

INTRACRANIAL TELANGIECTASIS

REPORT OF TWO CASES *

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The two following cases of probable cerebral telangiectasis seem to be of sufficient interest to warrant report.

REPORT OF CASES

CASE 1.—History.—A merchant, aged 21 years, was referred by Dr. Ransom of Hancock, Minn., July 14, 1920. The patient's family history was negative except that his father died of diabetes. When 7 years old, he was thrown from a horse and struck on his head, but he remembered no details of the accident; appendectomy was performed on him when he was 14; he denied venereal disease. When 8 years old, while convalescing from an attack of mumps, he had a severe convulsion and remained unconscious for fourteen hours. He recovered and was well for three weeks, when he had another mild attack. Since that time the number of attacks have varied from three in one day to one in six months. He described a typical attack as follows: It begins with a sensation of electricity in the left hand, after which he feels as if the hand were off. Within a few minutes, he notices a jerking of the fingers of the left hand, gradually involving the muscles of the entire left arm. When it reaches the shoulder, he becomes semiconscious and feels as if he were floating in space. During this semiconscious period, he has excruciating pain in the left arm. Then he loses consciousness for a period of from three minutes to two hours. When he recovers consciousness he is confused and talks irrationally for a short time and his left arm feels weak and awkward for several hours. He has abortive attacks during which the hand becomes numb and the arm jerks, without loss of consciousness. Frequently he can arrest severe attacks by placing a rubber band tightly around the left arm. For the past four years he has had a constant feeling of numbness in the left hand. He does not have headache.

Examination.—The neurologic examination was negative except that he had impaired tactile sense of the left hand and decrease of abdominal and cremasteric reflexes on the left side. The fundi were normal. There was no telangiectasis on the face or body. Examination made three days later when he was complaining of indefinite general disagreeable feelings, revealed in addition a positive left Babinski and Gordon reflex and left ankle clonus. These findings were variable. The laboratory findings were: hemoglobin, 98 per cent.; leukocytes, 7,800; red blood cells, 4,480,000; blood pressure, — 110 systolic, 55 diastolic; urine, normal. The blood Wassermann reaction was negative. Spinal Fluid: pressure somewhat increased, no globulin, 4 lymphocytes, negative Wassermann reaction and colloidal gold curve.

* Read before the Minnesota Academy of Medicine, April 14, 1921.

Diagnosis.—A diagnosis of jacksonian epilepsy due to irritation of the right motor region, cause undetermined, was made and craniotomy was performed Aug. 2, 1920, by Dr. Harry Ritchie.

Treatment and Course.—Enormously dilated blood vessels were found on the pia over the right motor region, arranged in the form of an irregular circle. These were ligated in four places and the dura was closed. During the next twenty-four hours the patient had twelve convulsions of such severity that chloroform had to be administered. The following night he had two more and was then placed on two-thirds grain of luminal three times a day. Up to the present time he has had only one unconscious seizure but at about weekly intervals he has had twitching of the left arm with blurred vision lasting a few minutes.

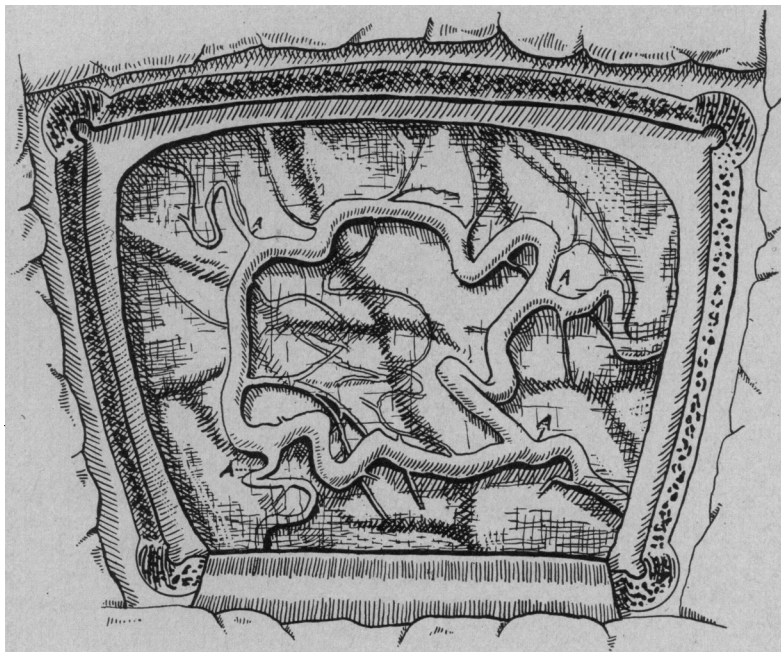


Fig. 1 (Case 1).—Jacksonian epilepsy; anterior upper margin of right ear. The vessels were ligated at A, A, A, A; $\times 1.3 +$.

Comment.—Sachs reports two similar cases and believes that the clinical picture is sufficiently typical to enable one to arrive at a definite diagnosis. The outstanding symptoms are jacksonian epilepsy at long intervals in a nonsyphilitic, with unconsciousness of long duration, no evidence of intracranial pressure, slow progression of symptoms and telangiectasis on the head or face. With proper surgical interference, he makes an optimistic prognosis. Our patient conforms fairly well to this clinical picture, with one important exception—there was no evidence of superficial telangiectasis. In one of Sach's cases, the angiomatous process was found in the dura with numerous connections with the pial vessels.

CASE 2.—History.—Our second case was that of a farmer, 26 years old, first seen in consultation with Dr. Robert Earl, July 8, 1915. His family history was negative except that the grandfather, mother and one sister suffered from periodic headaches. His personal history was unimportant except for an occasional headache prior to the accident. In July, 1914, while running, he struck the top of his head against a protruding heating pipe. He was unconscious for thirty minutes and was sent to a hospital where he remained for two weeks, during which time he was somewhat stuporous. Recovery was complete but ten months later he developed headache in the posterior mid-frontal region. This headache was progressive, more severe in the evening and so distressing that the patient threatened suicide. Within one month he developed definite mental symptoms—he thought that people were talking

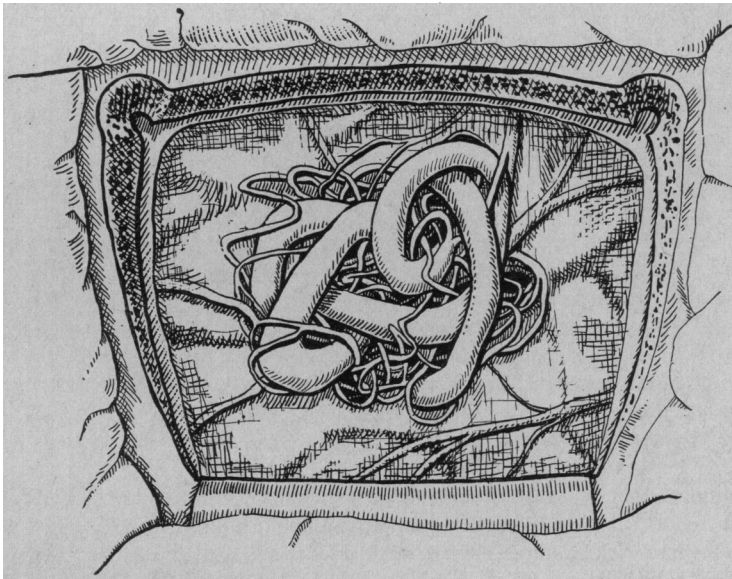


Fig. 2 (Case 2).—Multiple head injuries; inner canthus of the right eye; $\times 1.3+$.

about him and plotting against him and he threatened to harm them. He was also somewhat resistive and at times most unreasonable. Sleep and his general condition were satisfactory.

Examination.—Neurologic examination made one year after the accident was negative throughout except that both knee jerks were sluggish but normal on reinforcement; there was a small area of marked tenderness to pressure over the posterior portion of the right frontal region. All laboratory findings were normal. The spinal fluid was under slight pressure, the globulin test was negative, 5 lymphocytes per cubic millimeter; the Wassermann test and colloidal gold curve were negative. A roentgen-ray examination of the head was negative.

Diagnosis.—A diagnosis of beginning dementia praecox or of a cyst secondary to the trauma in the right frontal region was made.

Treatment and Course.—Because of extreme tenderness over this area, operation was advised. Dr. Robert Earl made a right frontal bone flap and found

the dura adherent to the skull. The dura appeared bluish, which was found to be due to a marked angiomatous condition on the surface of the brain cortex. This mass of blood vessels covered the entire operation field and appeared like a nest of bluish angleworms. Vessels were ligated in several places; bleeding was profuse but easily controlled. The patient made an uneventful recovery both physically and mentally.

Subsequent History.—The subsequent history is of interest. On Aug. 30, 1916, he was struck on the head by a small plank, which rendered him dizzy for a short time; after this he had occasional headaches. On June 6, 1917, he was blown up by an explosion, striking his head forcibly against a stone wall. His headaches were decidedly worse after that. On July 30, 1917, he stood for a time bareheaded in the hot sun, after which his headaches were so severe that large doses of morphin were required. Since October, 1916, spicules of bone had at times come through a discharging sinus at the site of operation. He was seen again Aug. 6, 1917. All neurologic findings were negative; the spinal fluid was normal. The old bone flap was loose at the median line and could be pressed in. On Aug. 8, 1917, Dr. Robert Earl transplanted two tibial grafts to fill the defect in the skull; the dura appeared normal except that there were extensive hemorrhages. He recovered and joined the Navy in November, 1917. In February, 1918, he was accidentally kicked on the head by another sailor and was unconscious for fifteen minutes, after which he recovered. It was found that he had a small depressed fracture over the right posterior frontal region. This depressed fracture was probably one of the tibial grafts which had become loosened by the kick. Ten days later, he developed generalized convulsions, as many as three in one week. During one of these attacks he threatened to kill the orderly with a razor. He was discharged from the Navy because of traumatic epilepsy. The skull wound discharged for about a year and then healed. During the year 1918 he had two more generalized convulsions. In July, 1919, he was again struck on the head by a small tree and was unconscious for several hours, after which he became acutely maniacal for two hours, during which time he attempted to kill one of the men with him. It required five men to put him in restraint. He recovered within an hour and remained in the hospital for two weeks when he returned to work. In July, 1920, he developed left foot drop and some frontal headaches. These continued until October when the foot drop improved, but the headache became more severe and memory was impaired. He was operated on Jan. 27, 1921, by Dr. J. A. Caldwell, United States Public Health Surgeon, at Cincinnati. The patient stated that the doctor had found depressed bone, removed it and filled the gap with a piece of rib. He has been perfectly well since that time, except for slight impairment of memory and the scars on his head. The neurologic examination on March 12, 1921, was negative except that the knee jerks were still sluggish, especially the left one.

LITERATURE ON THE SUBJECT

A true psychosis is not infrequent with cerebral lesions, especially when they involve the frontal region. Any form of mental picture may develop secondary to trauma. Under the head of post-traumatic constitution, Adolph Meyer¹ mentions paranoid states somewhat similar

1. Meyer, Adolph: *Am. J. Insan.*, January, 1904.

to the psychosis presented by our patient. The tendency to homicidal impulses in this case, on three separate occasions, secondary to trauma, is of rather infrequent occurrence.

Both Sachs² and Spiller,³ in their articles on cerebral telangiectasis, give a fairly complete review of the literature. In the majority of instances, patients presented evidences of telangiectasis on the face, head or somewhere on the body. In the discussion of Sachs' paper cases were reported by Starr, Angell, Gordon and Hunt.⁴ Steiner⁵ reported the cases of three families with hereditary hemorrhagic telangiectasis and reviewed twenty-five cases from the literature. The outstanding features were telangiectasis and repeated epistaxis. In none of his cases, however, was any mention made of cerebral involvement. Our two patients had no superficial telangiectasis themselves and knew of none in relatives.

Cerebral angiomas have been classed as congenital tumors, new growths or secondary to trauma. When associated with telangiectasis elsewhere, the congenital factor seems an important one. In reviewing the cases in the literature, trauma is of frequent occurrence. According to Sachs, in certain cases it seems highly probable that a trauma has either irritated a preexisting growth making it develop rapidly, or has actually been the cause of the new growth. Local inflammatory reaction, such as adhesions, connective tissue overgrowth or roughened inner layer of the dura over the dilated vessels, are further evidences of this. The history in my second case and the operative findings lead one to believe that trauma probably was an etiologic factor.

Surgery is the only treatment for these patients. Ligation of the dilated blood vessels has given surprising results in some instances. Sachs suggests that the blood vessels be ligated in such a way that no thrombosis forms, but that they collapse after ligation, thus preventing irritation of the cortex. Spiller calls attention to the danger of extensive ligation and reports two cases. In one case hemorrhage occurred into the brain and in the other hemiplegia resulted.

In our first case, the four main trunks were ligated without any serious after-effects except the series of convulsions within twelve hours after operation, in which the patient almost collapsed.⁶

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2. Sachs, E.: *Am. J. Med. Sc.*, October, 1915.

3. Spiller, W. G.: *Congenital Tumor of the Brain (Telangiectasis) and Associated Cerebral Movements*, *Arch. Neurol. & Psychiat.* **2**:50 (July) 1919.

4. *Nerv. & Ment. Dis.*, 1915, p. 634.

5. Steiner, W. R.: *Telangiectasia*, *Arch. Int. Med.* **19**:194 (Feb.) 1917.

6. In addition to the references given, the following may be of interest: Cassiver, H.: *Neurol. Centralbl.*, 1902, p. 32; 1910, p. 456.

Deetz, E.: *Virchows Arch. f. path. Anat.* **168**:341.

Kalischer, S.: *Berl. klin. Wchnschr.*, 1897, p. 1059.