



## Internal Medicine Flashcard

## An unusual cause of intracranial hemorrhage

Yusaku Kajihara

Department of Gastroenterology, Fuyoukai Murakami Hospital, 3-3-14 Hamada, Aomori 030-0843, Japan



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## 1. Case description

A 39-year-old woman with no significant medical history was admitted in the emergency room with loss of consciousness. Computed tomography revealed right-sided putamen hemorrhage (Fig. 1, Panel A). The patient had no risk factors for intracranial hemorrhage, including hypertension, older age, alcohol intake, and dyslipidemia. Cerebral angiography showed steno-occlusive changes at the terminal internal carotid artery (ICA) portion bilaterally, with the development of collateral circulation and no aneurysm (Fig. 1, Panels B and C). What is the diagnosis?

## 2. Discussion section

Moyamoya disease (MMD) was diagnosed by the abovementioned angiographic findings. MMD is a poorly understood occlusive disease involving large intracranial arteries, especially the distal ICA and the stem of the middle cerebral artery and anterior cerebral artery. The lenticulostriate arteries develop a rich collateral circulation around the occlusive lesion, which gives the impression of a “puff of smoke” (*moyamoya* in Japanese) on conventional x-ray angiography.

MMD has several unique clinical features, which include two peaks of age distribution at 5 years and at about 40 years [1]. The main

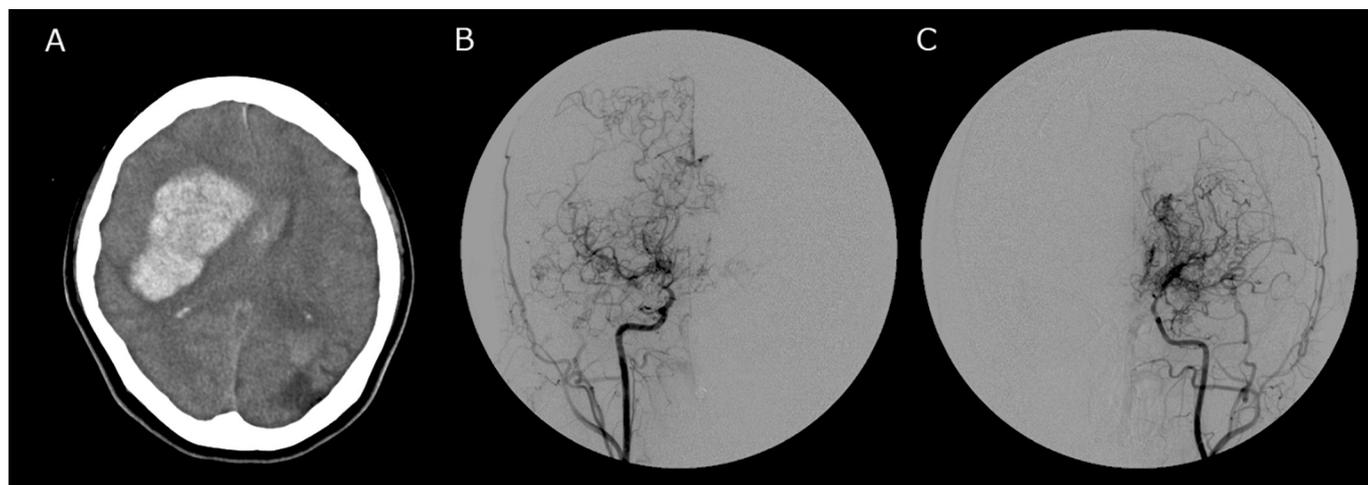


Fig. 1. Panel A: Computed tomography revealing right-sided putamen hemorrhage. Panels B and C: Cerebral angiography showing steno-occlusive changes at the terminal internal carotid artery portion bilaterally, with the development of collateral circulation.

E-mail address: [yukajihara-gi@umin.ac.jp](mailto:yukajihara-gi@umin.ac.jp).

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clinical manifestations of MMD are stroke and hemorrhage; unlike pediatric patients, who usually present with transient ischemic attacks or cerebral infarction, almost one-half of adult patients have intracranial hemorrhage that seriously affects their prognosis [2]. Extracranial-intracranial bypass surgery, which is often used for ischemic MMD, has preventive effects for adult patients with hemorrhagic MMD [2].

The incidence of MMD is high in East Asia, such as Japan [1]. In Europe, the incidence of MMD has been estimated approximately one-tenth of that in Japan [3]. MMD is a chronic progressive cerebrovascular disease and can cause permanent neurologic impairment. Therefore, early diagnosis and prompt treatment of MMD are important.

### Conflict of interest

None declared.

### References

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