Azygos Anterior Cerebral Artery Aneurysm Associated with Fenestration of the Anterior Cerebral Artery

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Summary: We report a case of a ruptured aneurysm occurring at the peripheral bifurcation of the azygos anterior cerebral artery with fenestration at the unilateral horizontal segment, A1. Further, hypoplasia was noted at the contralateral A1. It is considered extremely rare that an aneurysm is complicated by such a combination of anomalies. Presence of these anomalies might significantly influence hemodynamics, and hemodynamic stress might play an important role in the occurrence and growth of an aneurysm.

Key words: aneurysm — azygos anterior cerebral artery — hemodynamic stress — fenestration — cerebral angiography

Introduction

Azygos ACA (Alpers et al. 1959; Baptista et al. 1963; Pool et al. 1965), triple ACA (Baptista et al. 1963) and A1 fenestration (Alpers et al. 1959; Crompton, 1962; Baptista et al. 1963; Teal et al. 1973; Ito et al. 1981; Krayenbuhl et al. 1982) are known as anomalies related to the anterior cerebral artery (ACA). Each of them is a rare vascular anomaly, and only a few reports have been presented concerning an aneurysm associated with such an anomaly (Crompton, 1962; Katz et al. 1978; Kondo et al. 1979; Huber et al. 1980; Ito et al. 1981; Niizuma et al. 1981; Inagawa et al. 1983; Matsumura and Nojiri, 1984; Kitami et al. 1985). This paper is a report of a case of an aneurysm occurring at the peripheral bifurcation of the azygos ACA together with fenestration at unilateral A1 and hypoplasia at contralateral A1.

Case Report

A 56 year-old housewife noticed the sudden onset of severe headache and vomiting on May 17, 1979. Subarachnoid hemorrhage was revealed by a lumbar puncture at another clinic, and the patient was admitted to our clinic on May 23. On admission, she was stuporous and had neck stiffness and subhyaloid hemorrhages in both ocular fundi. CT scan disclosed severe subarachnoid hemorrhage in the longitudinal fissure of the cerebrum. Right carotid angiography revealed fenestration at A1 and a saccular aneurysm at the bifurcation of the pericallosal artery (Fig.1, 2, 3). Left carotid angiography demonstrated hypoplasia at A1, but this artery was not visualized beyond this segment. When left carotid angiography was conducted by compressing the right common carotid artery, contralateral A1 and M1 and one pericallosal artery were visualized.
Fig. 1. Antero-posterior view of right carotid angiogram revealing fenestration in the horizontal segment of the anterior cerebral artery and a ruptured saccular aneurysm at the peripheral bifurcation of the pericallosal artery.

Fig. 2. Magnified antero-posterior view of right carotid angiogram. A1 fenestration indicating by small arrows and saccular aneurysm, large arrow.

Fig. 3. Lateral view of right carotid angiogram showing a ruptured saccular aneurysm (arrow) of single pericallosal artery.

Fig. 4. Antero-posterior view of left carotid angiogram with contralateral carotid compression. Left hypoplastic A1 portion (small arrow), contralateral anterior cerebral artery with fenestration (large arrow) and single trunk of the pericallosal artery are revealed.
through hypoplastic A1, and an aneurysm was identified at the peripheral bifurcation of this artery (Fig. 4). It was concluded on the basis of these cerebral angiographic findings that bilateral ACA formed an azygos artery and that a saccular aneurysm was present at the peripheral bifurcation of this artery. In addition, fenestration was present at right A1 and hypoplasia at left A1.

Operation was performed three weeks after the onset of the hemorrhage and identification of azygos ACA and neck clipping of the saccular aneurysm occurring at the peripheral bifurcation of this artery were carried out. The postoperative course was uneventful. The patient is well without any neurological deficits.

Discussion

The horizontal segment of the ACA often shows morphological variations, and it has been known that an aneurysm of the anterior communicating artery is likely to occur in patients with hypoplasia or aplasia at unilateral A1 (Stehbens, 1963; Ferguson, 1970; Andrews and Spiegel, 1981; Fujimoto et al. 1981). Further, azygos ACA, triple ACA and A1 fenestration have been reported as vascular anomalies occurring at this site (Baptista, 1963); some patients with an aneurysm complicated by such an anomaly are reported in the literature (Crompton, 1962; Katz et al. 1978; Kondo et al. 1979; Huber et al. 1980; Ito et al. 1981; Niizuma et al. 1981; Inagawa et al. 1983; Benebetti and Curri, 1983; Matsumura and Nojiri, 1984; Kitami et al. 1985; Abe et al. 1985).

Azygos ACA may occur because two vessels, which should form two ACA, are fused during the formation of ACA from the medial branch of the primitive olfactory artery around the 40th day of gestation or because the median artery of the corpus callosum appearing around the 44th day of gestation persists and two ACA, which should be formed, are reduced. Fenestration of ACA may be ascribable either to partial persistence for unknown reasons of the plexal structure of ACA noted in the early stage of gestation or to incomplete fusion, but its etiology remains to be elucidated (Padget, 1948; Stehbens, 1963; Ferguson, 1970).

It is not necessarily easy to identify azygos ACA and A1 fenestration on cerebral angiograms. Azygos ACA may be identified more easily by unilateral carotid angiography with contralateral compression of the carotid artery rather than by bilateral carotid angiography (Niizuma, 1981). Azygos ACA was confirmed finally by operation in our case. On the other hand, care must be taken to identify A1 fenestration, because looping of Heubner’s artery, accessory middle cerebral artery and posterior cerebral artery may be identified erroneously as fenestration in the anteroposterior view. For this reason, it is necessary to take oblique and magnified views (Ito, 1981; Matsumura and Nojiri, 1984). A1 fenestration was identified using a magnifier.

To our knowledge, 39 cases of azygos ACA aneurysm have so far been reported (Pool and Potts, 1965; Katz et al. 1978; Kondo et al. 1979; Huber et al. 1980; Niizuma et al. 1981; Fujimoto et al. 1981; Benedetti and Curri, 1983; Kitami, 1985; Abe et al. 1985). Of these 39 cases, 35 had an aneurysm associated with azygos ACA at the peripheral end, one at the center and three at the origin. The incidence of azygos ACA itself is assumed to be about 2% (Alpers et al. 1959; Baptista et al. 1963; Huber et al. 1980; Ito et al. 1981; Kitami et al. 1985) and is a rare vascular anomaly. However, the incidence of an aneurysm in the vicinity of azygos ACA is considerably higher, though it differs among researchers. It was reported as 13.6% by Pool and Potts (1965), 13%, by Niizuma et al. (1981), 25.9%, by Fuji-
moto et al. (1981), 41.1%, by Huber et al. (1980) and 71%, by Kitami et al. (1985). According to Kitami et al. (1985), its incidence was as high as 71%, because a large number of the aneurysmatic lesions, which were non-ruptured or were as small as 3 mm in diameter, were detected as a result of detailed cerebral angiography. It is suggested from such a large number of aneurysmatic lesions at the peripheral end of azygos ACA that hemodynamic stress applied to this site may produce a significant influence on the occurrence and growth of the aneurysm (Stehbens, 1963; Ferguson, 1970; Perlmutter and Photon, 1976; Huber et al. 1980).

Fenestration of ACA has so far been demonstrated by cerebral angiography in 19 patients (Alpers et al. 1959; Stehbens, 1963; Perlmutter and Photon, 1976; Ito et al. 1981; Krayenbuhl and Yasargil, 1982; Inagawa et al. 1983; Matsumura and Nojiri, 1984; Kitami et al. 1985), this was noted in the distal half at A1 in all of them. The incidence of this vascular anomaly with an aneurysm was high (15 of 20 cases including this one), but an aneurysm coincident with fenestration has been noted only in three cases (two cases reported by Inagawa (1983) and one autopsy case by Crompton (1962). Crompton noted a defect in the tunica media at the proximal end of fenestration in the autopsy case and reported that an aneurysm might tend to occur at the vascular bifurcation on the proximal side of this anomaly. However, the number of patients with fenestration of ACA itself is limited, and the likelihood of a complication of this anomaly by ananeurysm is considered not to be high.

In the present case, fenestration was noted at unilateral A1 and hypoplasia, at contralateral A1. ACA formed a single azygos artery, and its peripheral bifurcation was morphologically exposed to considerable hemodynamic stress. The combination of these anomalies was considered to be closely correlated with the occurrence of the aneurysm.

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References


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