Vascular Malformations of the Brain.

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Development of the cerebral vessels is a very complex and rapid process; and it is not surprising that anomalies and malformations commonly occur.

Before the introduction of angiography by Egas Moniz (1933) these anomalies could only be studied by anatomists and pathologists We are now able to demonstrate and often treat them by surgical operations.

Minor anomalies are not usually of clinical significance; and they are discovered accidentally, e.g. double basilar artery. Ameli (1952)

On the other hand major malformations are of great clinical interest. During the last ten years a great deal has been achieved in diagnosis and treatment of these conditions.

These malformations can be devided into three main groups:

1. Aneurysms.
2. Telengiectases.
3. Angiomata.

Hæmangioblastomata and the conditions described by Lindau, Von Hippel and others are not included, as they are true neoplasms associated with vascular malformation in other parts of the body.

ANEURYSMS.

Intracranial aneurysms are not rare. Fearnsides (1916) found them in 0.85 of his autopsies. In 10% of the cases they are multiple.

According to Forbus (1930) the aneurysms which usually occur at the arterial forks are due to deficient development of arterial walls at these sites. But Dandy (1944) belived they owed their origin to peristence of part of the embryological vessels. The primitive trigeminal artery can be given as an example. This is the first communicating vessel between the carotid and basilar system. As the posterior communicating artery is developed the trigeminal artery disappears.

This artery may persist in the adult as a large vessel; or only a part of it remain at its origin, and later give rise to a berry like aneurysm.

Carmichael (1950) believes that developmental defects and arterial degeneration together are responsible for formation of these aneurysms.

There is no doubt that in some cases of aneurysms in olderly people atherosclerosis plays a part.

Rarely an infected embolus, as in cases of subacute bacterial endocarditis, may cause a so-called mycotic aneurysm. Syphilis plays no part in formation of intracranial aneurysms. Fearnsides (1916) found no evidence of syphilis on 51 autopsies performed on these cases.

As regards the site of these aneurysms; more than 75% of them occur at the three following positions: Middle Cerebral artery, Anterior Cerebral and Anterior Communicating, and the terminal part of the internal carotid artery, (Meadows 1952)

The size of the aneurysms vary greatly. (Fig. 1 & Fig. 2).

Occasionally more than one member of the family are affected. I have seen the condition in father and son.

CLINICAL MANIFESTATIONS

Three different syndromes can be described:

1. Subarachnoid hemorrhage due to leakage or rupture of the aneurysm into the subarachnoid space.

2. Intracerebral hematoma; when the aneurysm is adherent to or embedded in the cerebral substance.

3. Compression Syndrome.

SUBARACHNOID HEMORRHAGE.

Aneurysms are the commonest cause of subarachnoid hemorrhage.

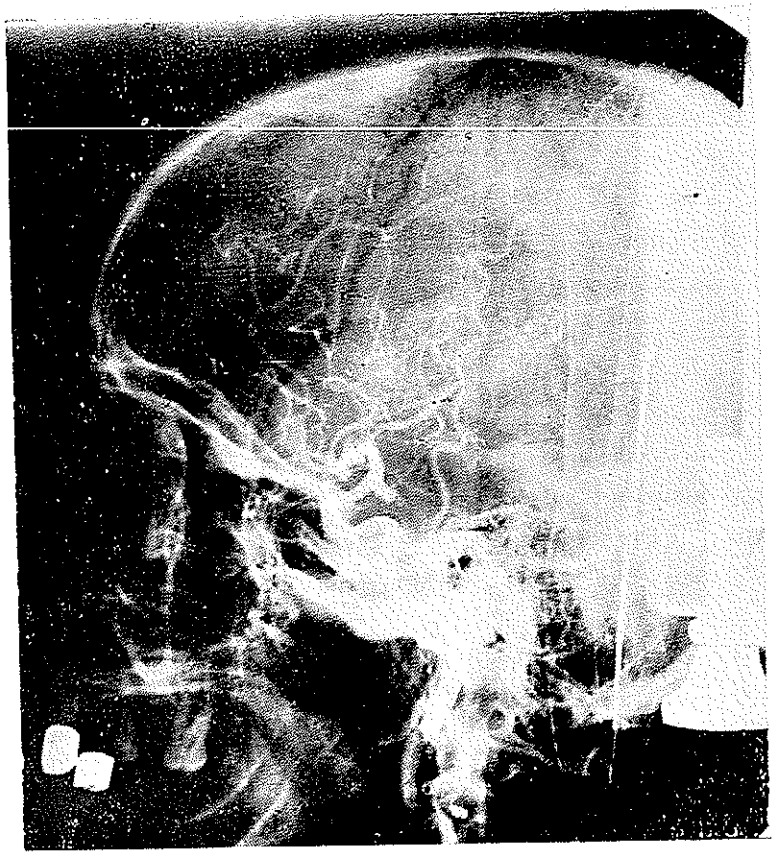

Fig. 1
A berry aneurysm arising from the terminal part of the internal carotid artery.

The condition is well known and the time at my disposal does not permit me to give a detailed account. A single case will be given to illustrate some typical features.

A married women aged 40 had had at least 3 attacks of sudden occipital headache followed by mental confusion and coma in the last 3 years.

On previous occasions she had regained consciousness after one or two hours; but this time she had remained semicomatous and delirious for five days.

On examination, that is on the fifth day, she had a temperature of 38° C with pulse of 80 per minute. She had severe neck rigidity and positive Kernig's sign. There was no paresis. Blood showed 12,000 W. B. C. with 75% polymorphs. Urine had albumin but no sugar.

Lumbar puncture gave a heavily blood stained C. S. F. The precipitate would not clott on standing and the supernatent fluid was xanthochromic. Carotid angiography on the right side showed a small berry aneurysm arising from the terminal part of the carotid artery. The right common carotid artery was ligated; followed 3 weeks later by ligature of the internal carotid artery.

Patient recovered and has remained free of symptoms for the last 18 months.

In hypertensive patients the condition may be mistaken for an ordinary vascular catastrophe.

The following case is an example. A lady of 55, a known hypertensive collapses in the street. Her friend puts the comatose patient in a taxi and takes her to her doctor. The blood pressure was 200/150; with no sign indicative of a hemiplegia. After a few days patient gradually regains consciousness; but complains of slight headache. 3 weeks after the first attack whilst walking in her house, she again falls down unconscious.

I was called to see this patient 4 days after the 2nd. attack. Then she was semicomatose, irrational and uncooperative when roused. She had a left hemiplegia. There was some neck rigidity present. Lumber puncture revealed a slightly blood stained C. S. F. Angiography on the right side demonstrated an aneurysm. (Fig. 1) The common and later

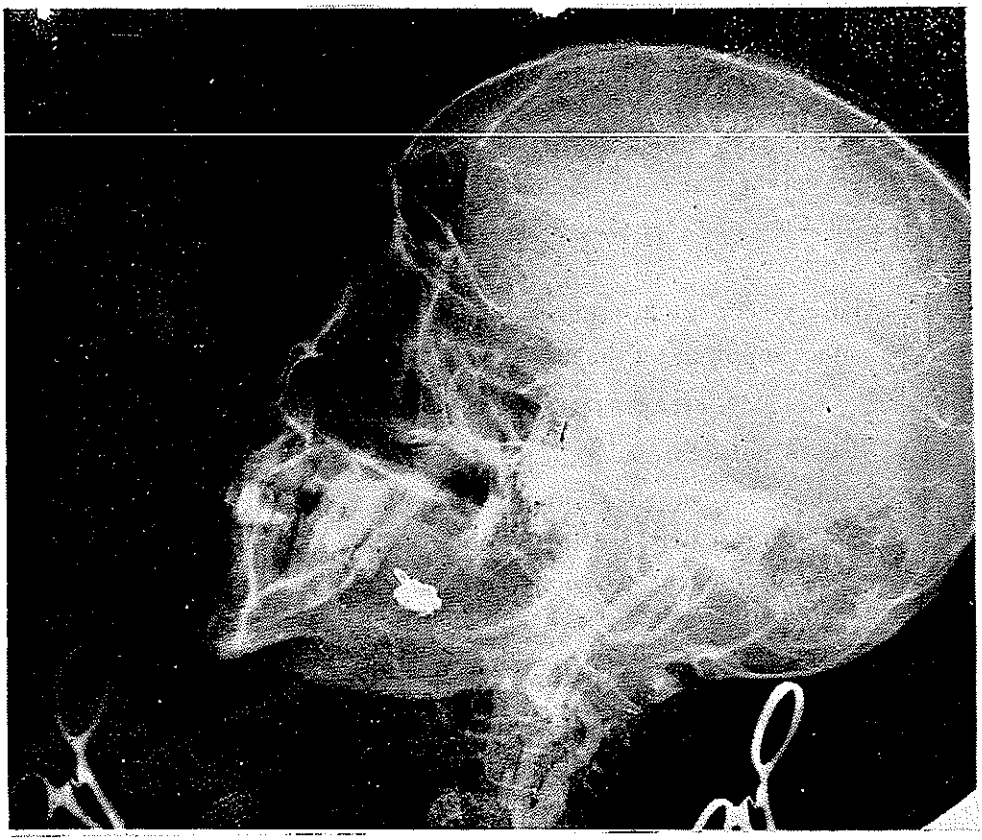

Fig. 2

A large aneurysm arising from the internal carotid artery; the first part of the middle cerebral artery is displaced slightly upswards.

the internal carotid arteries were ligated. Patient slowly recovered, and her hemiplegia also cleared up within 3 months of her operation, but she still complains of some burning sensation on the left side of the body.

PARALYTIC OR COMPRESSION SYNDROME

A number of cases have as their first clinical manifestation signs of pressure on one of the cranial nerves. The third, fifth and second cranial nerves are most often affected. The nerve affected depends on the site of the aneurysmal sac. The aneurysms arising from the anterior cerebral or anterior communicating artery are liable to compress the 2nd. nerve giving rise to changes in the visual fields and optic atrophy.

Paralysis of the 3rd. nerve is often associated with hypoaesthesia in the areas supplied by the 5th nerve.

INTRACEREBRAL HEMATOMA.

This is not a rare complication of an intracranial aneurysm, but is not widely recognised.

The clinical signs are that a rapidly growing intracranial mass. The signs would depend on the site of the aneurysm. (Fig. 2) shows an aneurysm which caused coma and hemiplegia in a 45 year old woman

TREATMENT.

Ligation of the common carotid artery, and later the internal carotid, is the main step in treatment of these aneurysms.

Direct attack on the aneurysm is becoming more popular. The manœuvres include; clipping the neck of the sack, application of a clip on the artery distal to the aneurysm; and application of pieces of muscle around or inside the sac.

TELENGIECTASES

These lesions, which are dilated vascular spaces, are rare, and most of these are incidental findings at autopsy. (Sugar 1951).

A condition which has been classified as telengiectases, and more wildely recognised is trigeminal encephaloangiomatosis, or Sturge-Weber Syndrome.

This consists of port wine stain naevus of the face, convulsions and abnormal calcification in the x-rays of the skull. The neavus and the calcification are on the same side. Calcification is supposed to be due to stasis in the dilated blood vessels. There are a large number of thin walled vessels in the cortex and meningeo. (Green 1945).

ANGIOMATA.

Two types are described; Arterio-Venous and Venous Angiomata. Arteriovenous Angiomata are much more common.

ARTERIOVENOUS ANGIOMATA.

Anatomically they are composed of three parts.

1. The feeding artery or arteries These are much larger than the cerebral vessels normally seen in the area.
2. A large mass of tangled vessels which form the major part of the angioma.
3. Large veins draining the blood into the venous sinuses. (Fig. 3).

There is no matrix, and therefore these are not true neoplasms. The veins contain arterial blood. The vessels are very thin and fragile. When a large angioma is surgically exposed, blood can actually be seen rushing in the vessels with each pulsation.

These malformations which very much vary in size, may occur in any part of the brain, but they are more often seen in the central area of the cortex.

Presence of such malformations has some influence on the size of the corotid artery, and scalp veins. Effects on the heart and ultimately on general circulation have been noted.

I have seen two children with large angiomata giving rise to hemiatrophy of the body on the opposite side.

There is no doubt that these malformations are congenital, and they often grow in size.

CLINICAL ASPECTS.

The condition manifests itself in at least three different ways.

1. Subarachnoid hemorrhage.
2. Intracerebral hemorrhage.

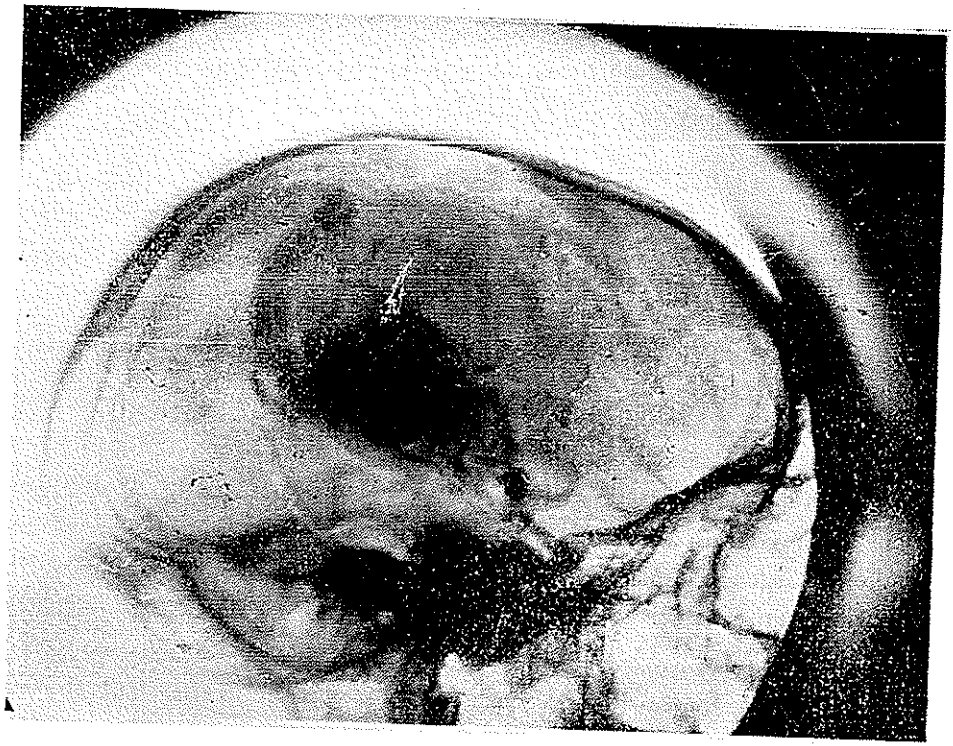

Fig. 3
A large arteriovenous angioma demonstrating the three distinctive parts: feeding arteries, mani angioma, and the large draining vein.

3. Jaksonian fits.

Three illustrative cases will be described.

CASE I.

A boy aged 12 whilst swimming hit his head against the side of the pool. He felt a little dazed and vomited. He was able to walk home and to bed. A few hours later he began complaining of severe headache and became drowsy. On admission to hospital he was semiconscious, and would not cooperate. His pulse was slow but his pupils were equal with normal reaction. Fundi were normal. He had a left hemiparesis affecting the face and the arm more than the leg

With a diagnosis of extradural hematoma Patient was taken to the operating theatre. A burr hole in the right temporal region was made. There was no extradural hematoma. A canula was inserted into the temporal lobe and 30 c.c. of dark blood was aspirated.

Patient's recovery was dramatic. An angiogram performed later showed a small angioma. (Fig. 4). This is obviously a case of bleeding into the brain substance.

CASE 2.

A boy of 14 was admitted to Pahlavi Hospital 4 hours after falling off his bicycle. He had not lost consciousness until an hour before admission. The loss of consciousness came on suddenly, following an acute headache.

On examination, he was deeply comatose with noisy breathing. There was head retraction with severe neck rigidity. Fundi did not show show any hemmorrhage. There was a right hemiplegia.

Lumbar puncture gave a heavily blood stained cerebrospinal fluid, under pressure. Two days later patient slowly regained consciousness, but he was totally asphasic. A carotid angiography was made, and an angioma around a mass, presumably a hematoma, was visualised in the posterior parietal region.

Patient's aphasia and hemiplegia gradually improved. Six months later patient was readmitted. He was still walking with a hemiplegic gait, and had a slight dysphasia of motor type.

Another angiography was performed. Now the angioma is moreclearly seen. This shows that the best time to remove an an-

Fig. 4
Small angioma which had caused an intracerebral hematoma after a slight injury.

gioma is soon after a hemorrhage, when the vessels are collapsed.

CASE 3.

A man of 22 was admitted with 6 years history of Jacksonian fits affecting the right side of the body, with increasing right hemiparesis. Recently the attacks had become more frequent and more severe. In some attacks the fits would become generalised with loss of consciousness. He had a slight dysphasia. Fundi were normal. There was a bruit audible on the surface of the skull on the left side He had a hemiparesis on the right side affecting face, arm and leg equally. There was a slight sensory disturbance of parietal type affecting the right half of the body.

A left carotid angiography was performed. This showed a large badly visualised angioma in the left parietal region. The angiography had to be repeated many times before we could obtain a decent picture. The reason was the very rapid circulation and the size of the angioma. (Fig. 5)

The patient was very anxious that something should be done for him. The condition and the operation hazards were explained to him. He was advised to return to Teheran six months later, and if he still wished to have the operation it would be carried out.

He was put on anticonvulsant therapy. On readmission he was much worse. His face was congested. His dysphasia had become more severe. His fits were not controlled, and often he had 10 to 12 fits daily. His hemiparesis had also become much worse. He insisted on having the operation.

Operation was performed under general anaesthesia with hypotension. I thank Dr Far for giving this anaesthesia which lasted for seven hours.

There was much bleeding from the scalp and diploic vessels. On opening the dura we were confronted with a huge vascular mass, pulsating and shaking like a mass of live worms. The skull flap was too small and more bone had to be removed. The vessels which we thought were arteries were clamped first Any bleeding had to be controlled with pressure, and pieces of muscle removed from the patient's thigh. Once we were able to take the feeding arteries, which were deeply situated, the whole angioma collapsed. We were then able

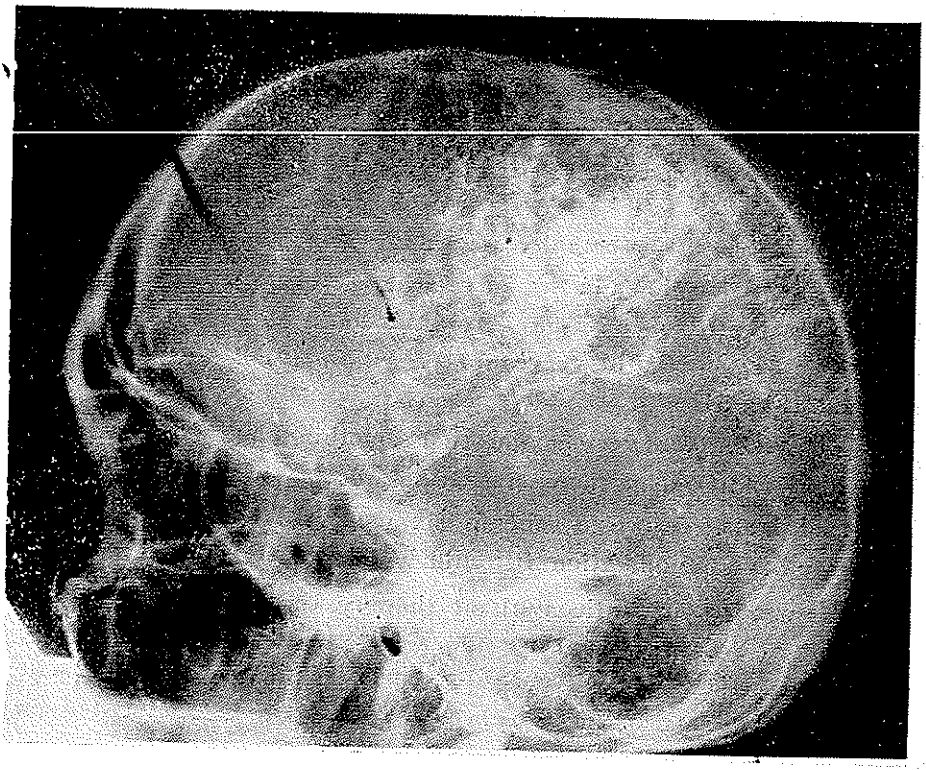

Fig. 5
A large arteriovenous angioma with at least three feeding arteries.

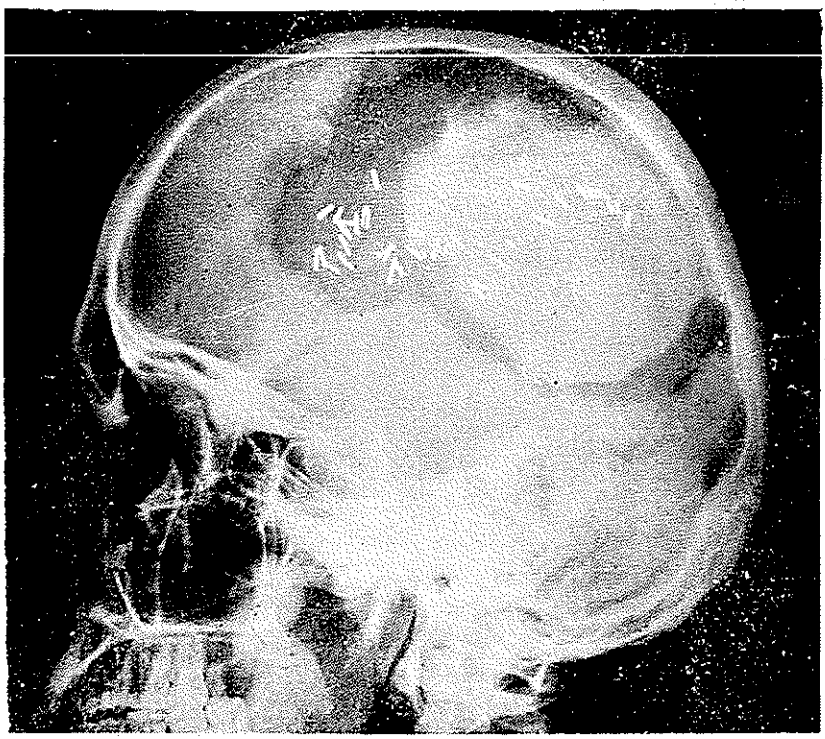

Fig. 6

Lateral view of the skull of the patient after the operation, demonstrating the extent of the angioma and the number of silver clips used.

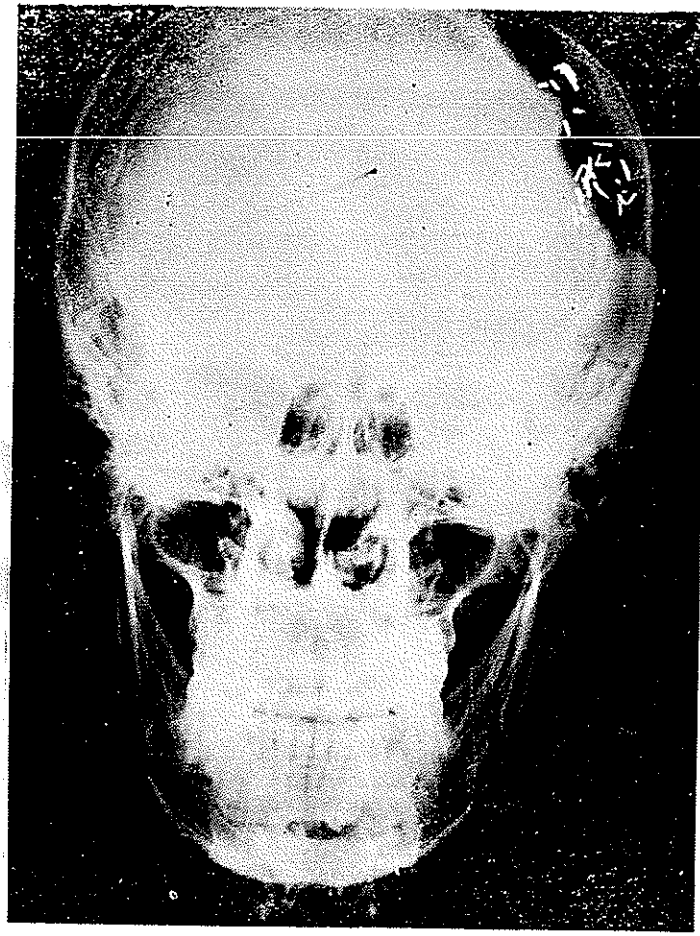

Fig. 7
Demonstrating the depth of the feeding arteries, although the angioma itself was superficial.

to remove the mass in too with the aid of endothermy. Patient received altogether eight pints of blood during and after the operation.

As Robert C. Bassett has said, (1951).

«It is a matter of record that there is no more formidable lesion dealt with by the neurological surgeon than the arterio–venous anomalies encountered in the brain.

Post operatively patient was completely aphasic and hemiplegic for two weeks. Then he began to improve. This improvement continued until his discharge from the hospital. At the time of his discharge his speech was better than it was before the operation. He looked normal and seemed to be more intelligent. The hemiparesis was no worse than it was before the operation. For 5 weeks that he was kept in hospital after the operation he had not had a single fit. This was in spite of no anticonvulsant therapy.

On his discharge he was advised to take one grain of luminal twice daily.

As without operation all these patients ultimately die from hemorrhage, I feel that it is advisable to operate the above patient had one of the largest angioma that I have seen, in the most specialised area of the cortex, neverthless the result justified the risk.

VENOUS ANGIOMA.

These are very rare. I have only seen one case demonstrated by angiography. This patient died 3 months later in another town, from hemorrhage. At autopsy they found no cause for hemorrhage. If the occipital region had been examined histologically, the angioma would have been demonstrated.

Résumé

L'auteur a cité quelques exemples des malformations vasculaires du cerveau (anevrismes, telangiéctasies, angiomes) et a montré qu'elles sont tout à fait curables par des techniques chirurgicales, même si elles étaient trop importantes.

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