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“Moyamoya” Vessels on the Tumor in the Sellar Region

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Introduction

A characteristic angiographic appearance of stenosis or occlusion of internal carotid arteries, numerous fine vascular network (so-called “moyamoya”) in the basal ganglia, and leptomeningeal and transdural anastomosis is well known as so-called “moyamoya” disease. There remains to be clarified whether this condition is an independent disease entity or a syndrome of various causes, and whether it is congenital or acquired. Kudo emphasizes unknown etiology. On the other hand, Debrun et al. reported a case of hypothalamic tumor with irradiation, in which carotid angiography disclosed the radiological picture associated with so-called “moyamoya” disease and states it a nonspecific radiological syndrome. Handa and Mori, and Mori et al. also reported the cases of tumor, irradiation and arteriosclerosis in association with angiographic findings of “moyamoya” vessels.

A rare case of intrasellar tumor with suprasellar extension, which was associated with bilateral stenosis of internal carotid arteries, “moyamoya” vessels on the tumor, leptomeningeal and transdural anastomosis, is reported.

Case report

A nine-year-old girl was admitted to Kyoto University Hospital with decreased visual acuity on the left. Since one year before admission, she had often suffered from headache, vomiting and epistaxis. Neurological examination showed blindness and optic atrophy on the left. On the right, visual acuity decreased 1.5 to 0.8 for one month and mild optic atrophy was observed. On physical examination, short stature and dry skin were noted. Vital signs were within normal limits and urinalysis was normal. No abnormalities were noted in the epipharynx. Plain skull films showed a ballooning of the sella and suture diastasis (Fig. 1). There were no calcification in and above the sella turcica. Angiographic

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study with Seldinger method was done. It was characterized by bilateral stenosis of internal carotid arteries, a fine vascular network in the suprasellar region, transdural anastomosis and displacement of dilated posterior communicating arteries. Left internal carotid angiography (Fig. 2 A, B), though it partially filled the external carotid artery (ECA), showed a marked narrowing and elevation of the sup-
raclinoid portion of the internal carotid artery (ICA). The anterior cerebral artery (ACA) was opacified via an abnormal fine vascular network, "moyamoya" vessels. They were mainly supplied by the branches of supraclinoid portion of ICA and partly from fine vessels in the ethmoidal region. The vertebral and basilar arteries were faintly opacified via the muscle branches of ECA. The middle cerebral artery was not visualized in the early arterial phase. In the venous phase, the venous angle was displaced upward and backward. The internal cerebral vein was also elevated. Left external carotid angiography (Fig. 3) showed that the middle meningeal artery (MMA) became dilated and tortuous, and it connected with the anterior falx artery. The left MCA was visualized via the posterior communicating artery (Pcom). Right common carotid angiography (Fig. 4) showed that the "moyamoya" vessels were more evident both in the suprasellar and ethmoidal region. Otherwise, it showed similar findings to the left one. Left vertebral angiography (Fig. 5A) showed that the MCA was filled by the markedly dilated Pcom. The Pcom and the head of basilar artery were elevated. On the base view (Fig. 5B), both Pcoms arched laterally and the horizontal portion of the right ACA was displaced anteriorly. The left counterpart of ACA was not filled. The computed tomography (Fig. 6) disclosed intra-and suprasellar round
Fig. 5. Left vertebral angiogram. A Lateral view, B : Base view. The middle cerebral arteries are opacified via the dilated posterior communicating arteries, which are displaced upward and lateralward. The right anterior cerebral artery, of its horizontal portion, is displaced anteriorly.

mass with homogeneous low density area. Ring-like wall was noted on the contrast enhancement. The third ventricle was elevated and displaced backward, but the foramen of Monro was not obstructed. Lateral ventricle was mildly enlarged. No calcification was shown in the intra- and suprasellar region. From these findings, clinical diagnosis was made as a craniopharyngioma with internal carotid arterial stenosis and subsequent angiographic changes like "moyamoya" disease. The right frontotemporal craniotomy was performed for decompression of optic nerves and tumor removal. The tumor compressed bilateral optic nerves upward and the right ICA laterally. Proximal to the dilated Pcom, the right ICA was collapsed but not hypoplastic. There were some blood stream in it (Fig. 7). Puncture of the tumor yielded gritty motor oil-like fluid, which contained cholesterin crystals on microscopic examination. There were many tortuous fine vessels on the tumor capsule and surrounding basal cortex of the frontal lobe.

Total removal was abandoned because these fine vessels made collaterals to the ACAs. Histological examination of surgical specimen showed only fibrous tissue. Postoperatively, $^{60}$Co irradiation of 5,000 rads was performed. Follow-up angiography at the completion of irradiation showed almost same appearance as the preoperative ones. Dural vessels were recanalized at operative dural incision.

Discussion

Angiographic study in the present case suggests that bilateral carotid arteries became stenotic by compression of the tumor in the sellar region and bilateral horizontal portions of the anterior cerebral arteries were also compressed. These manifestations would be
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Fig. 6. CT scan. Upper : precontrast study, Lower : postcontrast study. Sellar enlargement and round mass in the intra-and suprasellar region. The wall of slightly high density is enhanced on contrast study, but not in its interior. The third ventricle is elevated, but no enlargement of ventricular system is noted.

Fig. 7. Operative photograph. The right internal carotid artery is narrowed by the tumor (T), but distal to the posterior communicating artery (not shown), it becomes dilated. Tortuous vessels are observed on the optic nerve (ON) and tumor capsule. Arrow : Carotid bifurcation, PS : Planum sphenoidale, S : Spatula

favored to the development of fine vessels on the tumor capsule and extensive collateral circulation. Direct visualization at operation supports this view. Although histological study failed to show definitive diagnosis, craniopharyngioma is most probable because of age, its location and cystic content. On angiography\textsuperscript{11-15} craniopharyngioma is avascular tumor frequently with displacement of the surrounding structures. There were only two cases of craniopharyngioma with homogeneous vascular stain\textsuperscript{11-15}. NUMAGUCHI et al.\textsuperscript{9} reported an unusual case of craniopharyngioma with a dense capsular stain and occlusion of the internal carotid artery, which is similar to the reported case herein. They indicated two possibilities as a cause of unusual capsular stain; (a) well developed cavernous branches due to occlusion of the suprasellar segment of internal carotid artery and (b) radiation vasculitis. Numerous fine tortuous vessels on the tumor capsule in the present case are mainly derived from the supraclinoid portion of internal carotid artery, but not from the cavernous branches on angiography. They are partly from the vessels in the ethmoidal region, which
are supplied by the ophthalmic artery and internal maxillary artery. Regarding to the radiation vasculitis, this case had not any radiotherapy before admission, so that the possibility of radiation vasculitis was ruled out.

There are some other cases with tumors, irradiation, neurocutaneous syndrome and arteriosclerosis, in which angiographic findings are the same as in so-called “moyamoya” disease of unknown etiology. The present case suggests that compression of internal carotid arteries by the tumor is closely related to the development of collateral circulation. Although it is disputable that the case presented here is regarded as the so-called “moyamoya” disease, stenosis or occlusion of internal carotid arteries by the tumor can cause development of collateral circulation similar to the so-called “moyamoya” disease.

References