

Association between an aplastic basilar artery, unaccompanied by a primitive carotid–vertebrobasilar anastomosis, and multiple aneurysms on the dominant posterior communicating artery

Case report

SANJAY BEHARI, M.CH., HIMANSHU KRISHNA, M.S., MARAKANI V. KIRAN KUMAR, M.S., VIJAY SAWLANI, M.D., RAJENDRA V. PHADKE, M.D., AND VIJENDRA K. JAIN, M.CH.

Departments of Neurosurgery and Radiodiagnosis, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

✓ Basilar artery (BA) aplasia when unaccompanied by a primitive carotid–vertebrobasilar anastomosis is exceedingly rare. The association of BA aplasia with two aneurysms on the dominant posterior communicating artery (PCoA) has not been previously reported.

This 40-year-old man presented in a state of drowsiness and responded to simple commands only after being coaxed. He had complete left cranial third nerve palsy, right hemiparesis, and persisting signs of meningeal irritation. A computerized tomography (CT) scan revealed subarachnoid and intraventricular hemorrhage. An angiogram revealed BA aplasia. The right PCoA followed a sinuous course with multiple loops and provided the dominant supply to the posterior circulation. This vessel harbored two aneurysms, one at the origin of the PCoA from the internal carotid artery and the other at the looping segment just proximal to the brainstem. The left PCoA was extremely thin. The pterional transsylvian approach was used to clip the two aneurysms on the PCoA.

The hemodynamic changes produced by the BA aplasia may have produced alterations in the cerebral vasculature leading to aneurysm formation and consequent subarachnoid hemorrhage.

KEY WORDS • aneurysm • basilar artery • subarachnoid hemorrhage

DURING the early gestational period, the forebrain is supplied by the CA system. The hindbrain also receives contributions from this system via the primitive segmental arteries (the trigeminal, proatlantic, hypoglossal, and otic arteries). The association of BA hypoplasia with a primitive CA–VBA anastomosis, such as a persistent primitive trigeminal artery, is known;^{2,5,8,12,13,15} however, the presence of BA aplasia without such an anastomosis is extremely rare.^{4,7,10,11,17} In our patient, the BA aplasia was associated with two aneurysms on the dominant PCoA. In this paper we discuss the implications of BA aplasia and its associated anomalies on the hemodynamics of the cerebral circulation.

Case Report

Examination. This 40-year-old man experienced a sudden loss of consciousness that lasted 4 hours, 2 days before his admission to our hospital. On regaining consciousness, he remained drowsy and responded to simple commands only after being coaxed. He had a complete left third nerve palsy, right hemiparesis (Grade III), and persisting signs of meningeal irritation (Hunt and Hess Grade 3). A plain CT scan

(Fig. 1) revealed the presence of an SAH with a hematoma in the prepontine and left cerebellopontine angle cisterns, which compressed the left cerebral peduncle. The hematoma extended through the choroidal fissure into the left temporal horn and trigone. Blood was also evident in the bilateral frontal horns, the third and fourth ventricles, and the left sylvian fissure (Fisher Grade 4). The patient began a regimen of dexamethasone (4 mg/6 hours), nimodipine (40 mg/4 hours), and phenytoin sodium (100 mg/8 hours).

Right ICA digital subtraction angiography (Fig. 2 *upper* and *center*) and axial CT angiography (Fig. 2 *lower*) revealed a tortuous right PCoA that supplied the bilateral PCAs and SCA, as well as the distal segment of the BA. This PCoA was associated with two aneurysms. One aneurysm was located at the right PCoA–ICA junction and the second aneurysm was situated at the loop formed by the right PCoA, just proximal to its junction to the PCA. The rest of the BA was not visualized. Both anteroposterior (Fig. 3 *left*) and lateral (Fig. 3 *center*) VA angiograms demonstrated the posterior inferior cerebellar artery filling from the VA but, again, because the BA could not be visualized aplasia of the proximal basilar trunk was suspected. The left CA revealed an extremely thin left PCoA (Fig. 3 *right*).

Operations. At surgery, we performed a right pterional craniotomy to clip the aneurysm located on the right PCoA–ICA junction by using a 5-mm straight Sugita clip (Mizuho Co., Inc., Nagoya, Japan). The right anterior choroidal artery was very close to the aneurysm and had to be gently

Abbreviations used in this paper: BA = basilar artery; CA = carotid artery; CT = computerized tomography; ICA = internal CA; PCA = posterior cerebral artery; PCoA = posterior communicating artery; SAH = subarachnoid hemorrhage; SCA = superior cerebellar artery; VA = vertebral artery; VBA = vertebrobasilar artery.

Basilar artery aplasia

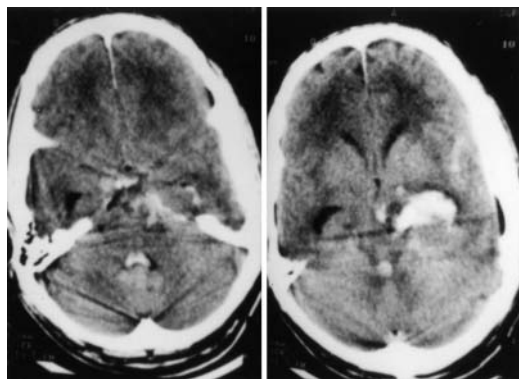


FIG. 1. Plain CT scans revealing an SAH with a hematoma in the prepontine and left cerebellopontine angle cisterns, which compresses the left cerebral peduncle. The hematoma is extending through the choroidal fissure into the left temporal horn and trigone. Blood is also evident in the bilateral frontal horns, the third and fourth ventricles, and the left sylvian fissure.

dissected away before the clip could be applied. The neck of the second aneurysm, which was located at the loop of the right PCoA, just proximal to its junction to the PCA, could not be visualized. This was because the tortuous loop formed by the distal right PCoA, from which the second aneurysm arose, was directed toward the left side and was hidden by the dorsum sellae.

Following the first surgery, the patient's motor strength on the right side deteriorated to Grade 1. A repeated CT scan allowed us to rule out a new hematoma or infarction. After 4 days, we performed a left pterional craniotomy, and by following the transsylvian approach, we were able to visualize the hypoplastic left PCoA and the third nerve in the interpeduncular cistern, trace the looping right PCoA up to the neck of the aneurysm, and treat the aneurysm by using a 7-mm straight Sugita clip (Mizuho Co. Inc.).

Postoperative Course. Following the second surgery, there was a gradual improvement in the patient's hemiparesis, but bronchial pneumonia developed and a tracheostomy had to be performed. Antibiotic medications and tapering doses of steroids were continued for 2 weeks. Oral nimodipine was administered for 6 weeks.

A repeated right ICA angiogram did not demonstrate any residual neck, but there was no filling of the right PCoA (Fig. 4 left). A left ICA angiogram, on the other hand, revealed a compensatory filling of the thin left PCoA, which had begun to supply the PCAs, SCAs, and distal BA (Fig. 4 right).

A neurological assessment performed 6 weeks later revealed that the patient was conscious, able to follow simple commands, and had persisting right hemiparesis (Grade 2), motor aphasia, and third nerve palsy. He tolerated well the removal of the tracheostomy tube. A repeated CT scan did not demonstrate any evidence of infarction or hematoma. At a follow-up examination performed after 3 months, the hemiparesis had improved to Grade 3.

Discussion

Embryogenesis of the Posterior Circulation

In the early gestational period, the posterior circulation



FIG. 2. Right ICA digital subtraction angiograms obtained in the anteroposterior (upper) and lateral (center) views and an axial CT angiogram (lower) revealing a tortuous right PCoA supplying the bilateral PCAs and SCAs, as well as the distal segment of the BA. This PCoA is associated with two aneurysms, one located at the right PCoA-ICA junction and the second at the loop formed by the right PCoA just proximal to its junction to the PCA. The rest of the BA is not visualized. The asterisks indicate the positions of both anterior clinoid processes and the arrows indicate the two aneurysms.

is supported by the paired plexiform longitudinal neural arteries. These arteries form connections with the ICA via the PCoA as well as via the primitive segmental arteries (the trigeminal, proatlantic, hypoglossal, and otic arteries). The caudal end of the PCoA develops into the PCA.² From approximately the 5th week of gestation, the primitive segmental arteries involute. The adult BA develops from the paired ventral longitudinal neural arteries after their fusion on the anterior surface of the neural tube. If the BA becomes the main supplier of blood to the developing PCAs, the PCoAs become smaller. If the latter vessels remain large, however, a "fetal origin" of the PCA persists. The VAs at C-1, which represent remnants of the proatlantic or first cervical segmental artery, also provide a symmetrical source of supply to the BA. The quality of the fusion process and the contributions of the regional segmental artery remnants to the posterior fossa supply determine the variations occurring in this region. Thus, an anomaly in the involution of the primitive segmental arteries or in the fusion of the paired plexiform longitudinal neural arteries may result in aplasia or hypoplasia of the BA.^{2,10}

Redistribution of Circulation and Pathogenesis

Maintenance of an adequate blood supply to the brainstem and cerebellum, in the case of a hypoplastic BA un-

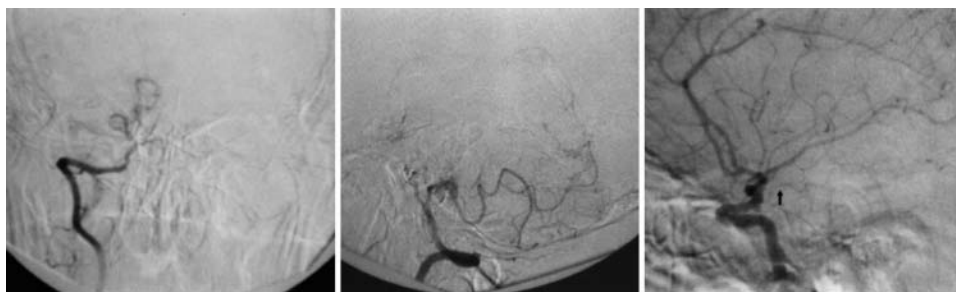


FIG. 3. Anteroposterior (*left*) and lateral (*center*) right VA angiograms demonstrating the posterior inferior cerebellar artery filling from the VA. Again the nonvisualization of the BA is indicative of aplasia of the proximal basilar trunk. A left CA reveals a thin left PCoA (*right, arrow*).

accompanied by a primitive CA–VBA anastomosis, is accomplished by the following: the PCoA, which supplies the distal segment; the VAs, which supply the proximal segment; the anastomoses existing between the pontine branches of the lower BA and both PCAs; and a persistent CA–VBA anastomosis of small caliber, which may not be visible on an angiogram.^{4,7,10,11}

Hegedus,⁷ studying the possible pathogenesis of a hypoplastic BA, has discussed two theoretical possibilities. According to the first, an injury occurring either during the perinatal period or in early childhood may impede the normal reproduction of smooth-muscle cells in the media and decrease the capacity of the artery to grow within the brain. Alternatively, an involution of the artery after it reaches full development may also be implicated. This is because an artery that becomes unnecessary during development undergoes regression, and the final size of the artery depends on the area of the brain that it ultimately supplies. If the supply of the VBA system is restricted to only a part of the brainstem and cerebellum, with a major contribution from the CA system through the large PCoAs, then due to a spontaneous involution within the VBAs, small-caliber or even aplastic arteries may result.

Review of the Literature

The association between BA hypoplasia and a primitive CA–VBA anastomosis, such as a persistent primitive trigeminal artery, is well known.^{2,5,8,12,13,15} Fortner and Smoker⁵ evaluated a persistent primitive trigeminal artery aneurysm by performing MR imaging and angiography and discussed the association of BA hypoplasia. Masaryk and associates¹³

and Schuierer, et al.,¹⁵ studied MR angiograms of the primitive trigeminal artery and found an associated hypoplastic proximal BA. Boyko and colleagues² found three cases in which there was a persistent trigeminal artery, in which all proximal BAs were hypoplastic. These authors emphasized the need to recognize this associated congenital variant and not to confuse it with an atherosclerotic stenosis or occlusion of the BA. The persistence of a primitive segmental artery between the CA and VBA systems is often associated with aplasia or hypoplasia of the proximal BA and the ipsilateral VA because there is no flow-related stimulus for the BA, proximal to the anastomosis, to develop along with the embryo.²

There are only occasional case reports of BA aplasia or hypoplasia that occurs without a primitive CA–VBA anastomosis, similar to that seen in the present study. Lasjaunias and coworkers^{10,11} found segmental aplasia of the BA in a 15-year-old boy. Hegedus⁷ presented three cases of hypoplastic BA. Chaturvedi and colleagues⁴ reported finding posterior circulation ischemia in the territory of the hypoplastic VBA system in eight patients. Seven of these patients also had at least one hypoplastic VA (bilateral in five cases) and in six patients both PCAs originated from the ICAs. In a report published before CT scanning was available, Szdzuy and Lehmann¹⁷ described a 56-year-old woman who presented with symptoms of brainstem ischemia due to hypoplasia of the BA and both VAs. In that case the diagnosis was based on findings on a conventional angiogram. In a situation similar to the present case, in none of these patients was there a coexisting primitive CA–VBA anastomosis.

An Alternative Hypothesis

An acquired occlusion of the BA from a dissection,^{14,18,19} resulting in a progressive dilation of the dominant right PCoA with the eventual development of aneurysms due to the hemodynamic overload, is an attractive alternative hypothesis. Our patient, however, experienced a sudden loss of consciousness and the development of neurological deficits following an SAH caused by the aneurysm located on a tortuous bend of the PCoA just proximal to the brainstem. This occurred just 2 days prior to his admission to the hospital and he was asymptomatic before this ictus. The occluded segment of the BA (the site for the proposed dissection) was not the site responsible for the SAH. It would be unusual for a temporal sequence of events to occur in which an acquired occlusion of the BA, the development of the two aneurysms, and their subsequent enlargement, as well

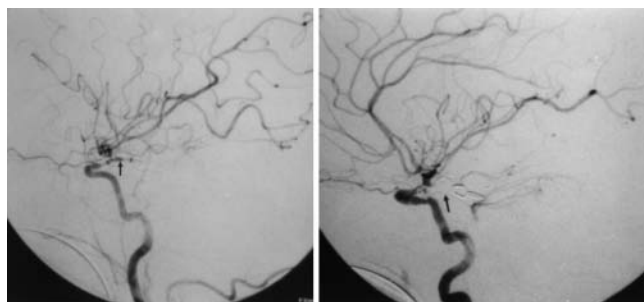


FIG. 4. *Left*: A repeated right ICA angiogram demonstrating no residual neck and no filling of the right PCoA (*arrow*). *Right*: Left ICA angiogram demonstrating a compensatory filling of the hypoplastic left PCoA (*arrow*).

as the rupture of one aneurysm, occurred within a short span of 2 days. The possibility that a dissection of the BA took place long before the aneurysmal SAH is also ruled out because it is highly unlikely that the patient would have been completely asymptomatic after a complete occlusion of the BA, especially when there was no alternative supply to the posterior circulation such as a primitive CA–VBA anastomosis.^{14,18,19} Angiograms of the posterior circulation did not reveal any signs of BA dissection such as an irregularity within the vessel lumen, a linear filling defect within the lumen, or the presence of an irregular aneurysm associated with focal luminal narrowing or irregularity.^{14,18} Both PCoAs, although of different sizes, were capable of supporting the posterior circulation in the presence of the aplastic BA. There was no clinicoradiological evidence of the existence of connective tissue disorders such as Ehler–Danlos syndrome, fibromuscular dysplasia, or moyamoya disease, which may cause intracranial dissections, vessel occlusions, or aneurysm formation. There was also no history of trauma or infection.¹⁸ Thus, a congenital aplasia of the BA rather than an acquired dissection was considered to be the more plausible explanation for the vessel occlusion.

Observations in Our Case

In our patient, the midsegmental BA aplasia was associated with a tortuous right PCoA, which was the major feeding vessel to the PCAs and SCAs as well as to the distal BA. The hypoplastic left PCoA was barely visible on the preoperative angiogram. Thus, a significant portion of the posterior fossa arterial supply was dependent on the right PCoA. The gross disparity between the luminal capacity of the right PCoA and the continuous demand on it to maintain a large portion of the posterior fossa circulation may have played a major role in the genesis of these two aneurysms.

While studying PCoAs in 126 cranial cavities of adult cadavers, Bisaria¹ found a junctional dilation of the PCoA in 6.3% of cases. Apart from the junctional dilation, other dilated portions of the PCoA were observed in 39.7% of their specimens. These were mostly fusiform in shape and a few showed ectasia of the arterial wall. Dilations were found in arteries with a diameter less than 2.5 mm. Bisaria,¹ Stehbens,¹⁶ and Fox, et al.,⁶ considered such dilations to be preaneurysmal and possessing the risk of leaking or rupturing, resulting in an SAH. Chase³ has suggested that gross anomalies of the cerebral vasculature (exemplified by the tortuous course of the dominant PCoA in our case) may be accompanied by structural defects in the wall of the vessels.³ Thus, the tortuous PCoA with aplasia of BA perhaps produced hemodynamic changes sufficient to expand two of the naturally occurring dilations of the PCoAs into aneurysms.^{1,3,9}

The underlying condition could only be detected when the distal aneurysm, located on a tortuous bend of the vessel just proximal to the brainstem (with its fundus pointed toward the left side), bled into the interpeduncular and left cerebellopontine angle cisterns and the ensuing hematoma compressed the left oculomotor nerve and the left cerebral peduncle. Following clipping of both the aneurysms, the important right PCoA did not show any filling. Fortunately, the circulation through the thin, yet patent, left PCoA compensated for this loss and the patient made a gradual recovery. Although a pterional approach from both sides was used to clip the aneurysms, perhaps drilling of the posterior

clinoid process by using a single approach or performing surgery via the subtemporal approach would have enabled access to both aneurysms during a single operation.

To the best of our knowledge, the association of BA aplasia with a dominant, tortuous PCoA that harbors two aneurysms and presents as an SAH has not been previously reported.

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Address reprint requests to: Vijendra K. Jain, M.Ch., Department of Neurosurgery, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Rae Bareilly Road, Lucknow 226014, India. email: vkjain@sngpi.ac.in.