Intracranial Arteriovenous Malformation

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I am grateful to Dr. Mitsuno, your President, for the invitation to be a guest speaker at this meeting of the Japanese Neurosurgical Society. I consider it an honor and have accepted the invitation with due humility. Both Mrs. Poppen and I have been able to renew our acquaintances with many of you. It has also given us the privilege of seeing your lovely country—an opportunity that both of us have anticipated with considerable enthusiasm.

I wish to present to you a review of the intracranial arteriovenous malformations that I have come in contact with in the past 30 years at the Lahey Clinic. This group of vascular malformations is of interest both because of the number and also because of the long-term survival of the earlier surgically treated patients. In addition, some of the patients were routinely used to give the surgeon specific and adequate information regarding the lesion that he was to come in contact with.

Intracranial arteriovenous malformations have been adequately described and classified by various authors in the last few decades. Norlen has reviewed the various classifications and suggests another of his own, based primarily on the anatomic and pathologic appearance of the lesions. Russell separates the blood vessel lesions into true tumors and angiomatous malformations similar to the classification made by Cushing and Bailey, who divided the cerebrovascular malformations into telangiectasia, venous angiomas and arteriovenous angiomas, and the true tumors into hemangioblastomas. Russell added cavernous angiomas to the cerebrovascular malformations. In a review of our series, we have met with a similar problem in that even among the members of our immediate neurosurgical and pathologic departments, different classifications have been used from time to time, such as, hemangiomas, arterial malformations, venous malformations, and the like.

The two major types of lesions that may arise from blood vessels, namely, the hemangioblastomas or the true tumors, and the hamartomatous...
formations of the blood vessels or cerebrovascular malformations, form two distinct entities as to symptomatology, prognosis and treatment. The vascular malformations are made up of blood vessels of adult structure, whereas angioblastomas are tumors with embryonic vascular changes and blood vessel forming cells. Therefore, the angiomatous lesions, which are composed of the adult vascular elements, can be divided into the sinus pericranii in which there is an outpouching of the dural sinuses or larger dural channels extending through the bone under the scalp. It becomes tense during coughing, sneezing, and straining. This is a rare lesion and in our experience we have encountered only two.

The meningeal angiomas may be angiomas of only the meninges, such as the meningeal varix and the Sturge-Weber syndrome. With the latter, there is an associated ipsilateral facial lesion made up of teleangiectatic vessels. The cortical lesion is usually located in the occipital area or it may be extensive and involve most of one hemisphere, with minute vessels overlying the small, narrow gyri with reactive calcification located in the deep layers of the cortex, readily seen on the ordinary x-ray films of the skull. In many of these patients there is an associated hemiatrophy of the skull on the side of the lesion. Angiomas in the same hemisphere are seen only on rare occasions. Clinically, the patients present themselves only because of seizures: homonymous hemianopsia is common, but the patient is rarely cognizant of this fact. A few of these patients are mentally retarded.

At the time of operation it may be difficult to determine whether the parenchymatous angiomas, such as the telangiectasia or minute capillary changes of the meninges, the parenchymal varix, the cavernous angiomas, or racemose angiomas are made up mostly of arteries or veins. Actually, it is also difficult to distinguish between the arteries and veins microscopically because of their maldevelopment. The cavernous or racemose angiomas are perhaps the more striking and ominous appearing lesions, particularly if they appear on the surface of the cortex and extend deep into the underlying tissues. I believe ominous is the proper word to describe the tumor when one considers the emotions that go on in the neurosurgeon's mind whose duty it is to remove the lesion. From the photographic standpoint, perhaps "striking" is the adjective that one could use in describing this lesion.

The angioblastic lesions which are made up of embryonic vascular producing tissue are composed of the angioblastic menigiomas. I am not referring to the vascular or bloody menigiomas, but to the angioblastic tissue that has some of the characteristics of menigiomas, as demonstrated by microscopic examination. Also, one encounters parenchymal angioblastomas, the so-called Lindau-von Hippel disease, which are found mainly in the cerebellar hemisphere. These are frequently associated with angiomas in
the retina on the same side as the cerebellar lesion. They may also be found in the pancreas, spleen, liver or kidneys. Erythrocytosis is reported to be frequently associated with these lesions; however, this has not been a factor in our group of patients.

From all practical standpoints, it would be advantageous to refer to the cerebrovascular malformations simply as large or small.

According to Olivecroma et al., arteriovenous malformations represent primitive vascular channels which tend to persist and develop, but in an abnormal fashion. Ordinarily, the majority of these primitive channels regress, with several vessels persisting in rather uniform courses and developing normally. This theory fits well with the different varieties, sizes and locations of vascular malformations, and certainly is adequate as a general explanation of the lesion. In our series of patients size and location appeared to be of more significance from the clinical and surgical points of view than the microscopic analysis.

Our series consists of 105 cases of arteriovenous malformations in which the diagnosis was proved by surgery, autopsy, or by angiography. We have discarded several cases in which a malformation was incidentally found associated with another lesion in the same hemisphere, such as a glioma or meningioma, on the grounds that neither the symptoms nor the course could be attributed to the finding of a malformation. Several more case histories were eliminated since the histories and findings were inadequately described.

Sixty-five patients were males and 40 were females. Approximately two-thirds of the group had the onset of symptoms before the age of 39, whereas only two had the onset of symptoms after the age of 60. The patient’s symptoms were most frequently initiated between the ages of 20 and 30.

Nine patients had bilateral lesions. Two patients represented combinations of multiple lesions occurring in the supratentorial as well as the infratentorial areas. There seemed to be no significant disparity as to the sites involved—either the right or left hemisphere. There was a vast preponderance of supratentorial lesions. Seventy-two of the malformations were classified as large and 33 small.

In the majority of patients we were able to satisfy ourselves as to the major vascular supply. In many patients it came from several of the major branches of the carotid or basilar arteries. The majority (56 cases), however, were primarily supplied by the middle cerebral artery and its branches. The major blood supply was from the anterior cerebral arteries in 38 patients, and from the posterior cerebral artery in 39. This was followed in frequency by the basilar artery and its branches, and malformations of the vein of Galen and the trigeminal artery. These seemed to little, if any, alteration
in the vascular supply associated with age, size or laterality. Neither size nor sex appeared to affect the location of the lesion in specific portions of the cortex. The cerebrovascular malformations did seem to have a preference for certain areas of the brain. The largest single group of malformations occurred in the parietal or central area of the cerebrum other than parasagittal. The next largest area of involvement was the frontal, followed by the occipital and parasagittal areas. Twenty-four occurred in the parietal or central area of the cerebrum, 18 in the frontal, 13 in the occipital, 11 in the parasagittal, 9 in the temporal lobe, 6 in the basal ganglia area, 5 in the Sylvian cleft, 4 in the corpus callosum, 2 in the ventricles, 2 in the hippocampal area.

Of the infratentorial lesions, five occurred within or around the brain stem, four within the cerebellar hemisphere, and one within the vermis.

Many of these vascular malformations are asymptomatic until bleeding or convulsions occur. A few are detected only as incidental findings during autopsy, death having occurred for other reasons.

Perhaps the single most common presenting symptom is headache, which occurred in 80 patients. However, if the patients with hemorrhage were excluded, only 40 had the presenting complaint of headache. In 19, the headache was of the migrainous type; the others had generalized headaches starting in a localized area or gradually developing into a generalized headache. Of the patients presenting themselves because of seizure, 15 had small and 35 had large lesions.

Subarachnoid or anterior cerebral hemorrhage occurred in 43 of the total 105 patients; 19 were in the group of 33 patients who had small lesions whereas 24 were in the group of 72 who had large lesions, indicating a greater proportion of hemorrhages in patients with the smaller malformations. Thirty-nine patients had motor deficits, usually as a result of intracerebral hemorrhage. Changes in vision also occurred usually in the patients who had an intracerebral hemorrhage or chronic choking of the optic discs. Change in vision usually was associated with the larger lesions, however, particularly if they were located in the occipital lobe.

Mental deficits occurred in 30 patients, 15 of whom had had intracerebral hemorrhage.

Size of the lesion appeared to play a definite part in the duration of symptoms. Of the 36 patients in whom the symptoms were present for less than one year, 18 had small and 18 large lesions. Of the eight patients in whom the duration of presenting symptoms was one to two years, two had small and six had large lesions. In the patients in whom the symptoms had been present between two and three years, 3 had small and 10 had large lesions. In patients with symptoms up to five years, 3 had small and
12 had large lesions. In those patients whose symptoms were of 10 years' duration, 5 had small and 13 had large lesions. In those with symptoms over 10 years, 3 had small and 12 had large lesions. Thus, it seemed that the longer the duration of the presenting symptoms, the greater the likelihood that the lesion was large. The occurrence of hemorrhage, however, did not seem to be related to the duration of the presenting symptoms.

X-ray films of the skull suggested the presence of an arterial malformation in 43 patients. Here, again, the larger rather than the smaller lesions tended to show positive signs. Ventriculography or pneumography was positive in 35 patients in demonstrating mass or localized atrophy, but not in determining the character of the lesion. Arteriograms were normal in only two of the patients in whom arteriography was performed, and both of these patients had small lesions. Electroencephalography had been carried out in only 32 patient: 15 were focally abnormal, 11 were normal, and 6 were diffusely abnormal.

**Treatment**

Six patients received no treatment other than anticonvulsive drugs. Three of these patients have had follow-up studies for more than three years, one over 10 years; one patient is dead as a result of a fatal hemorrhage, and another is symptom-free but has had follow-up observations for only one year.

Of the patients who were subjected to surgery, particularly the earlier patients who had not been subjected to arteriography but who were known to have a lesion, demonstrated by air studies or indicated by focal signs associated with increased intracranial pressure, only exploration and subtemporal decompression were carried out. Two of these patients died as a result of the operation or the condition. One patient has survived for many years, carrying out full activity. Of the patients who did not have associated increased intracranial pressure but who had exploratory operations only because of focal signs, seven were given x-ray treatment. Four improved and have returned to full activity, and two are dead; the condition of the seventh became worse temporarily as a result of operation, but after one year the patient returned to full activity.

Of those who received x-ray treatment alone, one patient improved and returned to full activity, and one is dead. Two have had follow-up studies for 5 to 10 years, and one has survived over 10 years. A few patients were subjected to only superficial coagulation of the vessels on the surface of the lesion. Of these, one patient improved and returned to full activity; one is well and symptom-free; one is dead, and one died as a result of the operation. Both living patients had a follow-up period of more than 10
years.

The remainder of the patients had what was thought to be complete excision of the malformation. Twenty-three improved and returned to full activity, but with a slight residual. Twelve were well and symptom-free; one was an invalid after surgery; one patient whose condition became worse for a time after surgery was later able to return to full activity. Six patients died as a result of the extirpation of the tumor. Of the patients who had total extirpation, 10 had follow-up studies for less than 1 year, 2 patients for 2 years, 8 for 3 years, 4 for 4 years, 9 for 5 years, 14 for 10 years and 3 for 15 years.

Of the four patients whose tumor was thought to be entirely removed but in whom subsequent angiograms showed incomplete removal, two died subsequently of fatal hemorrhage.

In some patients ligation of the internal carotid artery only was performed. Two of these improved and returned to full activity, but with some residual, however. One is completely well and symptom-free, and one is dead. Three have had a follow-up study of more than 10 years, and one considerably longer than 10 years.

Of the entire group of patients, 63 were considered to have had good results. There were 14 operative deaths, chiefly in the group with large lesions. Seventeen deaths occurred which apparently were not related to surgery. Four patients were invalids after the operation.

The procedures carried out on the above group of patients, therefore, were: (1) supportive treatment only; (2) exploration only; (3) exploration with subtemporal decompression; (4) exploration followed by ligation of the internal carotid artery; (5) ligation of the internal carotid artery only; (6) a combination of the above; (7) exploration with coagulation of the superficial portion of the malformation; (8) ligation of the major arterial trunks; (9) partial excision of the lesion; (10) complete excision, and (11) x-ray treatment only.

Good results were obtained in 37 of the 51 patients who had complete excision, in 11 of the 23 patients who had multiple procedures and in 4 of the 7 patient who had x-ray treatment. There is no question that, whenever the lesion can be excised without producing certain invalidism, extirpation is by far the better procedure. It is also true that patients under the age of 40 tolerated the operative procedures far better than those over that age, both as to mortality and morbidity. The morbidity and mortality rates vary also according to the location and the size of the lesion and, incidentally, on the experience of the surgeon who undertakes the operation.

I wish to present to you slides characteristic of certain of the lesions I have mentioned, and to discuss their operability and the final outcome.

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