Intracranial Atypical Fibromuscular Dysplasia With Ruptured Aneurysm

—Case Report—

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Abstract

A 53-year-old woman was admitted with severe subarachnoid hemorrhage due to rupture of an aneurysm associated with atypical intracranial fibromuscular dysplasia (FMD). Angiography demonstrated the aneurysm and very irregular form of the left internal carotid artery (ICA), the right ICA, and right proximal middle cerebral artery (MCA). Other arteries showed signs of atherosclerosis. The aneurysm was treated by embolization, but she subsequently died of shock of unknown cause. Detailed examination of serial angiograms detected enlargement of the aneurysm and progression of the irregular appearance of the ICA. FMD is a non-inflammatory and non-atheromatous arteriopathy that commonly affects the cervical ICA and sometimes the intracranial ICA. The association with saccular aneurysm is widely known and the prevalence of incidental aneurysms is higher than that in the general population. The common “string of beads” finding is easily distinguished from other vascular diseases, but non-specific findings such as “tubular stenosis” and “diverticular-like outpouching” are harder to differentiate. FMD is associated with various complications and appropriate periodic follow-up examination is required. Detailed analysis of serial angiograms may facilitate diagnosis of this condition.

Key words: intracranial atypical fibromuscular dysplasia, subarachnoid hemorrhage, ruptured aneurysm, Guglielmi detachable coil, string of beads, diverticular-like outpouching

Introduction

Fibromuscular dysplasia (FMD) is a non-inflammatory and non-atheromatous arteriopathy that affects the renal and cervical internal carotid arteries (ICAs).^{2-4,6)} The histological characteristics are intramural smooth muscle cell hyperplasia, fibrous dysplasia, and destruction of the arterial wall.⁶⁻⁸⁾ FMD causes alternating mural thinning and thickening, which appears as an irregular succession of dilatations and stenoses on angiography. This sign is commonly found in the bilateral extracranial ICAs at the level of the atlas or axis portion, and occasionally in the intracranial ICAs.^{2-4,6)} The diagnosis of FMD is based on the angiographic and/or histological features, but definitive diagnosis is sometimes difficult. Histological verification by biopsy is not available in most

cases, so a deductive diagnosis is usually based on the angiographic findings. The common “string of beads” finding is easily distinguished from other vascular diseases, but “tubular stenosis” and “diverticular-like outpouching” are much less common, and are not specific findings.^{2-4,6)} However, a firm diagnosis should be obtained, because FMD is associated with various complications, and appropriate and periodic follow-up examinations are required.

We describe a rare case of atypical intracranial FMD associated with a ruptured aneurysm, which appeared as concentric constrictions and protrusive dilatations of the bilateral intracranial ICAs, and could be distinguished from atherosclerosis or other vascular diseases by serial angiography in combination with clinical and laboratory findings.

Fig. 1 Left internal carotid angiograms in 1996, anteroposterior view (left) and lateral view (right), showing a left internal carotid artery (ICA)-posterior communicating artery aneurysm with 5.5 mm diameter and 4 mm neck. The C₁₋₅ portion of the ICA was mildly irregular which appeared similar to atherosclerotic change.

Case Report

A 53-year-old woman with left hemiparesis was admitted unconscious to our hospital on November 25, 2002. Her medical history included treatment for old atheromatous cerebral infarction in 1996. She had slight left hemiparesis, but maintained an ordinary daily life. An unruptured left ICA-posterior communicating artery (PcomA) aneurysm had been detected in 1996 (Fig. 1). The aneurysm was not very large, with 5.5 mm diameter, so we scheduled periodic follow up by magnetic resonance angiography. After treatment for the infarction and discharge, she did not return to our hospital, but she attended a local hospital for geographical reasons.

On admission, physical examination found no abnormalities except for mild obesity. She had no deformity, and no phacomatosis, erythroderma, or scleroderma of the skin. She did not smoke tobacco and did not have diabetes, and her blood pressure was normal. Her consciousness level was Glasgow Coma Scale 7 (E1V2M4). Computed tomography showed massive subarachnoid hemorrhage in the basal cistern and the bilateral sylvian fissures (Fig. 2). Hydrocephalus was also noted. Left internal carotid angiography demonstrated a heart-shaped aneurysm at the junction of the ICA-PComA (Fig. 3). The aneurysm was 12 mm in diameter, with a 4 mm neck directed posteromedially. The PcomA arose from the medial side of the aneurysm. Other angiographic findings included very irregular form of the left ICA from C₁ to C₅, and the same changes in the right ICA and right proximal middle cerebral artery (MCA), especially

Fig. 2 Computed tomography scan showing the sylvian fissures and basal cistern filled with massive hematoma.

Fig. 3 Left internal carotid angiograms in 2002, anteroposterior view (left) and lateral view (right), showing a heart-shaped aneurysm at the junction of the left internal carotid artery (ICA)-posterior communicating artery with 12 mm diameter and 4 mm neck. Segmental concentric constrictions (arrowheads) and protrusive dilatation (arrow) were seen in the intracranial ICA, but not in the other arteries. Comparison with Fig. 1 shows that the aneurysm had expanded, and the irregular form of the ICA had progressed.

from C₁ to C₅ (Figs. 3 and 4). The extracranial ICAs, anterior cerebral arteries, the left MCA, vertebralbasilar arteries, posterior cerebral arteries, aorta, brachiocephalic arteries, subclavian arteries, and renal arteries showed slight atherosclerosis, but most of these vessels demonstrated a smooth shape.

Fig. 4 Right internal carotid angiograms in 2002, anteroposterior view (left) and lateral view (right), showing similar changes to the left internal carotid artery (ICA) in the right intracranial ICA and the proximal middle cerebral artery. Segmental concentric constrictions (arrowheads) were seen, but no protrusive dilatation and no aneurysm.

Fig. 5 Postoperative left carotid angiogram, lateral view, showing the aneurysm was packed with Guglielmi detachable coils, but a slight neck remnant persisted.

No other aneurysm or vascular anomaly was detected.

Lumbar puncture and spinal drainage were performed to decrease the cerebrospinal fluid pressure, but her consciousness level did not improve. The aneurysm was embolized with Guglielmi detachable coils on Day 2 (Fig. 5). The postoperative course was uneventful and her consciousness level began to improve. She could eat soft food and express her intentions simply, but could not hold complicated conversations. She developed shock after a 1-week history of dull abdominal pain on January 21, 2003 (Day 57). Her blood pressure was below 50 mmHg and her

breathing was deteriorating. We administered dopamine and placed the patient on a ventilator system, but she died the next day (Day 58). Brain computed tomography scans, chest radiographs, electrocardiograms, and blood tests were reviewed, but the cause of her condition could not be established. Autopsy was not performed.

Discussion

Review of 321 published cases of aortocranial FMD described the localization of 423 aortocranial FMD sites, and identified at least 16 cases of intracranial FMD (about 3.5%).^{2,3)} Angiographical characterization of the shape of aortocranial FMD identified three distinct types: Type 1 showed typical “string of beads” appearance, and was the most common pattern in 89% of cases; Type 2 was “unifocal or multifocal tubular stenosis” found in 7%; and Type 3 was “atypical FMD expressed as diverticular-like outpouching” found in 4%.⁶⁾ The association with saccular aneurysm is widely known and the prevalence of incidental aneurysms is $7.3 \pm 2.2\%$, which is higher than the incidence of aneurysms in the general population (5.6%).¹⁾

Aortocranial angiography sometimes demonstrates irregular dilatations and stenoses at the arterial walls. The differential diagnosis includes dissection, atherosclerosis, systemic vasculitis, hypoplasia, collagen disease, meningitis, inherited disorder, and iatrogenic vasculopathy.^{4,5)} Several of these diseases could be excluded in our case based on the appearance, medical history, clinical features, and laboratory data. However, dissection, atherosclerosis, and hypoplasia could not be excluded, so detailed analysis of serial angiography was performed. The first angiography was obtained in December 1996 (Fig. 1), and the next in November 2002 (Fig. 3). Arterial irregularities had been pointed in 1996, and this condition had gradually progressed during the following 6 years. These irregularities appeared as segmental concentric constrictions and partial protrusive dilatations in 2002. These changes were limited to the intracranial portion of ICA, and the other arteries remained smooth. This progressive and concentric form was beyond the sphere of hypoplasia and dissection, and the sparing of the other arteries was not typical of atherosclerosis.²⁻⁶⁾ These constrictive and dilatative forms were very similar to “string of beads” and “diverticular-like outpouching,” and the progressive and sparing property was characteristic of FMD. The final diagnosis was atypical intracranial FMD based on these angiographic features.

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