Ruptured aneurysm arising from a basilar artery fenestration and associated with a persistent primitive hypoglossal artery

Case report and review of the literature

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Several cases have been reported in which a persistent PHA or a BA fenestration was associated with an intracranial aneurysm. The PHA is a rare remnant of one of five embryonal CA–BA anastomoses, and a fenestration of the vertebrobasilar system is a developmental anomaly that represents a normal variant. We report on a patient with a ruptured aneurysm at a BA fenestration that was associated with a left PHA. The patient was successfully treated by endovascular embolization.

Although many clinical cases of ruptured aneurysms associated with either a PHA or a BA fenestration have been reported, there has been, as far as we know, no case in the literature in which a ruptured aneurysm associated with both anomalies and no case in which endovascular embolization was used to treat a ruptured aneurysm associated with a PHA. This rare case is discussed and a review of the relevant literature is presented.

KEY WORDS • aneurysm • basilar artery fenestration • endovascular therapy • primitive hypoglossal artery

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Case Report

This 42-year-old woman experienced the sudden onset of a severe headache and was admitted to our hospital on the next day.

Examination. Neurological findings on admission were headache, neck stiffness, and left abducent nerve palsy. A CT scan demonstrated high-density areas in the prepontine cistern, indicating the presence of subarachnoid hemorrhage. Three-dimensional CT angiography revealed a persistent PHA on the left side, which entered the skull via the hypoglossal canal, and an aneurysm arising from the junction of the PHA and the BA (Fig. 1). A left ICA angiogram demonstrated a PHA originating from the C-2 vertebral level (Fig. 2). The bilateral VSAs and the bilateral PCoAs were not visible. Selective angiography of the PHA demonstrated a fenestration at the PHA–BA junction and an aneurysm at the proximal end of this fenestration (Fig. 3).

Operation. Following induction of general anesthesia, the patient underwent endovascular embolization of the aneurysm. A microcatheter (Excelsior SL-10; Target Therapeutics/Boston Scientific, Freemont, CA) was introduced into the aneurysm sac via the PHA and two Guglielmi Detachable Coils were placed in the aneurysm. The patient’s postoperative course was uneventful, and she was able to resume her normal life.

Postoperative Course. A postoperative angiogram confirmed complete obliteration of the aneurysm and a normal circulation (Fig. 4). The patient’s postoperative course was uneventful, and she was able to resume her normal life.

Discussion

It is well known that the PHA is one of the rare embryological remnants of five CA–BA anastomoses. Lie25 pro-
posed four criteria for the anatomical and angiographic definition of a PHA: 1) the artery arises from the ICA at the C1–3 level; 2) the PHA enters the skull via the anterior condylar foramen (the hypoglossal canal); 3) the BA is filled only beyond the point of entry of the PHA; and 4) there is no PCoA. Our case satisfied all four criteria.

Similar to BA fenestrations, the fragility of the vascular wall and/or hemodynamic stress due to the presence of a PHA are reportedly related to aneurysm formation. Including our report, 40 cases of PHA associated with ruptured aneurysms have been described in the literature.1,3-5,7-10,12,13,15,16,18,19,22,23,25,26,28,30,31,33-36,38,39,41-45 In 65.8% of these cases the patients were between 30 and 59 years of age (mean 45.6 years), indicating that the age of patients with ruptured aneurysms associated with a PHA is lower than that usually seen in patients with subarachnoid hemorrhage. In these reports the PHA was located on the right side in 20 cases, on the left in 19, and bilaterally in one case. With respect to the location of these aneurysms, 31.4% were located at the PHA–BA junction and 53% in the posterior circulation. In cases in which anterior circulation aneurysms were present, the distal anterior cerebral artery and the middle cerebral artery were involved in 13.7 and 11.8% of cases, respectively; in only 5.9% of cases was the anterior communicating artery involved and in no reported case was the ICA–PCoA involved. In cases in which there was a PHA, many aneurysms were located in the posterior circulation. The PHA is considered to have some association with aneurysms arising in the posterior circulation.

On the other hand, the presence of a BA fenestration, which is a rare anomaly, is due to the failure of the bilateral paired longitudinal neural arteries to fuse in the 7- to 12-mm-long embryo, when five anastomoses begin to regress and disappear. Basilar artery fenestrations most frequently have been located in the proximal BA trunk, close to the junction of the VAs (73%); 35.5% have been associated with vertebrobasilar junction aneurysms.40 Histopathological examinations of intracranial fenestrations have demonstrated structural defects in the media of the medial walls at the two ends of the fenestration.43,43,44 These muscular gaps and increased hemodynamic stress have been suggested as factors in the high incidence of aneurysms at the proximal end.43,43,44

As mentioned earlier in this paper, these vertebrobasilar system anomalies, a PHA and a BA fenestration, appear to share common embryological and clinical characteristics that may play a role in the frequent development of aneurysms. Although many cases of ruptured aneurysms associated with either a PHA or a BA fenestration have been documented, to our knowledge ours is the first report of a ruptured aneurysm associated with a PHA and a BA fenestration. We posit that the coexistence of two rare vertebro-

Ruptured aneurysm associated with BA fenestration and PHA

Fig. 1. Three-dimensional CT angiogram demonstrating entry of the left PHA through a hypoglossal canal (double arrows) and an aneurysm arising from the junction of the PHA and BA (single arrow).

Fig. 2. Left ICA angiogram demonstrating the PHA (double arrows) and an aneurysm (single arrow). The PHA originates from the left ICA at the C-2 vertebral level. The aneurysm arises anterior to this site from the PHA–BA junction.

Fig. 3. Anteroposterior (left) and lateral (right) selective PHA angiograms revealing the fenestration (double arrows) at the PHA–BA junction. The aneurysm is located at the proximal end of this fenestration (single arrow).
basilar system anomalies predisposed our patient to the development and rupture of this aneurysm and that hemodynamic stress and/or the inherent fragility of the vascular wall played important roles.

Regarding the treatment of aneurysms associated with a PHA, of the 31 patients who were previously reported to have undergone surgery, 28 underwent aneurysm clip placement. Although there is currently no consensus on the appropriate surgical approach, the VAs are usually hypoplastic or aplastic on both sides and the PCoAs are functionally absent; consequently, in these patients the PHA is usually the only feeding vessel for the posterior fossa circulation. Our search of the literature failed to uncover earlier reports on the use of endovascular embolization to treat aneurysms associated with a PHA. Considering the difficulties inherent in open surgery of aneurysms associated with a PHA, we suggest that endovascular embolization may represent a suitable treatment for aneurysms associated not only with a BA fenestration but also with a PHA.

Conclusions

We report a case in which a ruptured aneurysm was associated with a PHA and a BA fenestration was effectively treated by means of endovascular embolization. Considering the difficulties inherent in open surgery of aneurysms associated with a PHA, endovascular embolization for ruptured aneurysms associated with a PHA should be considered as one of the most suitable treatments.

References


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