Abnormal Cerebrovascular Network Related to the Internal Carotid Arteries

Akira Nishimoto, M.D., and Shinji Takeuchi, M.D.
Department of Neurological Surgery, Okayama University Medical School, Okayama City, Japan

Japanese neurosurgeons have recently observed in patients of their own race a variety of neurological disorders which are often transient and appear most frequently in young people. The typical angiographic appearance is that of narrowing or occlusion of both internal carotid arteries at the level of the siphon (C-1) together with a hemangiomatosus network at the base of the brain (Fig. 1).

These patients were initially reported as having a type of occlusive disease of the internal carotid artery. The entity has also been reported by other Japanese authors as occlusion of the circle of Willis, telangiectasia, or vascular malformation. In 1964 we collected 21 case reports from the Japanese literature, added three of our own, and concluded that the entity was unique to the Japanese. We reported the entity as “hemangiomatosus malformation of bilateral internal carotid arteries at the base of the brain” and noted the similarity of the abnormal vasculature to the vascular network in the embryo. We noted, too, that the malformation was primarily limited to the distribution of the internal carotid arteries. The term “cerebral rete mirabile” has some descriptive value although it is probably not entirely appropriate. The Western literature yielded only one similar case with bilateral involvement, and this patient was a Japanese woman. Tumors and Wood reported comparable cases in non-Japanese patients, but the occlusions were unilateral.

Since our paper in 1964, numerous other reports have appeared in the Japanese literature. Adding these to an additional number collected by writing all the neurosurgical clinics in Japan, we are now reporting a total of 96 cases.

Analysis of the Cases

Sex and Age. Of the 96 patients, 41 were men and 55 women. The onset of symptoms occurred before 10 years of age in 56 patients and before 21 years in 73 (Table 1).

Symptoms. There was considerable difference in the initial symptoms in patients, which could be correlated with age. In those who were under 21 years of age, weakness of a limb or limbs was the most common manifestation, occurring in 51 of 73 patients (Table 2). The weakness occurred suddenly, quickly disappeared, but often recurred. Convulsions, visual disturbances, and nystagmus were relatively common in this group. In those patients over 20 years of age, subarachnoid hemorrhage was the most common initial manifestation, occurring in 12 of 23; motor weakness was the next most common sign in this group. Only two patients under 21 years of age had a subarachnoid hemorrhage, and none of the older group had visual disturbances or nystagmus.

Prognosis. The majority of the patients in both age groups followed remitting courses and had to be classified in one group as recovering, unchanged or recurring. Typically, a patient shows a sudden onset of hemiparesis, recovers quickly only to have the finding reappear on the same or contralateral side; 66 patients in the younger group followed this course. Only four became progressively worse; an additional three died. There were no deaths in the older group; all 23 recovered unchanged or had a recurrence (Table 3).

Electroencephalographic Findings. Background EEG activity showed normal-to-mild slow-wave pattern generally in resting record. Generalized slow waves were easily activated by hyperventilation in most of the cases; these slow waves did not readily return to the normal pattern after hyperventilation had been stopped. We believe that these EEG findings indicate cerebral circulatory insufficiency but have little value in the diagnosis of the disease.
Angiographic Findings. The hallmark of this entity is the angiographic appearance of narrowing of both internal carotid arteries with a distinctly pathological hemangiomatic vascular network at the base of the brain (Figs. 2–4). Bilateral carotid angiograms were done in 72 of the patients; 55 of them showed the angiographic appearance described as typical of the entity. Usually the narrowing or occlusion was at the C-1 level, but some had narrowing extending to C-4 with no visualization of the posterior communicating or ophthalmic arteries. In 15 patients, the typical angiographic findings were noted on one side only, with atypical findings on the opposite side in 10 and almost normal contralateral angiograms in 5 of these. Only unilateral carotid angiograms were done in 22 patients, who are included in the series because they showed the typical abnormalities described and were presumed to have bilateral lesions on the basis of their symptoms and signs.

Two patients had repeat angiograms. One,