VENOUS ANGIOMA OF THE CEREBRUM.

REPORT OF A CASE WITH NECROPSY.

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INTRODUCTION.

ANGIOMATA are among the rarest of brain tumours. The majority of the cases recorded are examples of plexiform angiomas occurring in connexion with the cerebral arteries. Thus, D'Arcy Power¹ in 1888 reported a case of arterial angioma involving the angular gyrus, and Drysdale,² in 1904, recorded two cases of similar angiomas connected with the anterior and middle cerebral arteries; further cases have been published by Kalisher, Emanuel, Bergmann-Oppenheim³ and Simmonds.⁴ A few cases of cavernous angiomas have also been described by Bruns, Oliver, Stupple and Bielschowsky. La Villette collected 18 cases of both types occurring in various situations. These angiomas are most probably of congenital origin and are of slow growth; usually they produce only general symptoms and in some cases epilepsy had occurred. Cerebral angiomas consisting entirely of veins are apparently even rarer than either form of arterial angioma. In 1922, one of us (C. W.-D.) with Sir Charles Ballance⁵ recorded a case of extensive venous angioma of the cerebral cortex associated with mild hemiplegia and epilepsy of Jacksonian type. The nature of the cerebral lesion was revealed at operation, from which the patient made a good recovery (1921). Five years after operation (1926) the patient was admitted to the Albert Dock Hospital and there died as the result of hæmorrhage from the angioma. Owing to the kindness of Dr. W. Bonner-Morgan in notifying us, we were enabled to examine the brain post-mortem.

CLINICAL DESCRIPTION OF CASE.

A. J. A., age 31, was first seen by one of us (C. W.-D.) in August, 1920. He complained of "fits" and slight weakness of the left arm and left leg.

History of Illness: During 1916, while serving in the Navy, he began to experience occasional attacks of "tingling" on the left side of the face. After four months each attack was followed by transient loss of consciousness. This in turn shortly afterwards was followed by temporary weakness in the left arm and leg. The attacks recurred at intervals of about four weeks until, in March, 1917, he was invalided from the Navy as a case of ? disseminated sclerosis. The attacks continued to occur at irregular intervals up to the time of his coming under observation in 1920.

There was no history of previous illnesses or trauma and no family history of epilepsy or other nervous disorder.

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Condition on examination (August, 1920).—(1) *Symptoms*: The “fits” occur as intervals varying from a few weeks to a few days and each attack is usually preceded by an aura of nausea, vertigo, and tingling on the left side of the tongue and face; this is followed by loss of consciousness. Occasionally he realises that his left leg is twitching before he loses consciousness. The actual fit lasts only a few minutes, but on regaining consciousness he finds that the left arm and leg are powerless and numb and that he has difficulty in speaking; this state continues for about three hours following the fit and then passes off. During the attack his tongue has rarely been bitten and incontinence has not occurred. Apart from the attacks, he complains of some degree of weakness in the left arm and leg and of occasional frontal headache. There is no vomiting or failure of vision.

(2) *Physical Signs*: Pupils equal and react normally to light and accommodation; optic discs normal; no nystagmus; ocular muscles normal; all other cranial nerves, including face, tongue and ears, normal. No abnormality of sensation detected. All movements present in left arm, but left hand-grip distinctly weaker than right. No wasting, no incoordination of arms and no intention tremor. All left arm jerks brisker than those of the right. Abdominal reflexes fairly brisk, but right greater than left. All voluntary movements of left leg are performed, but there is slight spasticity. Knee and ankle-jerks brisk, left greater than right. No ankle clonus; plantar reflexes, right flexor, left indefinite (flexor response invariably on stimulation towards inner side, and occasionally extensor on proceeding towards outer side of sole).

Heart normal; pulse 76; blood-pressure 130 mm. systolic; sphincters normal; urine, no albumen, sugar or casts.

Diagnosis: A diagnosis of “Lesion (? nature) of Rolandic area of right cerebral cortex with Jacksonian epilepsy” was made.

The blood yielded a negative Wassermann reaction; the cerebrospinal fluid showed no lymphocytosis, and both globulin and Wassermann reactions were negative. X-ray examination showed no abnormality of the skull.

Subsequent Progress of Case: From August, 1920, the patient was taking 15 grains sodium bromide daily. Up till April, 1921 the attacks averaged only one or two per 14 days, several consisting simply of numbness and twitching of the left arm and leg, without loss of consciousness. Throughout this period there was no definite change in the physical signs. After April, 1921, however, the attacks increased in frequency and severity, averaging three or four a week, about one in three being accompanied by loss of consciousness. As the patient was deteriorating, operation was decided upon. This was performed by Sir Charles Ballance on November 19th, 1921.

Operations: Under intratracheal ether, the lower two-thirds of the right Rolandic area was exposed. The dura, which was tense, dark and thin, was opened, and on being reflected, the whole of the cortical area exposed was found to be occupied by enormous veins, the arachnoid being stretched over them; some of these veins were of the diameter almost of the little finger. The walls of the veins appeared very thin and at several points were adherent to the dura: during the reflection of the latter, rupture occurred at some of the sites of adhesion, the hemorrhage being controlled with gauze plugs (five in all). Owing to their very large size and to the fact that all disappeared beneath the margins of the bone bounding the opening, no attempt could be made at separation or removal of these veins; also, it was seen that the venous angioma extended far beyond the limits of the craniectomy opening in the skull. The patient stood the operation extremely well and was conscious within one-and-a-half hours of its termination.

A week later, a further operation was performed, the flap being turned down, the cortical surface cleared of blood clot and the gauze plugs removed, with the exception of a portion of one plug, which was firmly embedded in clot and young granulation tissue (Fig. 1). The veins were not thrombosed and appeared somewhat less prominent than

Fig. 1. Outer aspect of right hemisphere, showing the enormously dilated veins and thickened dura.

a week previously. A celluloid plate was cut to fit the opening in the bone and fixed in position with small tacks.

The patient made an excellent recovery from the effects of both operations, only complaining of occasional headache.

Later history of Case : For the first six months following operation, no definite epileptic seizures occurred; only on four occasions did the patient experience slight numbness of the left arm and hand, passing to the left side of the face. The physical signs remained practically as before, excepting that the left arm and leg appeared stronger. During the next eighteen months only four seizures with loss of consciousness occurred, the minor attacks of numbness and tingling down the left side being experienced once or twice in four weeks.

Following the above observations, the patient was lost sight of until May 9th, 1926, when he was admitted to the Albert Dock Hospital with a complaint of intense headache and fits. The note of his condition on admission states: "Pupils react normally, all left-sided reflexes greatly exaggerated as compared with right, with some spasticity and loss of muscular power. Heart, lungs and abdomen normal; pulse 80, respiration 22". The patient continued to complain of intense headache and finally developed coma of increasing depth, dying on May 19th, the tenth day of admission. During the 10 days in hospital the patient ran an intermittent temperature, varying from 97.5° F. in the morning to 100° F. at night, with a pulse-rate varying from 60 to 90.

NECROPSY.

Externally, a large operation scar on the right side of the head; a gap 3 in. by 2 in. in the right parietal region of the skull filled with a celluloid plate. On removal of this and framing the gap the dura was found much thickened and adherent. Under the celluloid plate many large and dilated veins were apparent. Over the venous tumour also the dura was much thickened and adherent, rendering separation and removal of the brain most difficult. The brain substance under the venous mass was much softened, the area of softening being most pronounced between the hemispheres. Death appeared to have resulted from subarachnoid hæmorrhage occurring from the angioma. The whole surface of the brain was hæmorrhagic and appeared unduly soft. Nothing of note was found elsewhere in the body.

PATHOLOGICAL DESCRIPTION OF CASE.

The specimen consisted of an adult brain, showing considerable general flattening of its convolutions, more especially those of the right occipital lobe. The nature, extent, and distribution of the lesion may best be described by reference to the two accompanying illustrations (Figs. 1 and 2). There was surface meningeal hæmorrhage, especially over the base, particularly in the interpeduncular space, inner surfaces of the temporal poles, and along both Sylvian fissures and lower aspects of the frontal lobes. Over the central two-fourths of the outer and upper part of the external surface of the right hemisphere there was a large mass of enormously dilated and tortuous surface veins mingled with thickened and adherent dura, in the upper part of which was embedded the remains of a gauze swab (Fig. 1). Some of the large veins were continued over the vertex, and, as shown in the vertical coronal section (Fig. 2), were numerous also on the inner aspect of the hemisphere, where the brain-tissue was extensively softened and destroyed. The largest of these veins (in their contracted post-mortem condition) were 7 or 8 mm. in diameter, and numerous smaller veins were also present in the sulci in the neighbourhood of the larger veins.

COMMENT.

Features in the case worthy of note are:—

- (1) The sensory aura beginning in the face.

- (2) The absence of any demonstrable change in cutaneous sensibility.
- (3) The absence of fundus changes.
- (4) The extremely large size of the veins forming the angioma.
- (5) The improvement following operation—presumably due to relief of local pressure.
- (6) Death as the result of hæmorrhage from the angioma.

By a curious coincidence, a somewhat similar case occurred in the same hospital in the same year (1921). This was a cerebral venous angioma consisting of vessels of much smaller calibre and occurring in a younger subject. The case was recorded by H. Campbell and C. A. Ballance.⁶ D. M. Greig⁷ also reported a case associated with hemiplegia, epilepsy and adenoma sebaceum in a youth, age 18. Fatal hæmorrhage occurred in a case of cavernous angioma of the left cerebellar peduncle described by Wergman.

As these venous angiomata are almost certainly congenital, it is remarkable that the increase in size to which all angiomata are liable was insufficient to produce symptoms in our case until the age of 27. In Campbell and Ballance's case, symptoms began at the age of 13 and in Greig's case, at 3 years. All three cases show the following features in common:—

- (1) First symptom, epilepsy, with relatively slow development of hemiplegia.
- (2) The presence of epileptic attacks of the Jacksonian type; preceded by a definite aura, and,
- (3) The absence of changes in the fundus oculi.

We desire to express our indebtedness to Dr. W. Bonner-Morgan and to the Staff of the Albert Dock Hospital for their courtesy in granting us every facility for the pathological examination of the case.

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Fig. 2. Vertical coronal section, showing distribution of the veins on inner aspect of right hemisphere.