Bilateral carotid vertebrobasilar rete mirabile

Case report

RONALD R. JONES, M.D., AND NICOLAS WETZEL, M.D.
Department of Surgery, Division of Neurological Surgery, Northwestern University Medical School, and Veterans Administration Research Hospital, Chicago, Illinois

A case of bilateral internal carotid artery hypoplasia with bilateral carotid rete mirabile is described in which the intracranial circulation was fed predominantly by external carotid anastomosis, the vertebrobasilar system, and the internal carotid system.

Key Words: rete mirabile, carotid artery hypoplasia, cerebrovascular circulation

One of the well-known causes of seizure in the adult, cerebral ischemia, is presumed to be the etiological agent in this patient. The pathological changes in this case are uncommon, however, and deserve reporting.

Case Report

This 43-year-old right-handed man was in excellent health until 1964 when he began having episodes of losing consciousness; there was no aura, tonic-clonic movements, tongue biting, or incontinence. The episodes lasted 10 to 15 minutes and were followed by confusion. From 1964 to 1967 the patient experienced seven such episodes. In August, 1967, following a seizure, he noted that his left hand and arm "felt funny." For this reason, the patient was hospitalized at the Veterans Administration Research Hospital, Chicago, for evaluation. There was no history of headaches, trauma, or family history of seizures. The review of systems was unremarkable except for a moderate alcoholic intake.

First Examination. Upon admission in August, 1967, the general physical examination was normal. The blood pressure was 130/84. Bilateral carotid bruits were present, more marked on the right. His mental status was normal. The cranial nerves, including visual fields, were normal. The left arm was mildly paretic and hypalgesic to touch and pinprick. The deep tendon reflexes were brisk bilaterally, and no pathological reflexes were present. No cerebellar abnormality was noted. Laboratory data including complete blood count, urinalysis, serum electrolytes, fasting blood sugar, and liver function studies were within normal limits. The electrocardiogram was also normal. Chest and skull films were normal, as was the brain scan. A lumbar puncture revealed clear fluid with normal pressure and protein content of 75 mg%. An electroencephalogram (EEG) showed bitemporal spikes with moderately severe slow waves in both temporal areas (right greater than left). A retrograde right brachial cerebral angiogram was interpreted as showing an arteriovenous malformation involving the right pos-
terior parietal lobe, the right temporal lobe, and the brain stem. Surgery was deemed inadvisable, and the patient was placed on anticonvulsant therapy.

Second Examination. Three months later in November, 1967, the patient was readmitted due to persistent seizures with 30 episodes of grand mal type occurring within the previous 45 days. These seizures consisted of generalized tonic-clonic movements, tongue biting, and urinary incontinence which were not preceded by an aura. Examination, both general and neurological, was unchanged; the medication was adjusted, and the patient was discharged.

Third Examination. Due to persistent seizures, the patient was readmitted in June, 1968, for reevaluation. The general physical examination was unchanged. He still had a right carotid and right temporoparietal cranial bruit. The cranial nerves were normal. The sensory examination revealed a decreased perception of all modalities in the left arm and leg, with no weakness but brisk reflexes and increased tone. Repeat brain scan and skull films were normal. The EEG again showed a bilateral frontotemporal slow wave (right greater than left). Right and left carotid angiograms and retrograde left brachial cerebral angiograms revealed a bilateral carotid rete mirabile, as well as ophthalmic rete and marked communication between the intracranial and extracranial circulation. A pneumoencephalogram was performed and was interpreted as normal. The cerebrospinal fluid (CSF) protein again was 75 mg%.

The medication was readjusted, the seizures were controlled, and the patient was discharged. To date, the patient is seizure free.

Discussion

In 1664, Thomas Willis described and recognized the importance of a basal vascular collateral system which today bears his name. Besides this basal system, there are two other collateral arterial anastomatic networks. These are:

1. The leptomeningeal arteries, which connect the major supplying arteries with each other by an arteriolar network over the cerebral and cerebellar cortex.  

2. The rete mirabile or internal-external carotid arterial anastomosis.

The former were demonstrated anatomically by Huebner in 1874 and roentgenographically by Fay in 1925. The latter network was described in adults and children by Vander Eckeen and Adams in 1953 and represented communication between the peripheral perforating branches of the external carotid artery and surface branches of the internal carotid artery. This network has been stated to be within a venous lake whether or not the site is intra- or extra-cranial. All of these types of collateral circulation are important because they offer alternative routes of circulation when the intracranial blood supply is reduced through normal channels due to pathological states.

The functional significance of two varieties of anastomotic vessels, namely, the leptomeningeal and rete mirabile, depends on many factors. The gradual increase in demands of a vascular intracranial lesion may cause a marked collateral supply. On the other hand, negative demands produced by a gradual occlusion of one of the great vessels at the base of the brain may result in adequate collateral takeover particularly in patients with young elastic vessels, provided there is adequate time and proper hemodynamics.

In any discussion of cerebrovascular abnormalities, one must distinguish between developmental anomalies and occlusive vascular disease. In this patient there was nothing to suggest that the condition was secondary to occlusive disease. On the other hand, embryologists have stated that the rete mirabile does not appear during the development of the cranial circulation in man, even though it does exist normally in lower mammal species. Therefore, it is difficult to explain the etiology of the condition.

The dysgenesis of the internal carotid artery we have reported, was described in 1913 by Fisher. His cases were secondary to occlusion of the internal carotid artery with collateral circulation being supplied by the circle of Willis and the contralateral internal carotid artery. He stated in his report, however, that occasionally the external carotid artery communicated with the intracranial circulation via the foramen ovale, foramen rotundum, and/or by a branch from the
Bilateral intracranial rete mirabile

middle meningeal artery. The latter was demonstrated in our case (Fig. 1).

Embryologic patterns are basic to an understanding and evaluation of these anastomoses and anomalies. The internal carotid arteries are formed from the cephalic prolongation of the dorsal aortic roots and the third aortic arch. At the 14 mm stage of embryonic development, the portion of the dorsal aortic root between the third and fourth arch diminishes and later disappears leaving the third arch to contribute the curved proximal portion of the internal carotid artery. That portion of the ventral aortic root which, from the first, has fed the third aortic arch persists as the common carotid artery.8

Priman and Christie9 believe that the hypoplastic vessels, namely, the internal carotid arteries, represent a congenital variation in which the carotid arteries apparently developed normally at first and take part in the formation of the circle of Willis. At a later stage of fetal life, degeneration and shrinking results from some unknown cause.

In our patient, both internal carotid arteries were hypoplastic from the bifurcation to the region of the siphon (Fig. 2). The siphon was absent and taken over by the carotid rete from which the anterior cerebral arteries originate. This suggests that the dorsal aortic roots bilaterally either failed to reach normal size or had decreased in size.

Many patterns of collateral circulation between the external and internal carotid arteries, secondary to developmental anomalies and occlusive vascular disease, are known.1,4,9,12,14,16 These channels are potential ones, however, and become obvious only when obstruction interrupts the normal pathways. This has been shown in our case in which the internal carotid arteries were not sufficient to supply the cerebral circulation. Nevertheless, the external carotid arteries enlarge and through numerous routes evident in the basal, frontal, and occipital areas10 supply the vascularity needed to the brain (Figs. 2 and 3).

The collateral circulation present in our case is a normal condition in many animals, thus suggesting the phylogenetic basis for such communications in man. For example, the main contribution to the circle of Willis

Fig. 1. Angiogram, early arterial phase. Anteroposterior projection shows the carotid rete (single arrow) in area of the carotid siphon. Middle meningeal arteries (double arrows) are seen communicating with intracranial circulation.

Fig. 2. Angiogram, arterial phase. Lateral projection shows the hypoplastic internal carotid artery (single arrow) from the bifurcation to the siphon. Carotid rete (double arrows) from which anterior cerebral arteries originate is evident. Ophthalmic rete (double cross arrow) is also present. Note the numerous rete in the frontal and occipital areas.
in the dog and cat is from the external carotid artery via a network of vessels, namely, the rete mirabile in the orbit, which corresponds to the ophthalmic collateral system seen in Fig. 2. On the other hand, in cattle and elephants, the occipital branch of the external carotid artery is the major supplying vessel through the vertebrobasilar system and in this way corresponds to the human occipitovertebral collateral pathway seen in Fig. 3.18

In addition to collateral branches from the external carotid artery, perforating collaterals from the anterior and posterior choroidal arteries are demonstrated around the quadrigeminal plate (Fig. 4). It has been postu-
Bilateral intracranial rete mirabile

lated that these collaterals may be caused by an increase in local flow secondary to arterial, arteriolar, and capillary vasodilation resulting from cerebral anoxia. It is also noteworthy that both middle cerebral arteries do not fill from the carotid system but from the vertebrobasilar system through the circle of Willis (Fig. 5).

There was a large thalamic blush in this patient’s angiogram which had to be distinguished from a tumor stain. The arteries that contribute to the thalamic blush are characteristically the anterior choroidal, lenticulostriate, artery of Huebner, posterior choroidal, posterior colosal, and thalamogeniculate branches of the posterior cerebral artery. The features that permitted the differentiation of a “striate blush” from a tumor stain were:

1. The presence of “occlusive” cerebral vascular disease, namely, bilateral dysgenic internal carotid arteries;
2. The orderly and regular appearance of the arteries that cause the blush, whereas in the tumor stain the vessels tend to be disorganized and irregular;
3. The absence of abnormally filling veins or tumor veins; and
4. The absence of arteriovenous shunting.

Therefore, we concluded that the blush was secondary to collaterals.

There are two other common pathological states exhibiting a rete mirabile exit in man besides the entity described here. One is a falx meningioma in which branches of the middle meningeal artery supplying the upper falx anastomose with branches of the anterior cerebral artery[10] supplying the anterior inferior portion of the falx. The other is stenosis or thrombosis of the internal carotid artery, in which case the meningeal branches of the ophthalmic arteries anastomose with anterior cerebral artery branches. The latter is most likely the most common of all three entities.

In review of the literature, the Japanese have reported[5,7,13] the greatest abundance of cases of rete mirabile not caused by occlusive vascular disease. Reports have also occurred in the French literature.[11] To date, most reported cases of the abnormality, whether unilateral or bilateral (as our case), have been in those patients of Japanese ancestry. Our patient has no such ancestry and is of some interest for this reason.

The most common symptoms of this anomaly in the Japanese reports were transient hemiparesis, seizures, and subarachnoid hemorrhage (SAH). All were exhibited in this patient except for SAH. Most cases were under 21 years of age and there was an equal sex distribution. The etiology is still unknown except as a compensatory mechanism to maintain an adequate cerebral circulation.

Summary

We have presented a case of bilateral internal carotid artery hypoplasia with bilateral carotid rete mirabile in which the intracranial circulation was fed predominantly by external carotid anastomosis, the vertebrobasilar system, and the internal carotid system.

References


Received for publication December 15, 1969.

Support by National Institute of Neurological Diseases and Blindness, Research Fellowship Award NB 02198.

Address reprint requests to: Ronald R. Jones, M.D., Division of Neurological Surgery, The Children’s Memorial Hospital, 2300 Children’s Plaza, Chicago, Illinois 60614.