with the clinical history, seemed to warrant the conclusion that the symptoms were those of myasthenia gravis pseudo-paralytica.

With this diagnosis he was transferred on November 12, 1907, to St. Luke's Hospital for further observation. Here the rapid variation in the severity of the symptoms from time to time was marked. I may mention, to illustrate this, that I attempted to bring him before the Neurological Society, but he had an attack of respiratory failure on the way to the meeting, his condition became alarming and I was forced to return to the hospital. After this he grew rapidly worse and had several similar attacks, in one of which he died on November 18, 1907, at 5 P. M. During the six weeks he was under observation the temperature was normal, the pulse varied between 60 and 90 and respirations between 20 and 40. Just before death the temperature rose to 100⅘, pulse 100 and respirations 40.

Post Mortem Report.—The autopsy was performed one hour after death by Dr. Francis C. Wood, pathologist of St. Luke's Hospital and Columbia University: Body emaciated. No rigor mortis. Cartilages and ribs rather prominent. Skeletal muscles not well developed but apparently normal. Lungs and heart present nothing abnormal on section. The thymus gland is unusually well preserved for the age and is considerably enlarged; extending from the upper border of the sternum, as a long, flat

boat-shaped mass and measuring about 11 by 5½ centimeters in width and one centimeter thick. Thyroid gland normal and mediastinal glands slightly enlarged. Abdominal organs: Coils of intestines collapsed. Liver, spleen, stomach, pancreas, kidneys and suprarenals are all apparently normal. Head: Brain apparently normal and weighs 3½ pounds. Pituitary body normal. Spinal cord normal. Specimens from different muscles, nerves, brachial and sacral plexus taken for further study.

Microscopical Examination.—The muscles, both voluntary and involuntary, including that of the heart and tongue; the detoid, brachial, extensors of the thigh, abdominal and pectoral, show no changes from the normal; there being no evidence of lymphocytic infiltration or degeneration. The thyroid and parathyroids show no lesion.

The thymus gland, which was enlarged, is normal except for an occasional area of focal necrosis. The Hassall's bodies are well marked. The lymphoid tissue is normal in distribution and amount. There is a moderate growth of connective tissue between the lobules, but no more than is normal in a child of this age. No lesions could be found in the nerve trunks. The lungs and heart are normal. The liver and kidneys show slight parenchymatous degeneration; the latter being slightly congested. The pituitary body and suprarenals are also normal.

Central Nervous System.—On inspection the pons, medulla and spinal cord were normal. Sections were made and examined from the following situations; through the fourth and first cervical segments, at the lower part of the medulla, through the medulla at the level of the eleventh and twelfth nuclei and four others higher up, of which one was through the vagal nucleus. The cells of the nuclei, the fibers, vessels and all the tissues in every section appear normal, hence any organic lesion of these parts of the nervous system may be safely excluded.

Summary.—Considering the clinical history, the symptoms and course of the disease in the case just cited; the diagnosis of myasthenia gravis pseudo-paralytica was justified. The initial symptoms came on after an operation for the removal of adenoids; they were rapidly progressive and later any emotional excitement caused an increase in their severity. During the six weeks he was under observation he had three attacks of respiratory failure after slight exertion and each was due to this cause. Examination of various muscles, done very thoroughly, did not show any tendency to muscle degeneration or the presence of lymphoid infiltration. The brachial plexus, the sacral and the nerves themselves were also normal. With the exception of a simple slight hypertrophy of the thymus gland, the glandular

structures were normal, including the thyroid, parathyroids and pituitary body. The examination of the central nervous system revealed no lesion or pathological condition to which the paretic symptoms might be attributed.

Conclusions.—In view of the above facts and the various pathological findings recorded by others in similar cases, we must conclude that the symptom complex of the disease is best explained on the basis of its being a nutritional disorder impairing the vital processes in the muscles from some unknown toxine.

In conclusion, I wish to express my thanks to Dr. F. C. Wood and members of the pathological staff of St. Luke's Hospital and also to Dr. M. G. Schlapp for his examination of the central nervous system.