Arterial Shrinkage

14

Satoshi Kuroda

Abstract

For these 50 years or longer, the diagnosis of moyamoya disease has been based on the information of the lumen of involved arteries, using cerebral angiography and MR angiography. However, this has led to some confusion in the diagnosis of the disease, such as in differentiating it from intracranial arterial stenosis caused by atherosclerosis. On the other hand, studies over the past 10 years have shown that arterial shrinkage appears specifically in affected arteries in moyamoya disease. This phenomenon may not only improve the accuracy of the diagnosis, but may also help to elucidate the still unknown etiology of the disease. In this chapter, I introduce a novel concept of arterial shrinkage specific for moyamoya disease and discuss how we should diagnose moyamoya disease more accurately than before.

Keywords

Moyamoya disease · Outer diameter · Arterial shrinkage · Diagnosis · Pathogenesis · Heavy T2-weighted image · Atherosclerosis

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14.1 Introduction

Moyamoya disease is characterized by progressive stenosis of the terminal portion of the internal carotid artery and its main branches such as middle cerebral artery and anterior cerebral artery. In response to disease progression, the perforating arteries, including the lenticulostriate artery, anterior and posterior choroidal arteries, and thalamoperforating artery, start to dilate and function as one of important collateral routes. Almost 50 years have passed since the moyamoya disease was first reported in an English-written journal in 1969 [1]. Since then, the stenosis of the terminal portion of internal carotid artery and its branches has been considered as the essence of the pathogenesis of moyamoya disease. In fact, this finding is an essential part of the diagnostic criteria even now. Pathologically, the affected arteries such as the carotid terminations include fibrocellular thickening of the intima, an irregular undulation (waving) of the internal elastic lamina, and attenuation of the media (*see Chap. 1*). Initially, cerebral angiography was mandatory for diagnosis, but with the development and widespread use of noninvasive MR angiography, MR angiography has been available since 1996 [2]. In pediatric patients, the diagnosis of moyamoya disease is relatively easy because of the paucity of diseases to differentiate. However, adults, especially the relatively elderly, often have difficulty distinguishing whether the stenosis of the affected arteries is due to moyamoya disease or arteriosclerosis, and the diagnosis is often difficult to make. This difficulty in diagnosis may be result from the fact that the diagnosis has been based only on information about the lumen of the affected arteries, regardless of the modalities used.

In this chapter, therefore, we introduce a novel concept of arterial shrinkage specific for moyamoya disease, which has become evident using novel imaging techniques such as heavy T2-weighted images, and discuss how we should diagnose moyamoya disease more accurately than before.

14.2 Arterial Shrinkage in Carotid Fork

As aforementioned, not rarely we encounter cases in which it is difficult to differentiate moyamoya disease from atherosclerotic intracranial artery stenosis. Through these experiences, I have long been eager to develop a different diagnostic method to more accurately diagnose moyamoya disease than ever before. At the same time, I have long remembered the scene of our first direct observation of the carotid fork suffering from moyamoya disease in 1987 when I was in my second year of residency. At that time, my mentor, Prof. Hiroyasu Kamiyama (*see Chap. 18*) performed a craniotomy on an adult male patient with unilateral moyamoya disease on the right side at Hokkaido University Hospital, Sapporo, Japan. The patient had an aneurysm at the bifurcation of the basilar artery and right superior cerebellar artery, so we planned to perform aneurysm clipping and surgical revascularization at the same time. We reached the carotid cistern through the right trans-Sylvian approach. Then, we noticed that the internal carotid artery and the proximal portion of the

middle cerebral artery and the anterior cerebral artery were discolored white and had a rough surface. More surprisingly, they significantly reduced in their outer diameters (Fig. 14.1). During surgery, Prof. Kamiyama and I were so surprised and excited by these findings that I still remember it as if it were yesterday. Since that day, my belief that the arteries involved by moyamoya disease would carry the pathophysiology completely different from that of atherosclerosis remains unchanged. Unfortunately, we did not publish the findings during surgery in this case. Therefore, when we published a review article on moyamoya disease in *Lancet Neurology*, we had the opportunity to report the first-ever direct observation of similar findings during clipping and bypass surgery for another case of moyamoya disease complicated by anterior communicating aneurysm [3].

On the other hand, the advances in MRI technology have made it possible to visualize these findings in moyamoya disease non-invasively in the last decade. Most of these findings have been obtained by heavy T2-weighted images in MRI. Typically, T2-weighted images are captured with a repetition time of 3000–5000 msec, echo time of 80–120 msec, and imaging time of 2–4 minutes using the fast spin-echo (fast SE or turbo SE) method. On heavy T2-weighted images, however, the contribution of T2 components is larger and the water signal

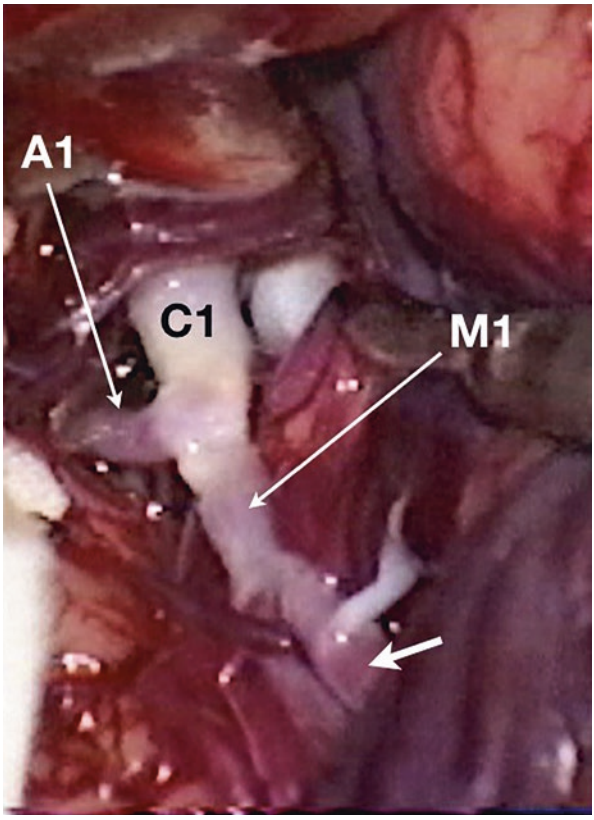

Fig. 14.1 Intraoperative photograph demonstrated that the internal carotid artery (C1) and the proximal portion of the middle cerebral artery (M1) and the anterior cerebral artery (A1) were discolored white and had a rough surface. Their outer diameter was much smaller than usual. Note that the middle cerebral artery distal to the anterior temporal artery showed normal appearance (arrow)

is further enhanced than T2-weighted images, resulting in an image with a good signal-to-noise (S/N) ratio. In contrast, the structures other than water are expressed as low signal intensity, making it extremely suitable to visualize the anatomical relationship between the water and adjacent structures. Therefore, heavy T2-weighted image is recognized suitable to visualize the luminal structures of the cochlea and salivary ducts. In the field of neurosurgery, heavy T2-weighted images have been also proved beneficial in understanding the anatomical relationship between the cranial nerves and offending vessels when performing microvascular decompression surgery for facial spasm, trigeminal neuralgia, and glossopharyngeal neuralgia. As the results, heavy T2-weighted image is also called as "MR hydrography" or "MR cisternography" [4].

Kaku et al. (2012) first reported that the outer diameter of ICA and M1 segment is significantly smaller in moyamoya disease than in the control and in M1 stenosis or occlusion, using 3D-CISS (3-dimensional constructive interference in steady state), one of heavy T2-weighted imaging technique. For example, the outer diameter of the horizontal portion of middle cerebral artery (M1) was 1.9 ± 0.4 mm in moyamoya disease, being significantly smaller than in M1 stenosis/occlusion (3.5 ± 0.6 mm, $P < 0.01$) and the controls (3.3 ± 0.5 mm, $P < 0.01$). This is the first study that denotes arterial shrinkage in moyamoya disease. However, this study included a significant number of pediatric patients, and thus may underestimate the outer diameter in moyamoya disease because the vessel size is smaller in children than in adults [5]. Using high-resolution MRI with a 3.0-Tesla MR apparatus, Kim et al. (2013) compared the outer diameter of the M1 portion between moyamoya disease ($n = 12$) and intracranial atherosclerotic disease (IAD; $n = 20$). They found that the outer diameter was significantly smaller in moyamoya disease (1.6 ± 0.4 mm) than in IAD (3.0 ± 0.5 mm, $P < 0.0001$) [6]. Ryoo et al. (2014) also reported similar results. Thus, they calculated a remodeling index as the ratio of vessel area at MCA to the reference vessel, and found that the value was significantly smaller in moyamoya disease than in IAD (0.19 ± 0.11 vs. 1.00 ± 0.43, $P < 0.001$) [7].

Using 3D-CISS technique, we quantified the outer diameter of the terminal portion of internal carotid artery (C1), the M1 portion, and the horizontal portion of anterior cerebral artery (A1) in 64 adult patients with moyamoya disease. As the results, the outer diameter was 2.3 ± 0.7 mm, 1.3 ± 0.5 mm, 1.0 ± 0.4 mm in the C1, M1, and A1 portions, respectively (Fig. 14.2). All values were significantly smaller than those in both M1 stenosis ($n = 6$) and the healthy controls ($n = 17$). On the other hand, the outer diameter of the basilar artery did not differ among 3 groups (Fig. 14.3). More importantly, there was a negative correlation between their outer diameters and Suzuki's angiographical stage ($P < 0.001$). In other words, the outer diameter of the affected arteries was found to decrease as the stage of the disease progressed. The finding strongly suggests that the outer diameter of carotid fork gradually decreases in parallel to disease progression (Fig. 14.4) [8]. In this study, we also evaluated the ipsilateral-to-contralateral ratio of the outer diameter of carotid fork in 5 children and 15 adults with "unilateral" moyamoya disease, because the values in the contralateral (*normal*) side are useful as the internal standards to prove the phenomenon of arterial shrinkage in moyamoya disease. As the results,

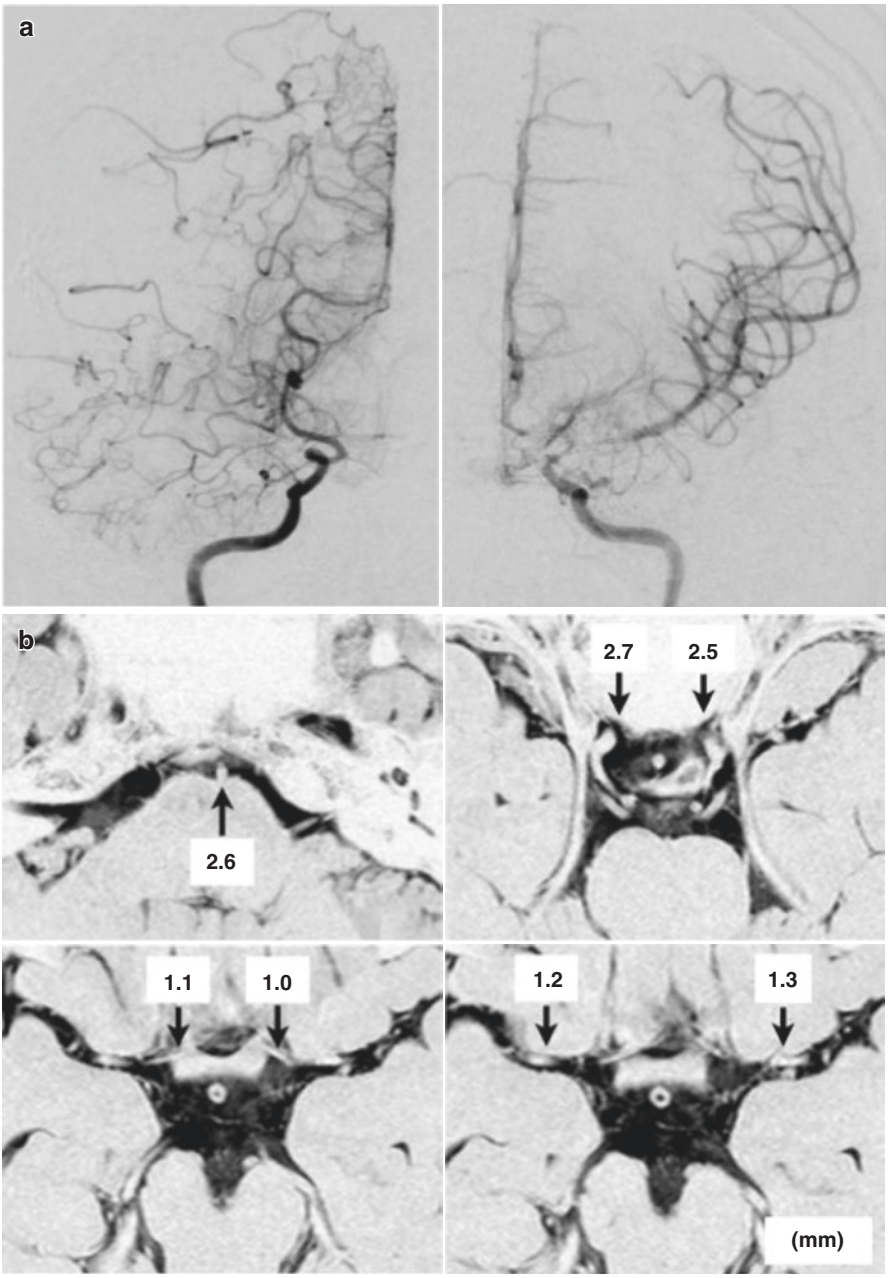

Fig. 14.2 Radiological findings in a 28-year-old female who experienced TIA due to moyamoya disease. Cerebral angiography (**a**) shows typical findings of moyamoya disease. On the other hand, 3D CISS images (**b**) demonstrate a marked reduction of the outer diameter of the terminal portion of the internal carotid artery (2.7 and 2.5 mm), horizontal portion of middle cerebral artery (1.2 and 1.3 mm) and anterior cerebral artery (1.1 and 1.0 mm) on the right and left side, respectively. In contrast, the outer diameter of the basilar artery was within normal limit (2.6 mm)

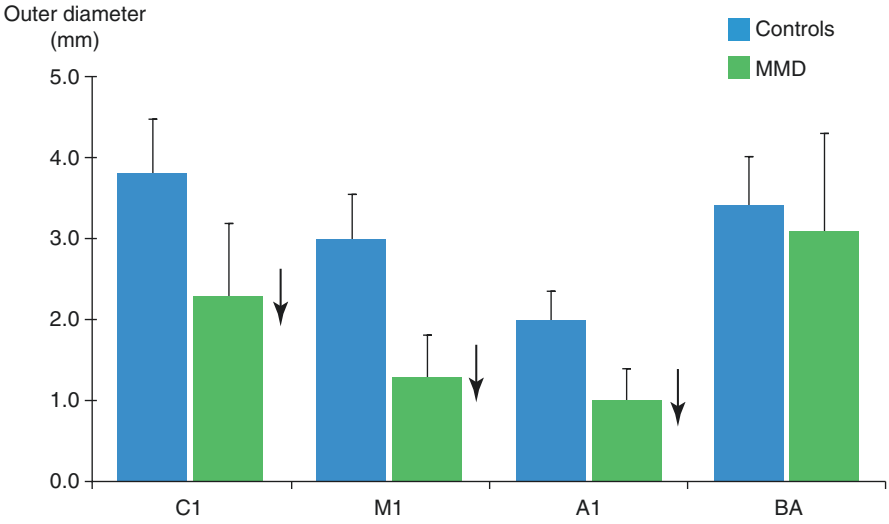

Fig. 14.3 A bar graph demonstrates a mean value and standard deviation of the outer diameter in the terminal portion of the internal carotid artery (C1), the horizontal portion of the middle cerebral artery (M1) and anterior cerebral artery (A1), and basilar artery (BA). Note that the outer diameter of the C1, M1, and A1 portion, but not of the BA, is significantly smaller in moyamoya disease than in the controls

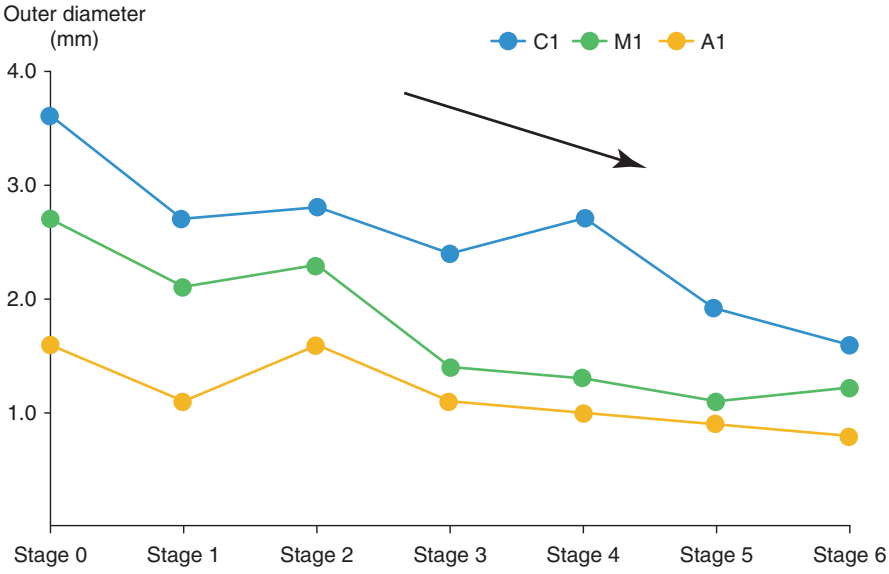

Fig. 14.4 A line graph shows that the mean value of outer diameter of the C1, Ma, and A1 portion gradually decreases in parallel to disease progression

the ratio was 0.68 ± 0.13 and 0.54 ± 0.15 in the C1 and M1 portion, respectively. The values were significantly smaller in unilateral moyamoya disease than in M1 stenosis (1.01 ± 0.09 and 1.06 ± 0.06, respectively) [8].

Subsequently, we also longitudinally evaluated the outer diameter of the involved arteries in non-operated 8 hemispheres that exhibited spontaneous disease progression during follow-up periods. Of these, 7 hemispheres were categorized into Suzuki's Stage 1–3 at initial presentation. In these 7 hemispheres, the outer diameter of the C1, M1, and A1 portion significantly decreased in parallel with subsequent disease progression to Suzuki's Stage 3–6. More interestingly, the outer diameter of the affected arteries was found to decrease with the progression of the disease stage and then continue to shrink progressively over the next 3–12 months. This longitudinal study could first prove that the outer diameter of involved arteries progressively decreases in moyamoya disease [9]. Therefore, progressive shrinkage in parallel with disease progression was confirmed in both cross-sectional study and longitudinal study. Subsequently, several investigators have shown this phenomenon in both Asian and non-Asian patients with moyamoya disease [10–13].

The number of reports on direct observation of the affected arteries during cerebral revascularization surgery for moyamoya disease is very limited, but the findings also support the above-mentioned radiological findings. In addition to the quantification of the outer diameters of C1, M1, and A1 portions using 3D-CISS as described above, we directly observed the affected arteries during surgical revascularization in three cases of adult moyamoya disease [8]. The findings were very similar to our previous observation (*see above*). Thus, the terminal portion of the internal carotid artery, as well as the horizontal portions of the middle cerebral artery and the anterior cerebral artery, was found to be white and their surface was irregular. Most notably, they significantly reduced in diameter in all cases (Fig. 14.5). For example, the outer diameter of C1 portion was 1.2 to 1.7 mm, being much smaller than normal controls (4.2 mm). Likewise, the outer diameter of M1 portion was much smaller in moyamoya disease than in normal controls (0.7–1.4 mm vs. 3.5 mm, respectively) [14].

In summary, the radiological and intraoperative findings over the last several years have revealed that not only does the lumen of the affected arteries narrow, but the outer diameter of the artery gradually shrinks as the disease progresses in moyamoya disease. This phenomenon is not observed in intracranial arterial stenosis due to atherosclerosis, and is believed specific to moyamoya disease at least nowadays. It has long been known that atherosclerosis leads to not only the luminal narrowing but also abnormal dilation of the entire artery, which is called positive or expansive remodeling. Therefore, negative or constrictive remodeling observed in moyamoya disease would be the completely opposite phenomenon. Furthermore, recent high-resolution MRI studies have shown that atherosclerosis causes eccentric lesion in the intracranial arteries, whereas moyamoya disease causes concentric lesions at the site of the stenosis [12, 15]. These observations strongly suggest that intracranial artery stenosis due to atherosclerosis and moyamoya disease is based on completely different pathophysiology (Fig. 14.6).

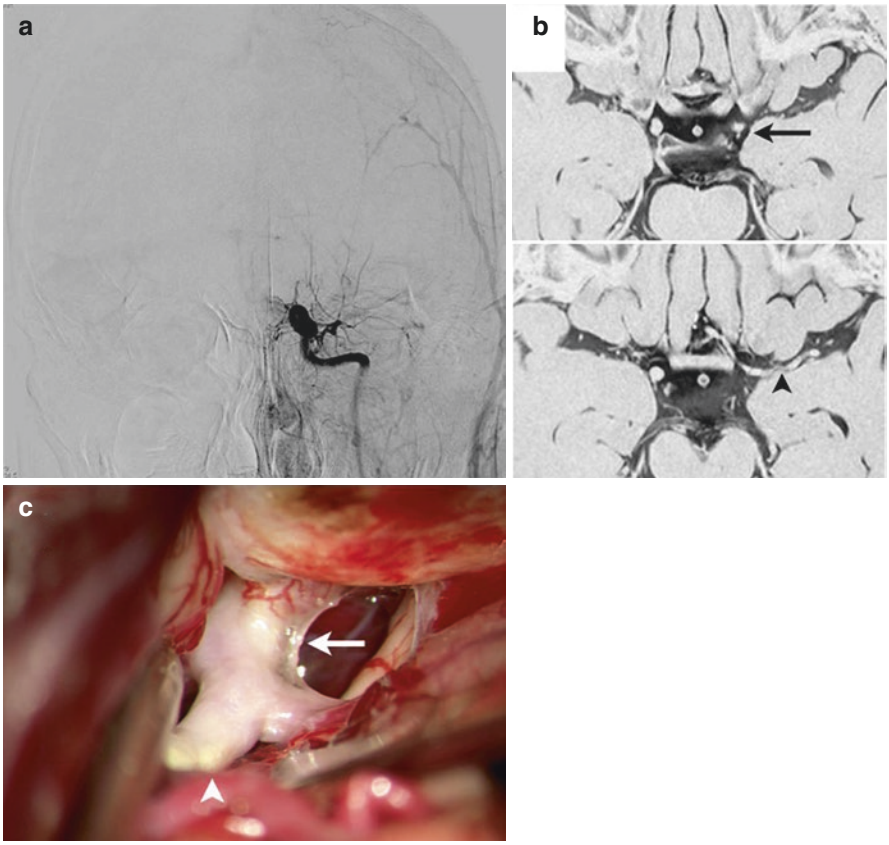

Fig. 14.5 Radiological and intraoperative findings of a 52-year-old patient with unilateral moyamoya disease on the left side. Left carotid angiography (**a**) demonstrates severe stenosis in the supraclinoid portion of the internal carotid artery. 3D CISS images (**b**) show a marked reduction of the outer diameter in the C1 (arrow) and M1 portion (arrowhead) on the left side. Intraoperative photograph during surgical revascularization onto the left hemisphere (**c**) reveals that the C1, M1, and A1 portion are discolored white and are markedly shrinked

It is well known that the intima of the affected arteries becomes thickened, while the tunica media become thinner in moyamoya disease. These pathological observations may be able to explain the mechanism of the reduction of the outer diameter of the affected arteries in moyamoya disease. Unique pathological changes such as duplication and waving are also known to occur in the elastic lamina of the affected arteries in moyamoya disease. However, the underlying mechanism has long been undetermined. If arterial shrinkage occurs in affected arteries of moyamoya disease, it may provide a clear explanation for these pathological changes in the elastic lamina as a secondary effect (*see Chap. 1*). In other words, moyamoya disease *per se* may be pathognomonically defined by both luminal stenosis and arterial shrinkage of the involved carotid forks with a progressive fashion (Fig. 14.6).

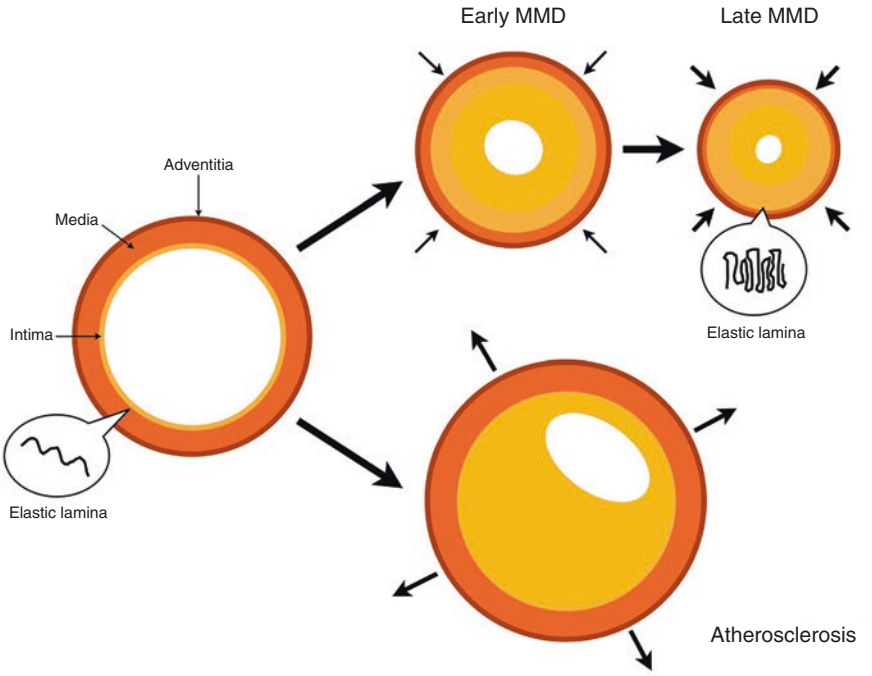

Fig. 14.6 A diagram shows the difference in pathophysiology between moyamoya disease (MMD) and atherosclerosis. Moyamoya disease causes concentric intimal thickening, medial atrophy, and the waving of the elastic lamina, leading to arterial shrinkage (constricting remodeling), but atherosclerosis causes eccentric intimal thickening and expansive remodeling

14.3 Arterial Shrinkage in Posterior Cerebral Artery

The posterior cerebral artery (PCA) is one of the important collateral routes in moyamoya disease. The PCA provides collateral blood flow to the frontal, temporal, and parietal lobes through leptomeningeal anastomosis, posterior pericallosal artery, and posterior choroidal artery. However, the PCA is also known to be involved in about 30% of patients with moyamoya disease. The stenotic lesions usually develop in the P2 or P3 segment of the PCA. Therefore, the occurrence of PCA involvement can easily cause ischemic symptoms not only in the occipital lobe, where the PCA essentially provides blood flow, but also in the parietal and temporal lobes, which receive collateral blood flow from the PCA. Clinical symptoms widely vary, including homonymous hemianopsia, headache attack in the temporal area, sensory aphasia, alexia, agraphia, and numbness of the contralateral extremities. However, there were no studies that denote whether arterial shrinkage occurs in the PCA. Therefore, we analyzed the outer diameter of the P2 portion of PCA in 72 patients with moyamoya disease. In this study, the stenotic lesion of the PCA was divided into 3 grades; Grade 0 (normal), Grade 1 (stenotic), and Grade 2 (occluded) on MR angiography. As the

results, we found that the outer diameter of PCA progressively decreased as the stenotic lesion occurred in the PCA. That is, the values for Grade 1 cases fell to about 75% of Grade 0 cases, and the values for Grade 2 cases were even smaller, falling to about 45% of Grade 0 cases. The results were similar for children and adults. The phenomenon was observed in both pediatric and adult patients. In addition, we could follow the PCA outer diameter over time during the progression of PCA disease in 4 hemispheres in 2 pediatric cases, and found that the PCA outer diameter also decreased with the progression of the disease. Although there are almost no studies that denote pathological findings of the stenotic lesions in the PCA, these radiological observations strongly suggest that similar pathological changes occur in stenotic lesions in PCA as in carotid fork. This fact fits well with the embryological knowledge that arteries more distal to the P2 portion of the PCA arise from the cranial ramus of the primitive ICA, as well as the MCA and ACA [16].

14.4 Arterial Shrinkage in Moyamoya Syndrome

The definition of moyamoya disease is rather complicated. The criteria for diagnosis are unique in that they have been based solely on information about the lumen of the affected artery on cerebral angiography and MR angiography for more than 50 years. When the disease was first discovered, moyamoya disease was defined as a disease of unknown etiology, and therefore, the patients with the same radiological findings but concurrent diseases have been diagnosed with moyamoya syndrome or quasi-moyamoya disease in children and adults, bilaterally or unilaterally. Hayashi et al. (2014) reported that about 5% of patients with moyamoya disease are diagnosed as having moyamoya syndrome [17]. Previously reported concurrent diseases are listed in Chap. 2. However, it is unclear from previous studies whether moyamoya syndrome is a completely different disease from moyamoya disease, or whether it is simply a combination of moyamoya disease and concurrent disease. Therefore, we hypothesized that if shrinkage in the affected arteries is specific to moyamoya disease, then it would be very useful to investigate whether this arterial shrinkage also occurs in moyamoya syndrome or not. In this study, we evaluated the outer diameter of the carotid forks in 9 patients with moyamoya syndrome. Concurrent diseases included neurofibromatosis 1 (NF1) in two cases, idiopathic thrombocytopenia (ITP) in one, autoimmune disease such as hyperthyroidism in 3, atherosclerosis in 3. Very interestingly, the outer diameter of the affected arteries markedly differed from the usual moyamoya disease, and there was a great deal of inter-case variability. In 7 of the 9 patients, as in the case of moyamoya disease, the diameter of C1 and M1 decreased as the disease progressed. In these 7 patients, the comorbidities included NF1 ($n = 2$), ITP ($n = 1$), autoimmune disease ($n = 2$), and atherosclerosis ($n = 2$). On the other hand, there was no reduction in the diameter of the affected arteries in the other two patients, both of whom were judged to have Stage 3 disease on cerebral angiography. Their comorbidities were autoimmune disease ($n = 1$) and atherosclerosis ($n = 1$) [18].

These observations strongly suggest that moyamoya syndrome (or quasi-moyamoya disease) is not a uniform disease entity, but can be divided into two pathophysiologically different disorders: the arterial shrinkage group and the non-arterial shrinkage group. The former is considered closely related to definitive moyamoya disease, but the latter may be essentially analogous to atherosclerosis, being divergent from definitive moyamoya disease. In more detail, there may be a significant number of cases that have exactly the same etiology as definitive moyamoya disease but are not diagnosed as moyamoya syndrome because they happen to have comorbidities. On the other hand, there may be some cases that are pathogenetically unrelated to moyamoya disease but are categorized as moyamoya syndrome because of both radiological findings similar to those of moyamoya disease and comorbidities. Anyway, such a distinction would have been highly unlikely when the diagnosis was based solely on information about the lumen of the involved arteries using cerebral angiography and MR angiography. This distinction was first made possible by observing the outer diameter of the involved arteries using heavy T2-weighted images. However, these findings are based on the study with a small sample size. Therefore, multi-center investigations with larger cohorts are warranted to further clarify the pathophysiology and disease entity of moyamoya syndrome [18].

14.5 Clinical Significance of Arterial Shrinkage in Moyamoya Disease

14.5.1 Endovascular Treatment

There are several reports attempting to dilate the stenotic lesions themselves with endovascular techniques for moyamoya disease. Rodruguez et al. (2007) reported that balloon angioplasty in adult patients with early moyamoya disease resulted in effective dilation of stenotic lesions [19]. Several groups of investigators also successfully performed balloon angioplasty and stent placement in children and adults with moyamoya disease or moyamoya syndrome [20–23].

However, Khan et al. (2011) analyzed clinical results of endovascular treatment for 5 patients with moyamoya disease. All of them repeated ischemic attacks after 0–4 months after endovascular treatments. Follow-up angiography revealed severe in-stent restenosis or occlusion in 4 of 5 patients. Finally, all of them underwent surgical revascularization to resolve ischemic attacks [24]. More importantly, Eicker et al. (2011) reported a 18-year-old cases that developed very massive subarachnoid hemorrhage several hours after stent placement of the supraclinoid internal carotid artery [25]. Considering the specific occurrence of arterial shrinkage in moyamoya disease, we should remind that these endovascular procedures may carry the risk of disruption of the affected artery, leading to life-threatening subarachnoid hemorrhage.

14.5.2 Differential Diagnosis of Moyamoya Disease

As aforementioned, it is not rare to encounter the difficulty to distinguish moyamoya disease from atherosclerosis-related intracranial arterial stenosis especially in elder patients. However, heavy T2-weighted image is quite useful to accurately identify moyamoya disease even in very elder patients (Fig. 14.7). This 80-year-old

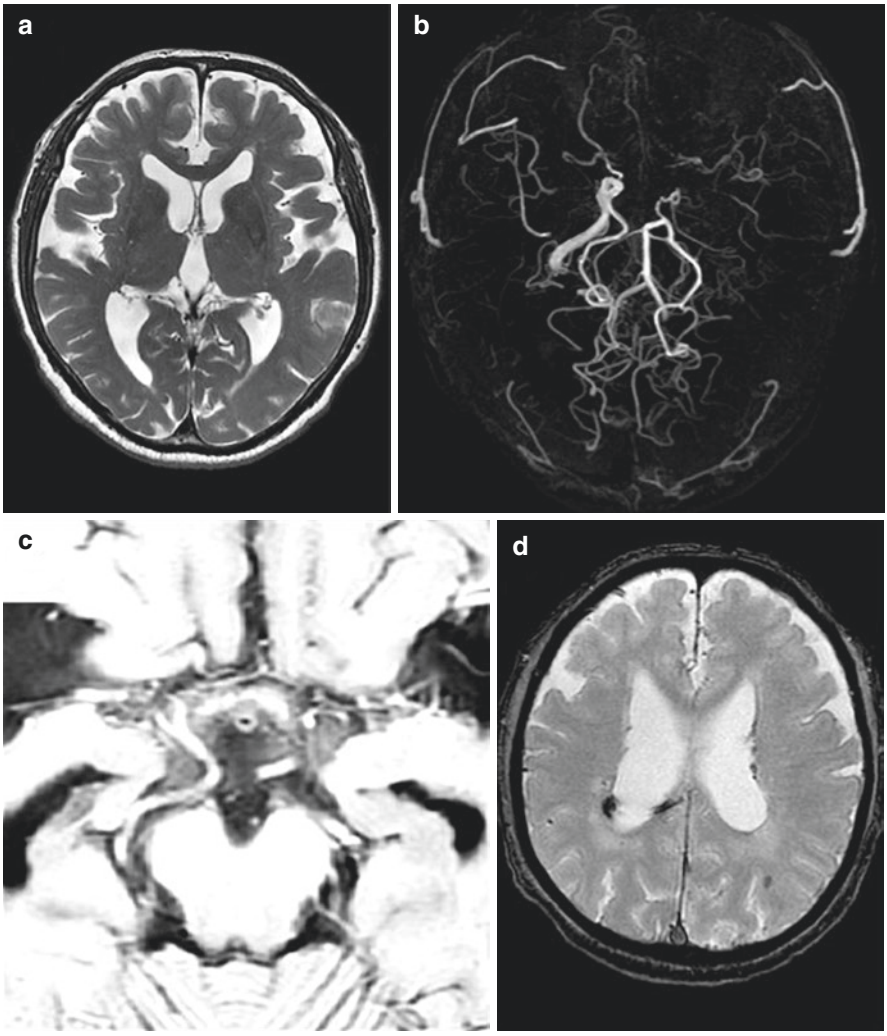

Fig. 14.7 Radiological findings of a 80-year-old male with moyamoya disease. T2-weighted image (**a**) demonstrates no parenchymal lesions, but MR angiography (**b**) reveals complete occlusion of the internal carotid artery at the supraclinoid portion on the right side and at the origin on the left side. 3D CISS image (**c**) reveals a marked arterial shrinkage in the carotid fork on both sides. Therefore, he was diagnosed as moyamoya disease. T2*-weighted image taken 6 months later (**d**) shows multiple bleeding in the subependymal layer of the lateral ventricle on both sides

male suddenly developed syncope and was referred to our hospital. T2-weighted MRI demonstrated no parenchymal lesions, but MR angiography revealed complete occlusion of the internal carotid artery at the supraclinoid portion on the right side and at the origin on the left side. At first, he was diagnosed as atherosclerotic carotid occlusion on both sides, but subsequently performed 3D CISS revealed a marked arterial shrinkage in the carotid fork on both sides. Finally, he was diagnosed as moyamoya disease and was conservatively followed up because of his high age. He was admitted to our hospital again because of sudden consciousness disturbance 6 months after the initial diagnosis. T2*-weighted image showed multiple bleeding in the subependymal layer of the lateral ventricle on both sides, which is the finding that strongly supports previous diagnosis of moyamoya disease.

As this case clearly demonstrates, cerebral angiography and MR angiography, which have been used to diagnose moyamoya disease, only provide information on the lumen of the affected arteries, making it difficult to exclude other diseases such as atherosclerosis. This fact has led to a great deal of confusion in routine clinical practice and research [26]. Therefore, we the Research Committee on Moyamoya Disease in Japan is preparing to revise the diagnostic criteria for moyamoya disease to include heavy T2-weighted image findings in addition to cerebral angiography and MR angiography.

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