pharyngioma in which the bilateral carotid siphons were occluded by the compres-
sion of the tumor, the anterior and the middle cerebral arteries were normally
visualized through the posterior cerebral arteries and the rami splenii in the
vertebral angiogram.

7) Angiographic examination of a case of 10 year-old boy which was
followed-up for a period of 5 years revealed that the collaterals from the rete
mirabile became more and more richer with a remarkable decrease in the juxta-
basal vascular networks. If the vascular networks are functioning as collaterals,
they should increase in vascularity as his age progresses and, therefore, the fact
that the vascular networks gradually decrease is very much in favor of its con-
genital origin.

The hypoplasia of the distal portion of the carotid siphon and the peculiar
juxta-basal vascular networks are probably due to some disorder in the vascular
development in the fetus stage. It is possible that the vascular networks have
developed as collaterals in the fetus stage to compensate poor cerebral blood
supply caused by the hypoplasia of the distal carotid siphon. This abnormal
vascular pattern may result in thrombosis or spasm leading to neurological deficits.
The vascular networks may not be demonstrated at that time and may later be
well demonstrated in the angiogram as symptoms and signs subside.

D-17. Idiopathic Occlusive Arterial Disorders in the
Cervical Region and the Main Trunks of the
Cerebral Arteries in Japan

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Since we have had no autopsy cases of such disorders in our clinic, we shall
discuss them mainly on the basis of clinical signs and cerebral angiographic
findings.

In our series of patients there were seven males and eleven females. The
age of onset of symptoms, was under 15 years in 15 cases (10 of them were under
11 years of age). The youngest was 4 years old and the oldest 25.

The dominant neurological symptoms were repeated transient focal signs,
in order of frequency: motor palsy, headache, episode of loss of consciousness,
numbness, speech disturbances, hemianopsia, etc. Of the 18 cases 14 had one
or more of these symptoms. In 16 cases, besides the above-described transient
symptoms, such deficits as mental retardation, involuntary movements, etc. were observed in various degrees. Slight EEG abnormalities were seen in most cases. Two adult patients had bloody CSF. Three had fever of unknown cause. A brother and sister in one family, two sisters in another had almost the same symptoms. One patient also had ichthyosis vulgaris. The past and family histories and other clinical findings were noncontributory.

The characteristic of angiographic findings were as follows:
1. Narrowing of the cervical portion of the internal carotid artery was observed in four cases. Occlusion or stenosis of the main trunk of the cerebral artery was found in all cases; bilateral lesions in the circle of Willis in 15 cases, and unilateral lesions in the circle of Willis and the contralateral origin of the middle cerebral and pericallosal arteries in the remaining three cases.
2. Two kinds of abnormal vascular network were found at the base of the brain; one in the basal ganglia, thalamus and subthalamic regions, and the other was a rete mirable in the narrow sense at the base of the brain, from extracranial to intracranial areas. In other regions, vascular anomalies were also found at the obstructive portion of the main trunk of the cerebral arteries, for example, the origin of the pericallosal artery; the brain surface; and inside the orbit, where communications are possible between the internal and external carotid arteries.

Among these abnormal vascular networks, lesions other than those at the base of the brain were apparently formed by collateral vessels. Therefore, we shall discuss the etiology of abnormal vascular networks at the base of the brain.

The first problem is whether or not arterial branches forming such a lesion exist at the base of normal brains. The main cerebral vessels which supply branches to the base of the brain are: internal carotid, posterior communicating, anterior choroidal, anterior, middle and posterior cerebral arteries. Some such perforating arteries were observed in normal cerebral angiograms, and they were rather clearly demonstrated in a case of occlusion of the internal carotid artery at its siphon. However, the distribution of the perforating branches studied angiographically by Kaplan and Ford in postmortem brains resembled remarkably the abnormal vascular network at the base of the brain. But this finding does not solve the problem. In most cases, occlusion or stenosis was located not distal to but proximal to or at the branching portion of the perforating branch. Moreover, there were some cases in which communications were present between the anomalous vessels at the base of the brains and the distal portion of the anterior and middle cerebral arteries despite the presence of scanty collateral vessels because of occlusion of the anterior and middle cerebral arteries. If these cases can be explained by the presence of collateral vessels, we have to examine whether or not there is an anastomosing branch between them.

In other author's reports on anastomoses, including autopsy reports, the following communications have been described: between the posterior pericallosal branch of the posterior cerebral artery and the pericallosal, lenticulostriate, and
anterior choroidal branches of the anterior cerebral artery and other perforating branches; between Heubner’s artery and the lenticulostriate, anterior and middle cerebral arteries; between anastomosing branches of the anterior and middle cerebral arteries; between the posterior communicating artery and the perforating branch of the internal carotid artery; between the perforating branches of the anterior choroidal artery; between the perforating branches of the middle cerebral artery; between the perforating branches extending to the subthalamic region which is supplied by the internal carotid and the main trunk of the other cerebral arteries; furthermore, among the communications between the perforating branches and the vessel on the brain surface, anastomoses have been reported between Heubner’s artery and the cortical arteries in the fronto-orbital region.

Granted these abundant anastomoses are present in brains at autopsy, we must still consider their time of origin. Even in cases of occlusion of both internal carotid arteries at its siphon, vascular abnormalities at the base of the brain were rarely seen. There was not always a correlation between the occlusive portion of the main trunk of the cerebral artery and the extent and the degree of its development. Therefore, it is assumed that such an abnormal vascularity can be formed only when the above-described abnormal anastomoses between the perforating branches are well-developed.

In conclusion, we consider at the present time that abnormal vascular networks at the base of the brain have the following features.

1. The usually clear demonstration in the arterial phase of the angiogram indicates that they are composed mainly of arteries and arterioles, though in some cases they resemble a capillary network. They were not seen in combination with angiomas of the face or other malformations; nor were they ever neoplastic or associated with arteriovenous fistulas. The main characteristic was their combination with occlusion or stenosis of the main trunk of cerebral artery.

2. In a poorly developed cases, disorders can be explained by occlusion of the main trunk of a cerebral artery, but well-developed cases cannot be explained only by such an occlusion, unless the abovementioned problem is solved.

3. This type of vascular abnormality was seen most often in children, and in our clinic we treated 2 pairs of young siblings. The abnormal vascularity was more advanced in children than in adults. Taking these facts into consideration, we presume that vascular anomalies at the base of the skull may be formed by extensive anastomoses of perforating branches which normally dilate or regress, but continue to exist, in the period from the gestational stage to the occurrence of symptoms due to stenosing disorders of the main cerebral arteries in both sides. Therefore we would like to call this vascular abnormality “cerebral arterial rete” in combination with the occlusive portion of one of the main cerebral arteries.