—Case Reports—

Shaggy Aorta Syndrome after Cerebral Angiography: A Case Report

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Shaggy aorta refers to an aorta with intimal roughening due to atheromatous aortic plaques. Catheterization and anticoagulation therapy can result in cholesterol emboli, potentially leading to systemic organ infarction. Contrast-enhanced computed tomography (CT) and transesophageal echocardiography are commonly used to diagnose shaggy aorta. A patient in his ninth decade of life had a history of right occipital lobe ischemic stroke, bilateral internal carotid artery stenosis, and shaggy aorta syndrome related to transfemoral cerebral angiography. Dysarthria occurred immediately after the procedure. Brain magnetic resonance imaging (MRI) confirmed cerebral infarction, and anticoagulant therapy was administered. Four days later, after observing numbness of the left 5th finger and purplish discoloration of the tips of the 2nd and 5th fingers, we performed contrast-enhanced CT and diagnosed shaggy aorta. There was no renal impairment or eosinophilia and the patient was discharged 16 days after the examination. Aortic MRI performed 1 month later revealed an unstable plaque in the vessel wall. Although we report our experience with a single patient, we recommend that patients scheduled for cerebral angiography, especially those with severe arteriosclerosis, undergo preprocedural aortic fast spoiled gradient echo MRI screening to avoid shaggy aorta syndrome. (J Nippon Med Sch 2025; 92: 409–413)

Key words: shaggy aorta, cerebral angiography, aortic MRI, screening, cholesterol embolism

Introduction

Percutaneous endovascular treatments for aortic, cardiac, and cerebrovascular diseases are now the standard; thus, careful evaluation of the vessel wall is important[1]. Shaggy aorta syndrome refers to diffuse aortic atherosclerosis that results in feathering of the aortic lining[2]. Catheterization and anticoagulation treatment can disintegrate and disperse fine cholesterol crystals that can embolize systemic organs in patients with atheroma. The prognosis for patients with organ damage due to shaggy aorta is poor, although the 1-year mortality rate of 64-87%[3] can be lowered to 23% with multidisciplinary treatment[4]. The cause of shaggy aorta syndrome is idiopathic in approximately 21% of cases and iatrogenic in 79% of cases. Catheter manipulation, anticoagulation therapy, and aortic surgery may lead to shaggy aorta syndrome, the incidence of which has increased with the greater use of endovascular procedures[5]. Although it may be beneficial to screen for shaggy aorta before angiography, no study has tested this hypothesis.

Nakayama et al.[6], Hagiwara et al.[7], and Kikuno et al.[8] reported patients with cholesterol embolism elicited by cerebral angiography. The present report describes a patient with shaggy aorta syndrome after cerebral angiography who subsequently developed a cerebral infarct and skin symptoms. Aortic MRI was later performed and showed intra-plaque hemorrhage, which was caused by the shaggy aorta syndrome. Our experience suggests the MRI screening before angiography is beneficial.

Case Report

A man in his eighties was receiving medication for hypertension, diabetes mellitus, and dyslipidemia. Antiplatelet medication was started for an atherothrombotic

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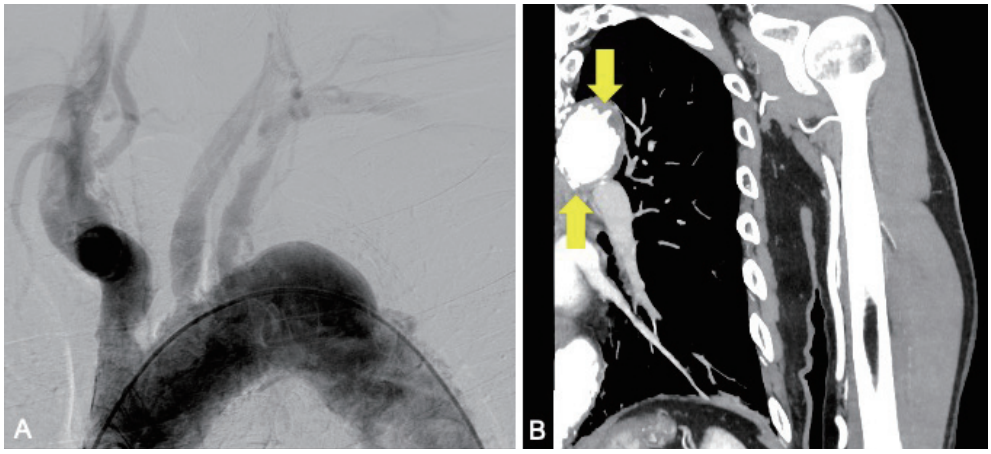

Fig. 1 Aortography and contrast-enhanced CT
(A) Aortograph showing numerous irregularities in the vascular wall.
(B) Contrast-enhanced CT (coronal image) showing a roughened aortic intima (arrows) due to atheromatous aortic plaques.

cerebral infarct that had developed 5 months earlier and elicited dyslexia. Carotid artery (CA) echography revealed bilateral internal CA stenosis. On admission to our hospital, the patient was alert and had no abnormal neurological findings, and his dyslexia was improved.

To determine the degree of CA stenosis without the influence of CA calcification and to evaluate intracranial collateral circulation, we obtained cerebral digital subtraction angiographs through the right femoral artery. The initial aortogram revealed multiple wall irregularities in the aortic arch and subclavian artery, which confirmed a diagnosis of shaggy aorta (**Fig. 1A**). A catheter was inserted into the bilateral common carotid and left subclavian artery, and cerebral angiography was completed. The patient reported dysarthria immediately thereafter, and minor left-sided facial paresis was confirmed. Cerebral infarction was suspected and heparin (4,000 IU) was injected intravenously. Later, continuous heparin infusion (1,000 IU/day) was started. Brain MRI showed a punctate acute cerebral infarct in the right cerebral hemisphere (**Fig. 2A**) and edaravone infusion was started.

On the next day, his dysarthria had improved but the mild left facial paresis persisted. Two days after angiography, the patient's condition was stable, but brain MRI showed worsening of cerebral infarction in the left occipital lobe and right frontal lobe (**Fig. 2B**). We discontinued heparin and started intravenous sodium ozagrel and oral cilostazol (200 mg twice daily) for suspected shaggy aorta or A-to-A embolism from the brachiocephalic artery plaque. Heparin was stopped because anticoagulant therapy was likely an exacerbating factor at this point. We did not suspect heparin-induced thrombocytopenia (HIT) because blood tests showed no decrease in platelet count; no HIT antibody test was performed[9]. The cerebral infarct did not increase (**Fig. 2C**). At 4 days post-angiography, the patient developed numbness of the left 2nd and 5th fingers and dark purple discoloration of the fingertips. A contrast-enhanced CT scan of the upper extremity showed no abnormalities but revealed a large extensive plaque within the aorta, leading to a diagnosis of shaggy aorta (**Fig. 1B**). The clinical course confirmed this diagnosis, and oral administration of beraprost sodium was started. Because the symptoms involving the fingers were mild, no skin biopsy was obtained. In the absence of renal dysfunction no additional blood tests were performed. Fingertip color improved, and the patient was discharged to home 16 days post-angiography.

One month later, aortic fast spoiled gradient echo (FSPGR) MRI showed multiple intra-aortic hyperintensities suggestive of intra-plaque hemorrhage (**Fig. 3**). During 9 months of follow up, the patient's symptoms did not worsen.

Discussion

Overview of Shaggy Aorta Syndrome

The clinical signs of shaggy aorta syndrome vary: approximately 50% of patients have renal dysfunction, skin symptoms are observed in 35-96% of patients, and neurological symptoms are present in 4-23% of patients. Patients may experience transient ischemic attacks, cerebral infarcts, disturbance of consciousness, amaurosis, and paralysis[5]. For diagnosis, it is useful to confirm the presence of cholesterol crystals in a biopsy specimen of the embolized organ; skin biopsies are diagnostic in 92% of pa-

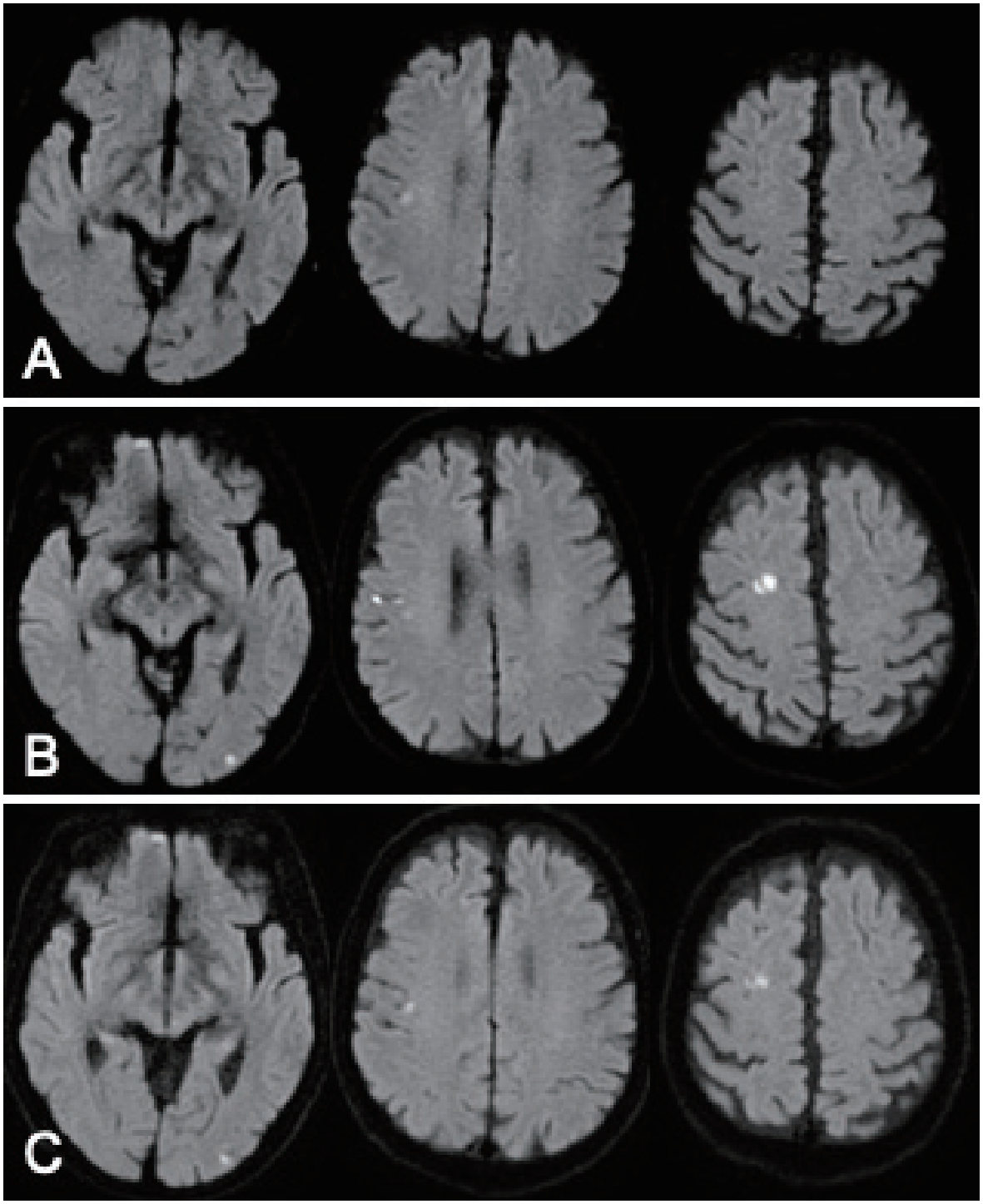

Fig. 2 Diffusion-weighted brain MRI scans (axial images).
(A) Scans on the day of admission showing a small cerebral infarct in the right frontal lobe.
(B) Scans 2 days later showing new cerebral infarcts in the right frontal lobe and left occipital lobe.
(C) Scans 5 days later showing no new cerebral infarcts.

tients[10]. Blood tests confirm eosinophilia in 80% of patients, and increased blood IL-5 levels and hypocomplementemia also suggest an immunologic mechanism[11–13].

Our patient was diagnosed as having shaggy aorta syndrome that developed after cerebral angiography. Cerebral infarction and peripheral vascular embolism of the fingers of the left hand were observed. However, because the finger symptoms were mild, no biopsy was performed.

Hasegawa et al.[14] and Minatohara[15] reported that corticosteroids, plasmapheresis, and prostaglandin preparations were useful treatments for shaggy aorta syndrome

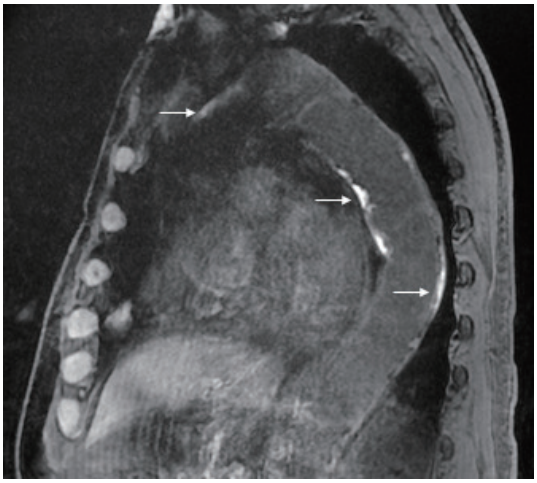

Fig. 3 Aortic FSPGR MRI
Aortic FSPGR MRI scan revealing multiple hyperintense plaques (arrows) in the ascending to descending aorta.

and stressed that triggers such as catheterization and anticoagulants must be avoided. We suspect catheter manipulation and anticoagulant therapy triggered symptoms in our patient. Patients with atherothrombotic cerebral infarction often undergo cerebral angiography to diagnose a lesion. Our experience with the present patient suggests that 3D CT angiography (CTA) should have been performed before cerebral angiography.

Preventing Shaggy Aorta Syndrome

To minimize complications related to shaggy aorta syndrome, at-risk patients should undergo prompt imaging assessment[1,16]. CT is important in the diagnosis, risk stratification, and management of aortic diseases, e.g., aortic dissection and intramural hematoma[17,18]. However, the accuracy of CT for ulcerating plaques and thrombi is unknown[19]. Shaggy aorta syndrome is diagnosed by contrast-enhanced CT scanning or transesophageal echocardiography (TEE). Fragile atheromatous lesions measuring ≥5 mm are visualized as ulcerative, possibly invasive lesions[20,21]. Aortic magnetic resonance angiography (MRA) can display blood vessel walls and thrombi without radiation exposure, and phase-contrast MRI scans can be used to analyze blood[22]. MRA is now almost as accurate as CTA for diagnosing aortic diseases[23]. Harloff et al.[24,25] reported that MRI is better than TEE for detecting aortic plaques.

In our patient, aortic MRA performed after the onset of shaggy aorta syndrome was useful for assessing the vessel wall. Although an MRI finding of intraplaque hemorrhage is not sufficient to diagnose shaggy aorta syndrome, atherosclerotic changes alert to its risk. MRI findings did not support a diagnosis of shaggy aorta in our patient; thus, additional studies are required in such cases.

Conclusion

Our experience treating a man who developed shaggy aorta syndrome after cerebral angiography suggests that aortic FSPGR MRI screening before cerebral angiography might benefit patients at risk of shaggy aorta. However, this recommendation requires confirmation in future studies.

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