PSEUDOSCLEROSIS OF STRÜMPELL-WESTPHAL IN FIVE MEMBERS OF A FAMILY.*

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INTRODUCTION.

IN 1878 Strümpell¹ reported a case resembling multiple sclerosis in which he found abnormal hardness and consistency of the brain. He suggested several possible causes, including hypertrophy or atrophy of the brain, senility, alcoholism, saturnism, and acute or chronic inflammatory processes. In 1883, Westphal² reported two cases resembling multiple sclerosis, but the microscopic examinations of the brain and spinal cord were negative. These cases showed progressive dementia, which appeared early in the course of the disease, and were characterized by absence of nystagmus.

Between 1883 and 1905 fourteen cases were reported in the literature in which it was noted that the brain and cord were unusually firm. Potts and Spiller³ reviewed these and concluded that one could not differentiate pseudosclerosis from disseminated sclerosis; they stated that the unusual firmness must be caused by a proliferation of the neuroglia even if it cannot be detected by a microscope. Hugo Weiss⁴ described the disease as characterized by headaches, dizziness, fatigue, changes of mood, facile laughing and crying, transient double vision, speech disturbances and rhythmic spasmodic movements of the extremities. Rebezzi⁵ thought that pseudosclerosis and multiple sclerosis were manifestations of the same disease-process and should be considered as one disease due to primary affection of the nervous elements with secondary increase of glia-cells. He proposed the name 'Westphal-Strümpell disease.' Oppenheim⁶ in his text-book states it is difficult, if not impossible, to differentiate pseudosclerosis from disseminated sclerosis in life.

In 1902, Kayser⁷ referred to the occurrence of peripheral annular pigment of each cornea in a patient with nervous symptoms similar to those of disseminated sclerosis. In each eye there existed an annular peripheral pigmentation of the cornea, which began at the limbus and gradually faded away centrally so that the central oval area of the cornea remained normal.

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The peripheral parts were made up of an accumulation of yellowish granules which became finer and more diffuse centrally, where the colour changed to a yellowish green. Fleischer⁸ later worked with this same patient, who subsequently developed more nervous symptoms; eventually he developed glycosuria, and a greenish-brown pigmentation of the head and hands became evident with a greyish discolouration over his chest and abdomen. At autopsy the liver was seen to be small and nodular; the spleen was enlarged. There were no gross or microscopic signs of multiple sclerosis. The cornea showed pigmentation of Descemet's membrane, extending centrally for about 1.9 cm. The deposit consisted of rounded and angular greenish-brown granules that appeared refractile at a certain angle. This pigment was also found in the liver, spleen, intestines and kidneys. A chemical examination of some of the granules in kidney and spleen seemed to indicate the presence of silver, but certain other tests made this chemical diagnosis improbable⁹. There was no history of the patient having had silver administered, while in argyrosis mental symptoms rarely, if ever, occur.

In 1912 Wilson¹⁰ described the disease known as progressive lenticular degeneration. He said it was a disease which occurred in young people, which is often familial, but not congenital or hereditary; it is essentially and chiefly a disease of the extrapyramidal motor system and is characterized by involuntary movements, usually of the nature of tremor, dysarthria, dysphagia, muscular weakness, spasticity and contractures with progressive emaciation; with these may be associated emotionalism and certain symptoms of a mental nature. It is progressive and after a longer or shorter time fatal. Pathologically it is characterized predominantly by bilateral degeneration of the lenticular nucleus and, in addition, cirrhosis of the liver is constantly found, the latter morbid condition rarely if ever giving rise to symptoms during the life of the patient.

In his paper, Wilson did not mention the corneal ring. Pollock¹¹ in 1917 reported a case of Wilson's disease with corneal pigmentation. Hall¹² in 1921 in a complete review of the literature states that the corneal ring is pathognomonic for the diagnosis of pseudosclerosis or of Wilson's disease. Of sixty-eight cases which he reviewed, the corneal ring was not mentioned in thirty-one. Of the remaining thirty-seven cases the ring was present in sixteen cases of pseudosclerosis and four cases of Wilson's disease and in one case of doubtful diagnosis. In five cases of Wilson's disease and eleven cases of doubtful diagnosis the ring was not present.

Hall reviewed the literature concerning the chemical nature of the pigment and was unable to settle the question. The possibility of its being some kind of an endogenous pigment—of the hæmoglobin group—which does not give the iron reaction, appears to be the most probable theory.

SYMPTOMATOLOGY.

Pseudosclerosis is frequently a familial disease affecting people usually between the ages of fourteen and twenty-six. It is a disease in which remissions frequently occur and itself may last fifteen to twenty years. The symptom-complex includes (a) neurological signs; (b) corneal pigmentation; (c) mental signs; and (d) cirrhosis of the liver.

(a) The neurological signs include:—

(1) Speech disturbances (the voice is slow, monotonous, indistinct, or staccato in type).

(2) The face is usually 'fixed,' with a tendency to grimacing.

(3) There is marked intention-tremor with some muscular rigidity, but rigidity is less marked than in Wilson's disease.

(4) Gait is of parietic-ataxic type.

(5) Cerebellar signs, as adiadokokinesis or dysmetria, occur.

(6) Exaggerated deep reflexes with retention of abdominals and plantar flexor responses are found.

(7) Occasional apoplectiform or epileptiform seizures¹³ may occur.

There is no pallor of optic discs, no nystagmus, no loss of bladder control.

(b) The finding of annular greenish-brown corneal pigment is considered pathognomonic of either pseudosclerosis or Wilson's disease¹⁴.

(c) Instead of showing euphoria, as is the usual finding in multiple sclerosis, the patients exhibit marked fluctuation in mood. They become irritable on the slightest pretext, and quarrel a great deal. At times they become maniacal, confused, or suicidal. They have frequent outbursts of crying, and are very untruthful. Mental deterioration is frequently found.

(d) The cirrhosis of the liver is usually asymptomatic, and is found only at autopsy. Occasionally during life the liver or spleen may be palpated or liver function tests may indicate liver damage.

PATHOLOGY.

Spielmeyer¹⁵ believes that the same disease-process is common to both Wilson's disease and pseudosclerosis and that a differentiation on histopathologic grounds is not possible.

Wilson's disease is held to be a degenerative process of the corpus striatum, the cerebral cortex showing no noteworthy changes. Alzheimer finds in pseudosclerosis (Westphal-Strümpell) the principal lesions in corpus striatum, optic thalamus, subthalamic region, pons and dentate nucleus of the cerebellum, but lesions also occur in other parts of the brain and especially in the cerebral cortex. The essential pathological picture is

thought to be the occurrence of the giant glia-cells with large lobulated nuclei poor in chromatin, and scanty cytoplasm.

Uchimura¹⁶ described a case in which the striatum and pallidum showed an increase in nuclei of a glial nature. These were of two types: (1) small cells filled with fat, and (2) typical large Alzheimer glia-cells. In the putamen there was perivascular infiltration with fat-laden cells. The most striking feature was the accumulation of such cells in the striatum. Similar changes were found in the external capsule, anterior commissure, and in the lowest portion of the internal capsule. The fatty degeneration was sharply localized.

Bielchowsky and Hallervorden¹⁷ give the clinico-pathological report of two cases of pseudosclerosis which at autopsy showed symmetrical fusion of the lesions in the frontal lobe.

DIFFERENTIAL DIAGNOSIS.

Multiple Sclerosis.—Pseudosclerosis is differentiated from multiple sclerosis by the absence of optic disc pallor, absence of nystagmus, lack of involvement of the bladder, intact abdominal reflexes, negative Babinski sign, and the mental picture, which differs from the euphoria of disseminated sclerosis.

Juvenile Parkinsonism.—Many cases of pseudosclerosis including two of the cases to be reported here have been diagnosed as postencephalitic Parkinsonism. A masklike facies is seen in both conditions. The tremor in Parkinsonism is constant at rest, and ceases on voluntary effort, while in tremor of pseudosclerosis is of the intention-type. In Parkinsonism the liver is not involved nor is the corneal ring present.

Wilson's Disease.—Some writers, including Higier¹⁸ and Rausch and Schilder¹⁹, regard Wilson's disease as a special form of pseudosclerosis. Wilson, in Lewandowsky's *Handbuch*, states that some cases of pseudosclerosis seem to be nearly related to progressive lenticular degeneration. Stocker²⁰ believes the mental condition is sufficient to differentiate between the two. In Wilson's disease the mentality remains intact longer and the patient is usually euphoric; in pseudosclerosis early dementia occurs with characteristic mental signs of irritability, attacks of violent temper, etc.

In Wilson's disease both hypertonicity and intention-tremor are seen, while in pseudosclerosis the predominant symptom is thought to be the latter. No swallowing difficulties occur in pseudosclerosis. Dysarthria and dysphagia are frequent and early signs in Wilson's disease, eventually leading to cachexia. As a rule no cerebellar signs are found; there are few or no remissions, and the disease has a shorter course of three to nine years.

ETIOLOGY.

Westphal and Sioli²¹ adduce evidence for the infective origin of pseudosclerosis. In their case the syndrome followed epidemic encephalitis.

Pathological examination showed cirrhosis of liver, Alzheimer glia-cells, proliferative changes in the vessels, and collections of small round cells. Campbell and Moore²² report the case of a printer with erysipelas who developed mental symptoms with marked tremor of hands and head and difficulty in articulation. At autopsy cirrhosis of the liver and typical large glia-cells in cortex, basal ganglia and midbrain were found. In this case the first nervous symptoms (prolonged sleep) appeared with an attack of facial erysipelas; a second attack of erysipelas was associated with a period of stupor; the patient died during a fourth attack. In Simpson's²³ case the father and mother were cousins. In the cases to be reported here the father and mother were first cousins.

PROGNOSIS.

Remissions and exacerbations have been reported, and patients have been known to live for fifteen or twenty years. The prognosis is considered better than in Wilson's disease, which usually becomes progressively worse.

TREATMENT.

No specific therapy is known. Liver therapy has been suggested. Some writers propose the use of decholin, a cholic acid derivative ($C_{23}H_{33}O_3COOH$), giving 10 cc. of a 20 per cent. solution intravenously. Decholin is intended to increase the functional activity of the liver.

PERSONAL CASE REPORTS.

Of the five cases to be reported here, two patients are at present in neurological hospitals, a third is on parole from a state hospital, and the other two, although living at home and going to school, are having difficulties in adjusting satisfactorily in the community.

Family History.—The patients are of Russian Jewish descent. Familial mental and nervous diseases are denied. The paternal grandfather died at 68 of heart trouble. The paternal grandmother died at the age of 57 following a cerebral accident. The maternal grandfather died at 40 of typhus. The maternal grandmother is living and well at the age of 80.

The father and mother were first cousins. The former died at age of 37 of pemphigus (eight years ago); a paternal uncle and aunt are living and well. The mother, age 40 years, has marked emotional instability. Her speech is expressionless. There is a suggestion of a corneal ring in her left eye. Her physical and neurological examination is quite negative. Icteric index was three. Direct and indirect Van den Bergh tests were negative. A maternal uncle and two maternal aunts are living and well. Three maternal uncles died in childhood, one of measles, one of pneumonia and one of typhus.

The mother had seven children and three miscarriages.

PREGNANCY 1.—A miscarriage at about six months following a fall down stairs.

PREGNANCY 2.—A boy, H. W., age 21 years, who at present is on parole from a New York State Hospital, where his case has been diagnosed as psychosis with psychopathic personality. It had once been considered postencephalitic Parkinsonism.

This patient reached the third year of high school at the age of 15. At the age of 13 he fell off a truck, hit his head, and was unconscious for two hours; at the age of 16 he had what was diagnosed as chorea.

At the age of 18 there was noted a gradual slowing of all the patient's movements. His speech became thickened, indistinct and at times explosive. He became quite unstable emotionally, laughing loudly at times; he also developed marked irritability and would quarrel with his mother and sisters and frequently was very brutal to them. He was at a State Hospital for thirteen months where he was considered to be mentally deficient. Within the past few weeks he has had two or three epileptiform seizures.

Physical and Neurological Examination.—Patient is fairly well developed and nourished. He is of short stature, with marked vasomotor instability and thick spadelike hands. Heart and lungs are negative. Abdomen is normal. Liver and spleen cannot be palpated. Pupils are equal, regular, react to light and accommodation. The corneal ring of pigment is present bilaterally. There is no nystagmus. Facies is masklike. There is a doubtful right facial weakness. Speech is dysarthric and explosive. Other cranial nerves are negative.

He has a slow simian gait. When in the Romberg position he tends to fall backward. He has good power in all his limbs. There is a slight intention tremor of the left hand, and bilateral adiadokokinesis. He requires considerable time to perform simple tasks such as buttoning his clothes. Deep reflexes are active and equal. Plantar flexion responses occur bilaterally. Touch, pain, position and vibratory sensations are intact. There is no astereognosis.

Laboratory Findings.—Blood Wassermann, negative. Icteric index, 5. Van den Bergh (direct and indirect) negative. Liver tolerance test (intravenous injection of bromsulphalein) normal.

X-ray of the skull was not obtained as patient refused to enter the wards and to have any further examination done, apparently because his first epileptiform seizure occurred after a venous puncture for blood for the Wassermann reaction.

PREGNANCY 3.—A boy, B. W., died at the age of 8 days. This was a case of instrumental delivery.

PREGNANCY 4.—A girl, R. W., age 17 years, was admitted to the Psychiatric Ward at Bellevue Hospital on September 30, 1931. She was transferred from a neurological hospital where she was said to have been 'excited, unmanageable, profane, seeking the company of men, having emotional upsets of laughing and crying.' Her case had been diagnosed as one of chronic encephalitis.

She was admitted to the neurological service at Bellevue Hospital on October 15, 1931, at which time she complained of (1) tremor of the head and upper extremities of three years' duration; (2) difficulty in walking with a staggering gait of three years' duration; (3) thickened slow speech for three years; (4) double vision of one week's duration.

At the age of 7 she was jaundiced and was told she had an enlarged liver; at the age of 10 she had rheumatic fever. She had pneumonia and influenza at the age of 14. She reached the seventh grade of school at the age of 11. She had only one menstrual period which occurred at the age of 15.

Patient believes that all her neurological symptoms occurred after she had pneumonia and influenza three years ago.

Physical and Neurological Examination.—Patient is a fairly well developed and well nourished girl. Heart and lungs are negative. Abdomen is normal. Liver and spleen cannot be palpated. Fields and fundi are negative. The corneal ring is present bilaterally. There is no nystagmus. Pupils are slightly irregular and unequal, the left being larger than the right; both react

promptly to light and accommodation. Speech is slurring and monotonous. The gait is spastic, with a positive Romberg sign. Marked intention-tremor is present in both upper limbs; this can be easily demonstrated in the finger-finger and the finger-nose tests. Bilateral adiadokokinesia is present. The deep reflexes are hyperactive and equal; the abdominals are sluggish but can be elicited. Negative Babinski signs present. Sensation is normal.

While in the ward patient was irritable and quarrelled a great deal with her sister and with other patients; she constantly sought attention from physicians and nurses. She would lose her temper readily and then would cry in a very loud manner, disturbing the others.

A psychometric examination revealed that the patient had an intelligence quotient of 68 per cent. on the Stanford-Binet scale.

Laboratory Findings.—The urine showed a faint trace of albumen. No casts were present. Manometric examination showed an initial spinal fluid pressure of 90 mm. water pressure. No cells were seen in the spinal fluid. There was no globulin and no sugar. Colloidal gold curve: 0001100000. Blood and spinal Wassermann tests: negative.

Blood Picture.—Red blood count, 3,940,000; hæmoglobin 80 per cent. (Tolquist); white blood count, 8,050, with 58 per cent. polymorphonuclear leucocytes. Non-protein nitrogen, 30; blood sugar, 100, and creatinin, 1.0. Icteric index: 6.0. Van den Bergh tests (direct and indirect), negative. Basal metabolism test was -12 per cent. Liver function test showed an abnormal dye retention; at the end of two hours 15 per cent. of the dye was present in the blood stream. X-rays of skull and spine were negative.

PREGNANCY 5.—A girl, D. W., age 15 years, was admitted to the neurological service at Bellevue Hospital on October 4, 1931, with the complaints of (1) tremor of both hands of two years' duration; (2) difficulty in talking for two years; (3) trouble in sleeping for two years; (4) double vision present occasionally during last two months; (5) occasional dizzy spells present during the past few months, usually associated with menstrual periods; (6) protruding anal mass present for a few weeks.

Patient's past history was essentially negative except for an injury to her right foot eight years ago, a piece of glass having infected her right heel. Since then she has been treated intermittently in several orthopædic hospitals, and skin grafts have been done. A resection of the posterior tibial branch of the nerve to the right heel was also performed. Alcoholic injections of the posterior tibial nerve were given but the pain in the heel has persisted.

For the past two years patient has noticed progressive weakness and an intention-tremor of her hands, accompanied by the other symptoms above mentioned. The mass protruding from the anus bleeds and is painful whenever patient defæcates.

Physical and Neurological Examination.—Patient is a rather poorly developed and nourished girl. Heart, lungs, and abdomen negative. Liver and spleen not palpable. Condylomata acuminata present. Right foot-drop present. Scars on abdomen on both sides present, said to be source of skin grafts. Cicatrix of right heel present, tender to touch. Right leg is shorter than the left.

Fields and fundi are normal. No nystagmus. Bilateral corneal ring present. Pupils are slightly irregular and unequal, the right being larger. Both react promptly to light and accommodation. Marked slurring of speech. Tremulous tongue. Patient drags right leg in walking. Positive Romberg sign. Marked intention-tremor of hands. Bilateral adiadokokinesia. Deep reflexes hyperactive but equal. Abdominals cannot be elicited. Bilateral plantar flexor responses.

Psychometric Tests.—Two years ago she had an intelligence quotient of 103 per cent. At the present time the I. Q. is 73 per cent. on the Stanford Binet Scale and 64 per cent. on the Army Performance Tests, while the psychologists report that the patient is very co-operative and that the findings are representative.

Laboratory Findings.—Urine, negative. Blood chemistry showed sugar, 98; creatinin, 1.3; and non-protein nitrogen, 35. Icteric index, 6.0. Van den Bergh tests (direct and indirect), negative. Blood Wassermann, negative. Manometric test showed an initial pressure of 80 mm. water with an open system. Spinal fluid showed no cells, globulin or sugar. Spinal Wassermann was negative and the colloidal gold curve was 0011000000. Liver function test showed no retention of the dye after two hours. The blood examination revealed a red blood count of 3,740,000, with 75 per cent. hæmoglobin; and a white blood count of 12,350 with 58 per cent. polymorphonuclear cells.

Basal metabolism was + 14 per cent., but examination was not considered satisfactory because of the restlessness of the patient.

X-ray of skull, dorsal, lumbar and sacral spine was negative.

The patient presented a marked problem in the ward because of her mental aberrations. She showed marked emotional instability, laughing or crying in a very loud manner on the slightest pretext. She quarrelled almost constantly with the other patients. She told lies constantly about the doctors, nurses and patients. She threatened suicide several times. She left the ward several times and was found in other parts of the hospital flirting with other patients. She made sexual advances to the various janitors and orderlies whom she encountered. She would often cry for hours at a time when reprimanded for her behaviour and would keep other patients awake at night.

It must be remembered that she was suffering some pain because of the condylomata which were being treated with deep X-ray therapy. However, a few tablets of placebo were sufficient to stop the crying spells even when she complained of considerable pain.

Her behaviour finally became such that it was necessary to transfer her to the psychopathic department.

PREGNANCIES 6 AND 7.—Twins—girls. The mother could not tell if the twins were identical; the delivery occurred at home without the aid of a physician. They were said to be so similar in appearance that until a year ago even the mother could not distinguish between them. At the present time the resemblance is not very noticeable.

(6) Y. W., age 13½ years, attends school. She has had to repeat two grades and now is in the 7B Grade. She is restless, quarrelsome, insolent. She has an intelligence quotient of 116 per cent. but has marked difficulty in adjustment in school.

The positive neurological findings here include the corneal ring bilaterally, a slow slurring speech and some adiadokokinesia.

This patient would permit no laboratory examinations. She made several appointments to see examiner but did not keep them.

(7) R. W., is in the eighth grade although she had an I. Q. of 110 per cent. She has had to repeat some of her school work. She, too, is irritable and restless.

The positive neurological findings here include the corneal ring bilaterally and a slight speech defect.

PREGNANCY 8.—A non-induced miscarriage at four months.

PREGNANCY 9.—A girl, F. W., age 11 years.

This patient is in the sixth grade at school; she is very quarrelsome, frequently runs away from home, and is entirely uncontrollable. When she visits her sisters she is restless, sarcastic, and quarrels constantly. She would permit no

physical or neurological examination except for an examination of her eyes. No corneal ring was present. Eye movements, pupils, etc. were normal.

PREGNANCY 10.—A miscarriage.

SUMMARY.

The literature regarding Westphal-Strümpell's pseudosclerosis is briefly summarized and the differential diagnosis between this condition and possible allied neurological conditions (especially multiple sclerosis, Wilson's disease, and juvenile Parkinsonism) is discussed. The Kayser-Fleischer corneal ring is mentioned as a pathognomonic sign of pseudosclerosis or Wilson's disease. Case-reports of five children in one family having this corneal ring with unusual neurological signs and mental symptoms are recorded. The parents of these children were first cousins.

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