

Moyamoya Disease Associated with Craniopharyngioma

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Tsujii N, Kuriyama T, Iwamoto M, Shizuki K. Moyamoya disease associated with craniopharyngioma. *Surg Neurol* 1984;21:588-92.

A case of coexisting moyamoya disease and craniopharyngioma in an 8-year-old boy is reported. The patient had been suffering from recurrence of transient right hemiparesis with headache and vomiting during the three years before admission. After the diagnosis of both moyamoya disease and craniopharyngioma, evacuation of the cystic fluid of the craniopharyngioma, encephalomyosynangiosis, and internal irradiation through an Ommaya drainage system were carried out. The symptoms gradually improved after these procedures. The relation of moyamoya disease to craniopharyngioma is discussed. We consider that internal irradiation is one of the best means of treatment for a cystic craniopharyngioma associated with moyamoya disease.

KEY WORDS: Moyamoya disease; Craniopharyngioma; Internal irradiation; Intracranial occlusive disease

Moyamoya or "puff of smoke" [15] is a term derived from the appearance of the cerebral angiographic picture. Typically, there is an abnormal vascular network at the base of the brain associated with an occlusion of the circle of Willis [14]. The etiology of moyamoya disease, however, still remains controversial. Though this is generally believed to be an acquired process, familial cases have occasionally been reported [11,17,18]. There are also some other cases associated with tumor [5,8,9], irradiation [14,16], neurocutaneous syndrome [6], and arteriosclerosis. In these, angiographic findings of moyamoya vessels are the same as those without other accompanying diseases. We report a case of moyamoya disease accompanied by craniopharyngioma, and we present a review of the literature.

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Case Report

An 8-year-old boy was admitted to Wakayama Red Cross Hospital with hemiconvulsion on the right side on January 30, 1982. In the 3 years before admission, he had suffered from recurrent episodes of headache with vomiting and convulsions of the right hand, and also attacks of transient hemiparesis when he cried out.

Examination

Neurological examination on admission showed disturbance of the visual acuity of the right eye with optic atrophy, hemiparesis of the right side with hyperreflexia and pathological reflexes. Plain cranial x-ray examination disclosed a saucerlike sella turcica with suprasellar calcification (Figure 1). An electroencephalogram showed diffuse continuous high voltage delta waves, especially over the right hemisphere. Plain computer tomography (CT) showed a mild atrophy of the brain. There was a round low-density area circumscribed by a high-density zone in the sellar to suprasellar region (Figure 2). The suprasellar lesion did not show a definite contrast enhancement. Neither of the intracranial internal carotid arteries nor the middle cerebral arteries were enhanced in the suprasellar cistern or in the sylvian cisterns. An angiogram of the left cerebral artery revealed a network of numerous fine vessels (moyamoya vessels) with non-filling of the A₁ segment of the anterior cerebral artery and poor filling of the M₁ segment of the middle cerebral artery. The left internal carotid artery was narrowed and shifted laterally probably by the tumor (Figure 3). The findings in the angiogram of the right carotid artery were similar to those of the left side (Figure 4). Collateral vessels from the external carotid arteries to the intracranial region were not visualized. Endocrinologic examination revealed panhypopituitarism. Moyamoya disease associated with a craniopharyngioma was suggested by these findings.



Figure 1. Plain x-ray film of the skull (lateral view) showing a saucerlike sella with suprasellar calcification.

Operation

Biopsy of the cyst wall and evacuation of the cyst fluid were carried out through a frontotemporal craniotomy on February 10, 1982. After the frontal lobe was retracted, we saw that the right internal carotid artery was narrowed and displaced laterally by a cystic tumor. The right optic nerve was also compressed by the tumor. There were many tortuous fine vessels in the basal part of the frontal lobe. The content of the cyst was a gritty motor-oil-like fluid. The cyst was drained into the subgaleal space of the temporal region using an Ommaya drainage system. Because the superficial temporal artery was too small, superficial temporal artery–middle cerebral artery anastomosis was not performed. Instead, an encephalomyosynangiosis was carried out using the temporal muscle in order to increase blood flow and prevent ischemia of the brain [3].

Postoperative Course

After the operation, the symptoms improved gradually. However, frequent taps of the Ommaya reservoir were necessary to treat overretention of fluid causing headache and vomiting. On February 22, cystograms with metrizamide instilled into the Ommaya reservoir revealed an irregular shaped cyst (2.2 cm × 3.4 cm) in the suprasellar region (Figure 5). The volume of the cyst was calculated to be about 12 cm³, and no leakage of the dye was seen. Because the patient's liver function was abnormal as a side effect of anesthesia at this time, the internal irradiation was postponed until his liver function returned to normal. On May 11, he was given



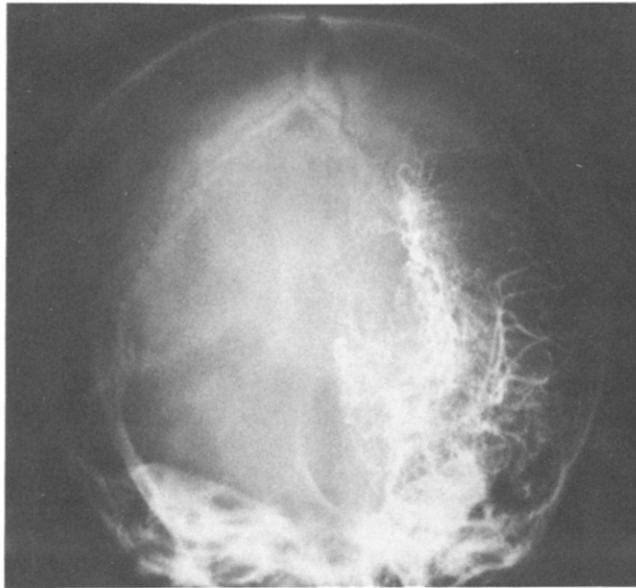
A



B

Figure 2. (A) A CT scan demonstrating mild atrophy of the brain. (B) Note a round low-density area circumscribed by a high-density zone in the suprasellar region. Neither the suprasellar lesion nor the Sylvian cisterns are enhanced by contrast medium.

intracystic administration of 1.0 mg of bleomycin daily for 5 days. On June 5, internal irradiation was performed by the injection of 10 mCi of ¹⁹⁸Au-colloid. Secretion of the fluid stopped 2 weeks after this injection. There were no side effects. The cyst did not change its size, probably because of calcification of the tumor's capsule. The patient was discharged from the hospital on June 29, 1982, having only a mild paresis of the right hand



A



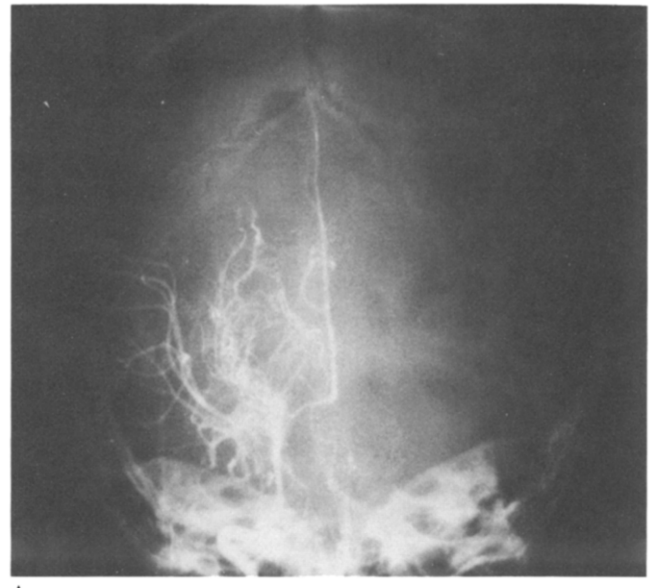
B

Figure 3. Angiogram of the left carotid artery, anteroposterior (A) and lateral (B) views, showing a fine vascular network (moyamoya vessels) with nonfilling of the A₁ segment of the anterior cerebral artery and poor filling of the M₁ segment of the middle cerebral artery.

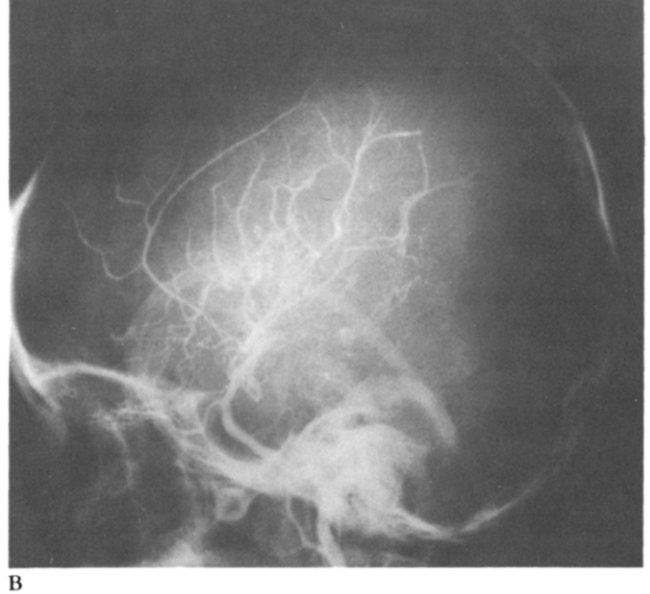
and disturbance of vision in the right eye. The patient has been healthy without aspiration of the cyst fluid to the time of writing this report, March 1983.

Discussion

The etiology of moyamoya disease has not yet been established [9]. An acquired etiology is plausible, but there are also a few reports of familial occurrence, which may suggest a congenital etiology [14]. There are some



A



B

Figure 4. Right carotid angiogram of the right carotid artery, anteroposterior (A) and lateral (B) views, demonstrating similar findings to those in the left side. Note the internal carotid artery to be compressed and displaced by the tumor.

other cases suggesting a development of collateral channels after the occlusion of the internal carotid arteries secondary to intracranial tumors, arteriosclerosis, and so on, in which angiographic findings are the same as those in so-called moyamoya disease of unknown etiology [2]. Takeuchi [19] advocates that an abnormal vascular network secondary to the known etiology (moyamoya phenomenon) should be differentiated from an idiopathic moyamoya disease.

We presented here a case of moyamoya disease associated with craniopharyngioma. The association of these

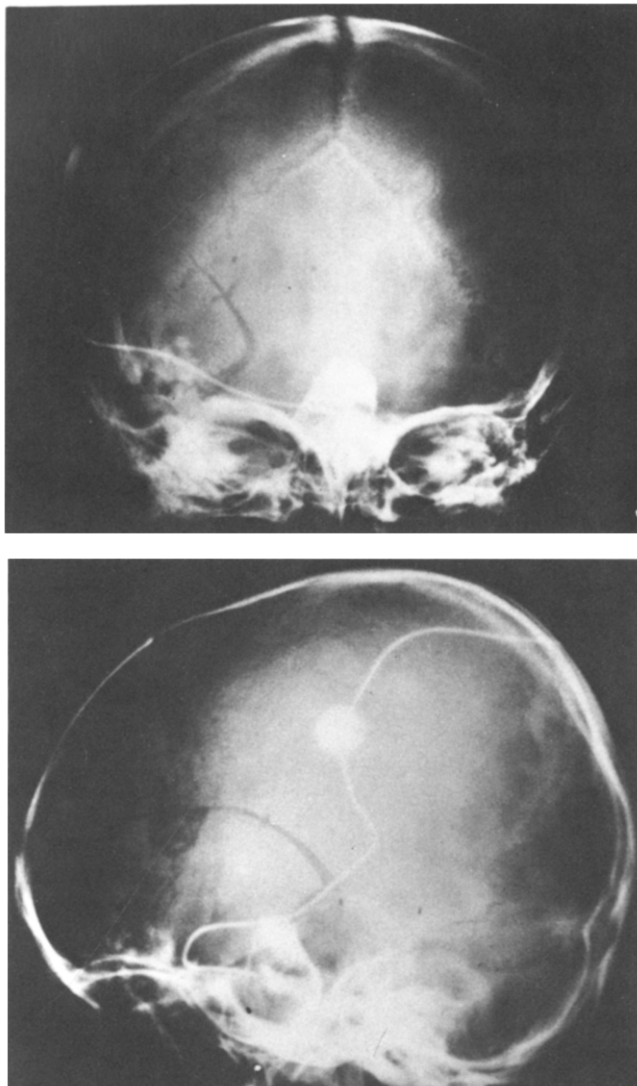


Figure 5. Cystograms showing an irregularly shaped cyst in the suprasellar region.

two lesions may be explained by coincidence or by development of collaterals secondary to stenosis of the internal carotid artery by the tumor. Only two cases of coexistence of moyamoya disease and craniopharyngioma have been reported in the literature [2,9]. Stenosis or occlusion of the internal carotid artery is a rare phenomenon in cases of intracranial tumors except for meningiomas [8] and pituitary adenomas. Such a vascular change associated with craniopharyngiomas has been reported only in three cases [5,9]. In two of these three cases, radiation therapy had been given before the appearance of the vascular change. In the other case, the moyamoya change of vessels was thought to be attributable to enlarged collateral vessels that were secondary to stenosis of the internal carotid artery due to compression by the craniopharyngioma. Mori et al [9] presented

4 cases of moyamoya vessels associated with brain tumor, and concluded that "moyamoya" disease was not an independent disease entity, but a neuroradiological syndrome caused by an occlusion of the circle of Willis and might result from a variety of causes. They also described four possible situations in which moyamoya changes might occur: compression of the carotid bifurcation by a slowly growing basal tumor; secondary arterial changes by radiotherapy of a basal tumor; in association with a neurocutaneous syndrome; and coincidence. Kojima and Waga [5] reported a case of stenosis of the internal carotid artery due to combined effects of direct compression by craniopharyngioma and radiation of the arterial wall. Kaku et al and Mizukawa et al [8] reported moyamoya disease associated with a meningioma. A case of moyamoya disease associated with glioma was reported by Pecker et al [13]. In these cases, the association was thought to be coincidence. Ishikawa et al [2] reported moyamoya vessels associated with craniopharyngioma and suggested that compression of the internal carotid artery by the craniopharyngioma was closely related to the development of collateral circulation, namely, moyamoya vessels. We suppose that the moyamoya vessels and craniopharyngioma in our case had developed independently, because the M₁ segment of the middle cerebral artery was occluded even though it was away from the tumor and received no direct compression by the tumor. However, compression by the tumor might have contributed to the development of moyamoya vessels in our case.

Kobayashi et al [4] reported that internal irradiation [1,4,7,10] was effective for cystic craniopharyngioma. This treatment is suitable for large cysts that are thought to be difficult to remove completely, for recurrent cysts resistant to previous treatment, or for multiple cysts. A safe and adequate dose to the cyst wall ranges from 9000-30,000 rads for craniopharyngiomas [4]. Retraction of the brain during an operation may injure blood vessels. Moyamoya vessels may be more vulnerable to irradiation than normal vessels. Therefore, internal irradiation should be applied to cystic craniopharyngioma associated with moyamoya disease to damage moyamoya vessels as little as possible. The Ommaya drainage system was effective in aspirating cystic fluid in the present case. This system is recommended in the treatment of a cystic craniopharyngioma associated with moyamoya disease because it is convenient for the internal irradiation and intracystic administration of bleomycin [12].

We wish to express our thanks to Dr. Norihiko Komai, Professor and Chairman, and to Dr. Seiji Hayashi, Associate Professor, both of the Department of Neurosurgery of Wakayama Medical College, for their constant guidance and kind advice.

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