FOUR CASES OF CAROTID-BASILAR ANASTOMOSIS ASSOCIATED WITH CENTRAL NERVOUS SYSTEM DYSFUNCTION

BY

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Various anomalous anastomoses between the carotid and basilar arteries have been reported. Krayenbühl and Yasargil (1957) classify three types. The first and by far the most frequent type is a persistent primitive trigeminal artery. The second in frequency is a persistent hypoglossal artery. The third type is a persistent primitive acoustic artery. We have found only the first type in our angiographic review.

Persistence of the primitive trigeminal artery was first reported by Quain (1844). The first report of angiographic demonstration was not until 1950 (Sutton). In a review of the literature in 1960 (Jackson and Garza-Mercado), only 33 case reports were found. Krayenbühl and Yasargil in their book on angiography describe four cases demonstrated by angiography. Their first case may be an anomaly other than a persistent trigeminal artery, since the vessel appears to originate at the junction of the C1 to C2 portion of the carotid artery. The vessel illustrated in their last case, described in a discussion of an arteriovenous malformation, may be a branch of the external carotid artery. Lindgren (1954) also reproduced angiograms of two cases.

The incidence of this lesion reported in the literature has varied from 1% to 0.001% (Jackson and Garza-Mercado, 1960). As pointed out by Harrison and Luttrell (1953), the true incidence of this lesion is very difficult or impossible to ascertain. We found an incidence of approximately 0.1% at the Indiana University Medical Center, and in a series of 76 angiograms at the New Castle State Hospital, three cases were demonstrated for an incidence of 4%.

Case Reports

Case 1.—A 7-year-old white girl was admitted to the Indiana University Medical Center Riley Hospital on July 5, 1950, because of headaches and unusual behaviour. One week before admission, she suffered progressively severe frontal headaches and epigastric distress. Two days before admission, she became agitated and had intermittent episodes of delirium in which she expressed fear of having her head cut off. She had no fever and no recent upper respiratory symptoms. The gestation period, birth, and neonatal period were uneventful. She weighed 7 lb. at birth. She walked at 9 months and said some words at 18 months. She had a speech impediment, and she was also clumsy and stumbled frequently. She had considerable difficulty in learning at school. For a year before admission, she had had episodes at night during which she would cry and talk to herself. At these times she could be awakened only with difficulty.

On examination, the patient was cooperative, but she was not orientated as to place and time. Her head and eyes were deviated to the left most of the time, but otherwise the general physical and neurological examinations showed no abnormalities. The optic discs appeared normal. During her stay in hospital, she deteriorated progressively. She developed a mild left hemiparesis and left homonymous hemianopia. Three lumbar punctures revealed a normal pressure, and spinal fluid examination was entirely normal, with a protein content of 15 mg./100 ml. The colloidal gold curve was 1111100000.
Blood chemistry was normal. Serial electroencephalograms showed high-amplitude, arhythmic slow waves in all leads, maximal in the right fronto-temporal region with evidence of progression. Pneumoencephalograms were normal. Cerebral angiography was normal except for the demonstration of a persistent trigeminal artery on the left (Fig. 1). The patient's parents signed her out of the hospital on August 1, 1959.

Case 2.—A 38-year-old white woman was admitted to New Castle State Hospital in 1951 because of uncontrolled right Jacksonian motor seizures and severe psychotic depression. A history of traumatic birth with subsequent right hemiparesis was obtained from the commitment papers. She did not walk until she was more than 3 years of age. During the first few years of her admission, she would refuse to do anything and sit by herself hour after hour complaining of 'radio voices' cursing her and talking about her. On Rauwolfia and phenothiazine drugs, she adjusted quite satisfactorily to the hospital routine and became a model patient.

On examination, she had a spastic, hyper-reflexic, right hemiparesis with a right-sided Babinski sign. The right extremities and right side of the face were hypoplastic. Position sense was decreased in the right fingers. With the patient sitting, ophthalmodynamometric pressures were 40/110 in both eyes. Psychological testing revealed her intellectual function to be between borderline and mild mental deficiency. There was also evidence of specific organic impairment of the left hemisphere and of affective disturbances.

Repeated electroencephalograms revealed rhythmic 4 to 6 c/s slowing and spikes from the left Sylvian leads.

Bilateral carotid angiography on June 24, 1959, showed a persistent trigeminal artery on the right (Fig. 2). Multiple small angiomas were present in the left middle cerebral artery distribution. Cerebral blood flow by the Kety-Schmidt nitrous oxide technique was measured twice on July 6, 1960. The first test indicated a flow of 40 ml. of blood per 100 g. of brain per minute, and the second 36 ml. of blood per 100 g. of brain per minute.

Case 3.—A 16-year-old white girl was admitted to New Castle State Hospital on September 30, 1958, because of generalized seizures, lapses of consciousness associated with bizarre behaviour, and violent combative behaviour between seizures. When medicated, she had seizures only at night. Seizures began at 30 months of age, following measles and presumed measles encephalitis. They were chiefly psychomotor seizures, but occasionally she had generalized convulsions. She described a brief aura of pain over the forehead and a sensation of spinning inside her head. During these times she was assaultive and destructive and it was a problem to separate the seizure activity from her usual personality. Because she frequently attacked other patients and attendants, she had to be placed in the hospital security ward.

On examination, she was an extremely large, heavy, white girl who was otherwise normal physically and neurologically. Her Wechsler-Bellevue intelligence quotient was 70. Some of the decrease was thought to be the effect of anticonvulsant drugs. Brachial blood pressures with the patient sitting were 110/70 mm. Hg in both arms. Ophthalmodynamometry, with the patient sitting, revealed retinal artery pressures to be 10/65 in the right eye and 30/80 in the left eye. The retinal artery pressures in the supine position in the right eye was 20/72 and 42/75 in the left eye.

Serial electroencephalograms showed generalized rhythmic slowing and multiple spikes, the maximal abnormality being in the right hemisphere. Bilateral carotid angiography was performed on August 2, 1960, and was normal except for a persistent trigeminal artery on the right (Fig. 3).

Case 4.—A 46-year-old white man was first seen on April 12, 1960, at New Castle State Hospital. He had been committed because of mental deficiency from birth, left-sided Jacksonian convulsions for four months, and progressive left hemiparesis for four months. He had been unable to learn at school, but he had made a marginal adjustment working at simple repetitive tasks and living under the supervision of a brother. He could not read or write but could print his name by rote. Four months before admission, he had his first Jacksonian seizure, beginning in the left arm, and afterwards a left hemiparesis. He gradually deteriorated so that two weeks before admission he was unable to walk, could not eat solid foods, experienced considerable nausea and vomiting, and complained of severe frontal and occipital headaches.

A pilonidal cyst was excised in 1944. His mother died at the age of 44 from an unknown cause. She had been confined to a mental hospital because of psychotic behaviour. Both of his parents had diabetes mellitus. Two brothers and one sister were living and well.
Physical examination revealed an acutely ill white male, who was obtunded and somnolent. His speech was slurred. He could not walk alone, but when supported he had a left hemiplegic gait. He had a left homonymous hemianopia and bilateral papilloedema. Touch perception was reduced on the left side of the face, and he had a left supranuclear facial paresis. Simultaneous auditory stimulation was ignored, or suppressed, on the left side. The tongue protruded to the left. He had a spastic left hemiplegia, complete in the arm, but with slight function in the leg. His blood pressure was 130/85 mm Hg and the pulse was 100. On the left side he had hypaesthesia and hypalgesia, suppression of bilateral touch stimuli, absence of position sense, astereognosis and atopognosis. Muscle stretch reflexes were exaggerated on the left side. Abdominal reflexes were absent on the left. The Babinski, Chaddock, and Conda signs were present on the left. The clinical impression was of an infiltrating glioblastoma multiforme of the right hemisphere, with maximal involvement in the posterior
Sylvian region. The patient was transferred to a general hospital. There, bilateral carotid angiograms indicated a massive lesion in the parietotemporal region and a persistent trigeminal artery on the right (Fig. 4).

A craniotomy performed April 14, 1960, revealed an infiltrating glioblastoma multiforme in the right posterior Sylvian region. For internal decompression, the posterior part of the parietal lobe and occipital lobe were removed. On November 19, 1960, when last examined, the patient was essentially unchanged neurologically.

Discussion

The embryogenesis of the trigeminal artery was described by Padget (1948). The anatomical relationships with the third, fourth, fifth, and sixth cranial nerves and the cavernous sinus have been well diagrammed by Harrison and Luttrell (1953). Kepes and Kernohan (1958) found that in about half the cases the artery perforated the dorsum sellae, and in the other half coursed lateral to the dorsum sellae to join the basilar artery. No case has been reported in which this condition existed bilaterally. There is no significant difference in the occurrence on the two sides or between the sexes. This anomaly is frequently associated with unusually small vertebral arteries and a small proximal basilar artery to the level of the shunt. Saltzman (1959) reported eight cases and made an angiographic classification into two types. In the first type the entire basilar artery system distal to the anastomosis was filled through the shunt. This was accompanied by poor filling of the posterior communicating artery. In the second type the shunt angiographically supplied mainly the superior cerebellar arteries, while the posterior cerebral artery received its supply through the posterior communicating artery. The distal end of the basilar artery was poorly visualized in this group. He classified one case as a combination of the two types. Aneurysms not directly related to the shunt are also probably more frequent (Schaerer, 1955; Murtagh, Stauffer, and Harley, 1955).

Various clinical syndromes have been suggested as related to a persistent trigeminal artery. A persistent weakness at the site of the carotid origin of the embryonal trigeminal artery has been suggested as a cause of intracavernous carotid aneurysms and carotid-cavernous fistulas (Sugar, 1951), although from necropsied cases this is probably not the most frequent site (Dandy and Follis, 1941). Only one patient, a 33-year-old woman, had a history of trigeminal neuralgia (Jackson and Garza-Mercado, 1960), but she had evidence of disseminated nervous system involvement, suggesting that the persistent trigeminal artery was not necessarily causally related to the trigeminal neuralgia. Subarachnoid haemorrhage has been purported to arise from this vessel (Kloss, 1953; Saltzman, 1959). Case 2 of Harrison and Luttrell (1953) had a massive intracerebellar haematoma at necropsy, but no mention was made of identifying the bleeding site. To our knowledge, bleeding from a persistent trigeminal artery has not been demonstrated at necropsy.

From the reported cases, it seems apparent that no characteristic clinical syndrome is associated with this condition. However, from our material, it does seem likely that the frequency of this anomaly is greater in patients with retarded mental development dating from infancy. All four of our patients were intellectually deficient, three of the patients being confined to a chronic neurological hospital.

In the New Castle State Hospital, a chronic neurological institution, the incidence of the anomaly was three in 76 angiograms. The incidence at the Indiana University Medical Center was only one case in 1,000 angiograms. The probability of the difference in incidence at the two hospitals being significant cannot be evaluated statistically because of the few positive cases. It does seem, however, that the difference is more than coincidental.

Ophthalmodynamometry was performed on two of our patients. Retinal artery pressures were the same in one and significantly different in the other. The difference may be explained by the head of pressure being decreased in the ophthalmic artery distal to a large shunt.

Summary

Four cases of carotid-basilar anastomosis demonstrated angiographically are reported. Although no characteristic clinical syndrome is associated with this anomaly, our cases suggest that there is a higher incidence of this condition in patients with retarded mental development dating from infancy. Carotid-basilar anastomosis is a possible cause of significant differences in retinal artery pressures.

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References

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