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Hypoplasia of the Basilar Artery

Three Case Reports

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Summary. Three patients, aged 65, 70, and 80 years, respectively, with hypoplastic basilar artery are presented. To our knowledge no similar case has been reported in the literature till now. In one of the cases, the hypoplasia of the basilar artery was an incidental finding. The other two patients had transient neurological symptoms considered characteristic of circulatory insufficiency in the vertebrobasilar system. One of the latter two patients died of acute myocardial infarction and the other patient died after developing occlusion in the cervical part of the right internal carotid artery. The exact nature of hypoplastic basilar artery is uncertain. The possible mechanisms of its occurrence are discussed.

Key words: Basilar artery – Hypoplasia

Introduction

The anomalies of the basilar artery are rare in comparison to that of the other intracranial arteries constituting the circle of Willis [3, 8, 25] with short segments of duplication reported to be the most common variation [8, 23, 25]. In exceptional cases the nonunion of the vertebral arteries has also been observed representing the persistence of the embryonic state [1, 18, 20]. McCullogh described a case in which the basilar artery was completely plexiform [19] and Lasjaunias et al. found segmental aplasia of the basilar artery in a 15-year-old boy [15]. The basilar artery may also exhibit considerable variability in caliber in patients with persistent carotid-basilar anastomosis [3, 25].

The purpose of this paper is to present three cases of hypoplastic basilar artery since no similar case could be found in the available literature.

Case Reports

Case 1. A 80-year-old woman with hypertension of several years duration was admitted to our department because of sudden onset of left-sided hemiparesis. Neurological examination revealed central facial weakness and moderate spastic hemiparesis with exaggerated tendon reflexes and pyramidal signs on the left side as well as bilateral horizontal nystagmus; she was alert. Angiography was not performed. The CSF obtained by cisternal puncture was clear and its protein content normal. A diagnosis of vertebrobasilar circulatory insufficiency was made. The patient's neurological condition improved rapidly and she had no neurological symptoms 4 days after her admission. The patient died suddenly on the 14th day. General autopsy disclosed an acute myocardial infarction as the cause of death. Furthermore, a generalized atherosclerosis was noted. The arteries at the base of the brain exhibited diffuse thickening of their wall. The basilar and vertebral arteries were of strikingly small caliber with an external diameter of about 1.2–1.4 mm almost in their entire length (Fig. 1). The other arteries of the circle of Willis appeared to be normal in size. Sectioning the brain coronally showed no macroscopic alterations. Microscopic examination revealed diffuse changes in the whole brain which are held to be characteristic of ischemia. There was no evidence of infarction.

Case 2. A 65-year-old woman with a 6-year history of hypertension and atrial fibrillation developed left-sided hemiparesis suddenly on the morning of her admission. She had been admitted to a local hospital 4 years previously because of

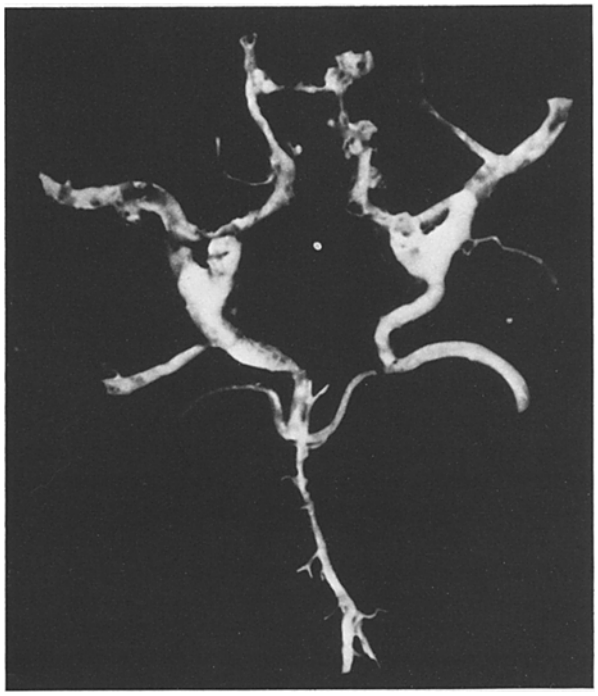

Fig. 1. Case 1. Photograph of the dissected circle of Willis. The basilar and vertebral arteries show unusually small caliber

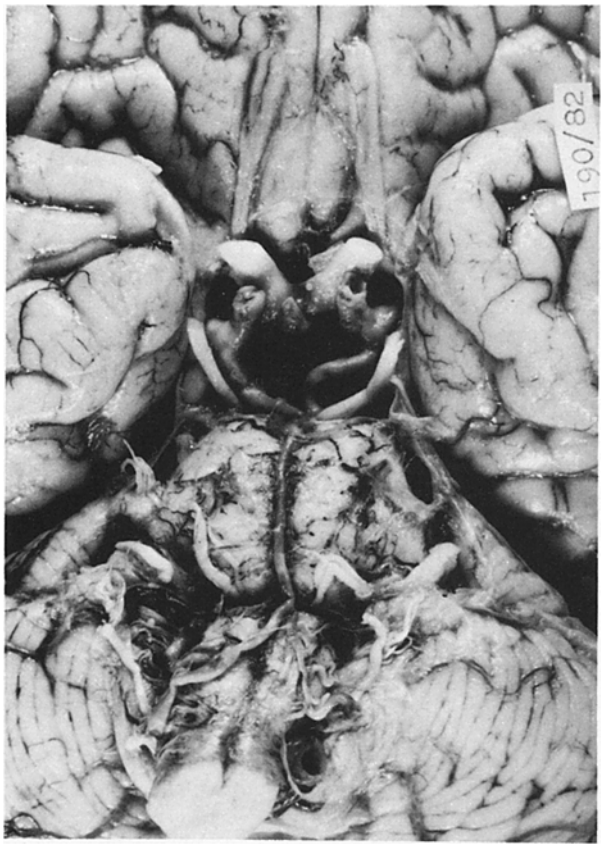

Fig. 2. Case 2. Photograph of the base of the brain. The basilar and vertebral arteries are greatly reduced in external diameter

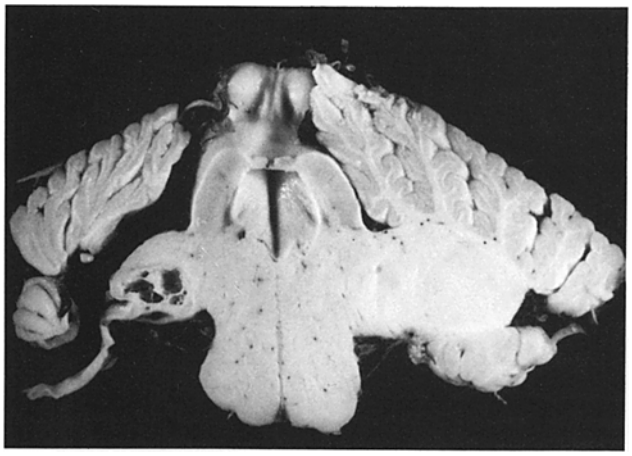

Fig. 3. Case 2. Photograph of the brain stem. The left middle cerebellar peduncle is atrophic and contains cavities of various sizes

dizziness, vomiting, and gait instability and was found to have nystagmus to the right as well as moderate flaccid hemiparesis without pyramidal signs and severe incoordination of the extremities on the left side. Angiography had not been performed. The patient had been treated with vasodilator agents and was discharged with minimal residual symptoms after 1 month. Her condition had remained unchanged until the day of admission to our Department. At this time neurological examination revealed central facial weakness and severe flaccid hemiparesis with pyramidal signs on the left side. Her head and eyes deviated to the right, and she was somnolent. Cisternal puncture yielded clear CSF with moderately elevated protein content. Right percutaneous carotid angiography disclosed total occlusion of the internal carotid artery about 4 cm above its origin. Despite treatment with the usual drugs the patient's neurological and general condition gradually deteriorated. She developed bronchopneumonia and died of cardiorespiratory failure on the 10th day in hospital. General autopsy revealed advanced generalized atherosclerosis, left ventricular hypertrophy of the heart and extensive confluent bronchopneumonia on both sides. The arteries at the base of the brain exhibited moderate atherosclerosis. The entire basilar artery was of unusually small caliber with a diameter of about 1.6 mm (Fig. 2). The vertebral arteries showed a similar caliber. The other arteries of the circle of Willis were of the usual size. On sectioning the brain coronally a recent infarction was found in the whole right hemisphere. On the left side, cavities of various sizes could be observed in the atrophic brachium pontis representing old infarction (Fig. 3).

Case 3. A 70-year-old man with a history of hypertension and diabetes mellitus for several years was referred to our department because of the weakness of the left extremities developing suddenly. Neurologically, he was found to have central facial and hypoglossal lesions, spastic hemiparesis more pronounced in the arm, exaggerated tendon reflexes with pyramidal signs and hypesthesia on the left side. He also had conjugate deviation and adversion of the head to the right. He was drowsy but responsive. The blood sugar was 14.4 mmol/l, and other routine laboratory findings were normal; CSF obtained by cisternal puncture was clear and had a normal protein content. Right percutaneous angiography demonstrated total occlusion of the internal carotid artery close to its origin. Despite the usual treatment the neurological symptoms of the patient worsened rapidly. He developed fever and died of bronchopneumonia 3 days after his admission. General autopsy revealed advanced generalized atherosclerosis, bilateral bronchopneumonia, left ventricular hypertrophy of the heart, bilateral arteriolosclerotic nephrosclerosis and arteria renalis duplex on the left side. At the base of the brain, the blood vessels constituting the arterial circle showed diffuse thickening of their wall. The basilar and both vertebral arteries were of remarkably small caliber. The basilar artery had a diameter of approximately 1.8 mm and the vertebral arteries were about 1.5 and 1.0 mm, respectively, in diameter. The coronal sections of the brain exhibited recent infarction being partly hemorrhagic in the whole right hemisphere. No infarction could be found in the brain stem or in the cerebellum even on microscopic investigation.

In each case, the posterior cerebral arteries originated from the internal carotid arteries and the posterior communicating arteries were larger than the basilar artery.

The hypoplastic basilar arteries were sectioned serially and specimens were taken from the other major intracranial arteries on both sides. The sections were stained with hematoxylin-eosin, orcein, elastic van Gieson, periodic acid-Schiff and Gömöri's method for reticulin.

Except for the unusually small external diameter, the basilar arteries showed a normal structure with moderate concentric or eccentric subendothelial fibrosis in each case (Fig. 4A and B). The internal elastic lamina was relatively well preserved. The thickened intima and the media contained a dense network of fine reticular fibers with no considerable accumulation of collagen fibers (Fig. 5). The other major

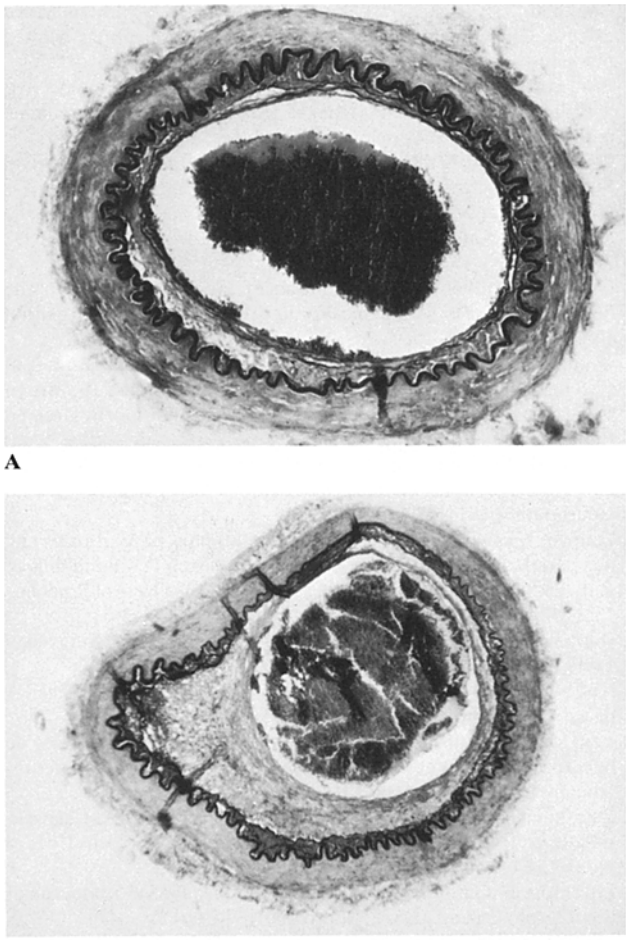

Fig. 4A, B. Microscopic appearance of the undersized basilar artery. (**A**) in case 1; (**B**) in case 3. Elastic van Gieson × 100

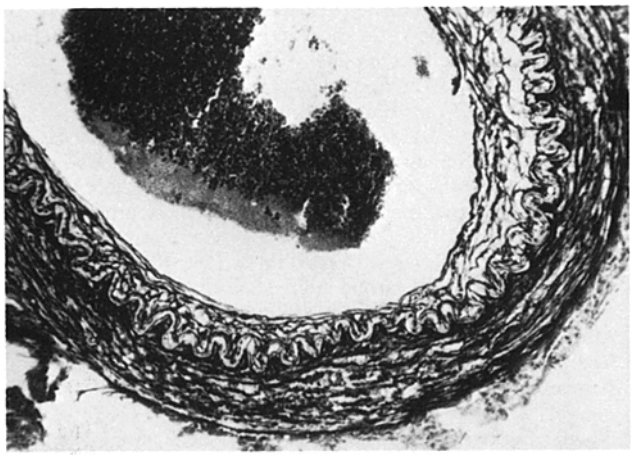

Fig. 5. Case 1. A part of the diminutive basilar artery. The arterial wall contains normal amounts of reticular fibers. Gömöri's method for reticulin × 160

intracranial arteries exhibited changes characteristic of aging and they were free from severe atheromata.

Discussion

Scanty case reports can be found in the literature on unilateral and bilateral hypoplasia of the internal carotid artery [5, 11, 14, 16, 17, 22, 24, 26], but no paper could be encountered dealing with a similar condition restricted to the basilar artery.

Our third patient died after developing occlusion of the right internal carotid artery and hypoplasia of the basilar artery was an incidental finding. The other two patients had neurological symptoms attributable to the circulatory insufficiency of the vertebrobasilar system. One of the two patients had a total recovery with no any evidence of definite infarction on postmortem investigation. In spite of the fact that the other patient developed thrombosis in the right internal carotid artery before death, she was also found to have an old infarction in the left middle cerebellar peduncle corresponding to the border zone of the territories supplied by the short and long circumferential branches of the basilar artery. This finding explains her neurological symptoms which had occurred 4 years previously.

Hypoplasia of the internal carotid artery has been reported to remain asymptomatic [11, 22], and rarely can it cause cerebral ischemia [26]. In the majority of cases, intracerebral or subarachnoid hemorrhage was found associated with hypoplasia of the internal carotid artery [5, 14, 16, 24]. The origin of bleeding was attributed either to dilated vascular anastomotic channels [5] or to a berry aneurysm of the anterior communicating artery coexisting with hypoplasia [14, 16, 24].

Hypoplasia of the basilar artery in contrast to that of the internal carotid artery seems to cause symptoms if any, only at advanced age. The age of patients with the latter condition varied between 17 and 56 years. The age of our patients at the time of the occurrence of symptoms being characteristic of vertebrobasilar insufficiency was 61 and 80 years, respectively.

According to the literature there are some possibilities of maintaining adequate blood supply to the brain stem and cerebellum even in cases of undersized basilar artery. Within the skull the most important potential circuit is through the circle of Willis [8, 25]. Some anastomoses exist between the posterior cerebral and cerebellar arteries [2, 7, 12, 27], and between the pontine branches of the lower basilar and both posterior cerebral arteries [7]. Since in each of our cases the posterior cerebral arteries originated from the internal carotid arteries and since the posterior communicating arteries were larger than the basilar artery itself, the communications through the circle of Willis may have had an important contributory role in the maintance of blood flow in the vertebrobasilar system. The persistence of carotid-basilar anastomoses existing in early embryonic life could not be verified at autopsy. Naturally, persistent carotid-basilar anastomoses of small caliber can be overlooked but in such cases, they contribute little to vertebrobasilar circulation [25].

Unfortunately, vertebral angiography was performed in none of our cases because of the advanced age of one of the patients and because of the symptoms of the other two patients being characteristic of circulatory insufficiency in the internal carotid system. Therefore, the possible collateral circulation could not be visualized in this way, and discussion of the role of hypoplastic basilar artery in the development of thrombosis in the right internal carotid artery would be merely speculative.

The cause of hypoplasia of the basilar artery is unknown as is that of the internal carotid arteries. In cases of hypoplastic internal carotid artery, several authors suppose that a malformation is responsible for the unusually small caliber of the artery [16, 24, 26]. In the knowledge of the development of

arterial system [6, 9, 21], however, the rare malformations of the basilar artery mentioned in the Introduction are easier to understand than its unusually small caliber at an advanced age. Nevertheless, one may suppose that an injurious effect acting either at the perinatal period or in early childhood may impede the normal reproduction of the smooth muscle cells in the media from maintaining the capacity of the artery to grow with the brain. Several diseases occurring in the perinatal period such as basal meningitis, arteritis and arterial occlusion as well as trauma in early childhood have been incriminated in the pathogenesis of hypoplasia of an artery [5, 16]. The intact lamination of the arterial wall and the absence of diffuse atrophic process together contradict the possibility of a pathologic process acquired early in life being the direct cause of the small caliber of the basilar artery in our cases.

Haltia et al. reported two cases of Moya Moya disease in which all constituents of the circle of Willis including the basilar artery and the intracranial portion of the vertebral arteries were similar to the basilar and vertebral arteries of our patients [10]. However, Moya Moya disease involves more the anterior part of the arterial circle and usually becomes manifest before 50 years of age [4, 13]. Furthermore, in Moya Moya disease considerable thickening and folding of the internal elastic lamina are held to be the most common pathological features [4, 10, 28]. Microscopic examination of the basilar arteries revealed the structure of the arterial wall to be proportionally similar to that observed in cases of hypoplastic internal carotid arteries.

Some authors assume that the unusually small caliber of an artery would be the result of involution from some unknown cause after reaching full development [17, 22]. It is well-known that an artery that becomes unnecessary during development undergoes regression [6, 25] and that the size of an artery depends on the area of brain that it ultimately supplies [25]. In each of our cases, the basilar and vertebral arteries supplied only the brain stem and the cerebellum which were normal in size. The carotid system obviously contributed to the maintance of adequate blood supply of the vertebrobasilar territory through the large posterior communicating arteries.

Summarizing the possible pathogenesis of hypoplastic basilar artery two mechanisms are to be taken into consideration. 1) Some injurious factor may disturb the normal reproductive cycle of the smooth muscle cells in the media during embryonic life resulting in the small caliber arteries and change in the hemodynamic state. 2) The persistence of perfusion from the carotids to the vertebrobasilar system could result in the small caliber arteries.

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