Intraarterial schwannoma in horizontal segment of middle cerebral artery causing subarachnoid hemorrhage

Case report

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A spontaneous subarachnoid hemorrhage (SAH) from the middle cerebral artery is most commonly caused by the rupture of saccular aneurysms and rarely by fusiform aneurysms or arterial dissections/dissecting aneurysms. To the authors’ knowledge, this is the first report of an intraarterial neoplasm causing an SAH. A 44-year-old woman presented with an SAH in the basal cisterns. Subsequent internal carotid artery angiography demonstrated a small bulge on the superior wall of the horizontal (M₁) segment of the middle cerebral artery. However, a pterional craniotomy revealed a well-circumscribed solitary tumor with a diameter of 15 mm involving the superior wall of the M₁ segment as the cause of the SAH. Pathological examination demonstrated typical findings of a schwannoma, elongated cells with tapered, spindle-shaped nuclei and indistinct cell borders, and diffuse immunoreactivity for the S100 protein.

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**KEY WORDS** • artery • angiogram • middle cerebral artery • schwannoma • subarachnoid hemorrhage • vascular disorders

A spontaneous SAH in the basal cisterns is most commonly caused by the rupture of saccular aneurysms arising in the circle of Willis. The MCA, in particular its horizontal (M₁) segment including the bifurcation, is the third most common location for ruptured saccular aneurysms after the ICA and anterior communicating artery. Uncommon lesions of the intracranial proximal arteries causing an SAH include fusiform aneurysms and arterial dissections/dissecting aneurysms. Meanwhile, mycotic and oncotic aneurysms commonly affect distal cerebral arteries.

We report on a case of a schwannoma arising in the wall of the M₁ segment of the MCA and causing an SAH. To our knowledge, this is the first report of an intraarterial neoplasm causing an SAH.

**Case Report**

*Presentation and Examination.* This 44-year-old woman presented with mental deterioration after a sudden bursting headache in May 2008. Computed tomography scanning demonstrated a thin yet diffuse SAH in the basal cisterns (Fig. 1 left). Subsequent left ICA angiography demonstrated a small bulge on the superior wall of the M₁ segment of the MCA (Fig. 1 right, arrow). Lateral lenticonstratate arteries seemed to arise from the superior wall of the M₁ segment just distal to the lesion. The lesion was misinterpreted as a ruptured dissecting aneurysm on the basis of the radiological findings.

*Operation.* Sylvian fissure dissection via a pterional craniotomy revealed the M₁ segment of the MCA. In the middle of the M₁ segment, the superior wall of the artery was involved with a well-circumscribed solitary tumor that had a diameter of 15 mm (Fig. 2A–C). The tumor was yellow, yet the blood-infiltrated part was red and was thought to be the origin of an SAH. Being soft and friable in consistency, the tumor was easily resected. However, the remaining arterial defect was large, and the lenticonstratate arteries were found to arise from the inferior remnant wall of the M₁ segment. End-to-end anastomosis was used to repair the M₁ segment (Fig. 2D). The extent of the arterial defect only allowed slight trimming of the free end of the vessel and required significant tension to approximate the edges.

*Histopathological Examination.* Histological examination of the specimen revealed an intraarterial mass lesion. In one spot, the vascular wall was ruptured and the tumor appeared to penetrate into the wall, and this area was assumed to be the origin of the bleeding (Fig. 3A).
The lesion consisted of elongated cells with tapered, spindle-shaped nuclei, variable chromasia, and inconspicuous nucleoli (Fig. 3B). While these cells had ample pink cytoplasm, there were no discernible cell membranes. Hardly any intranuclear pseudoinclusion was identified. Mild to moderate nuclear atypia was observed in some areas, yet it was considered a degenerative change. No necrosis or mitotic figures were observed in the lesion (Fig. 3B). Immunohistochemically, the tumor cells showed diffuse immunoreactivity for the S100 protein (dilution 1:2000, Dako) (Fig. 3C), yet it was negative for smooth muscle actin (1:100, Dako) (Fig. 3D), epithelial membrane antigen (1:300, Dako), and CD34 (1:100, Dako). The Ki-67 (1:300, Dako) labeling index showed a regional variability, ranging from 3% to 12%. Based on the histological examination and immunohistochemistry results, an intraarterial schwannoma was diagnosed.

Postoperative Course. The patient was hemiplegic, and the repaired MCA was occluded postoperatively. Endovascular recanalization was attempted without success.

Discussion

An oncotic intracranial aneurysm is a rare yet well-known entity. Tumor embolization or contiguous tumor growth can lead to neoplastic cellular invasion of the cerebral arteries, resulting in aneurysm formation commonly in distal arteries. Metastatic deposits from an atrial myxoma, bronchogenic carcinoma, renal cell carcinoma, or choriocarcinoma and contiguous growth of gliomatous tumors have already been reported in relation to oncotic aneurysms.2,4,10,12,16,19 However, this is the first reported case of a primary intraarterial neoplasm developing an intracranial aneurysm. This is the third pathogenesis of oncotic aneurysms, in addition to metastatic and contiguous types.

An intravascular schwannoma was previously reported by Gaudi et al.9 In that case a middle-aged woman presented with a subcutaneous nodule overlying her posterior calf, and the cutaneous schwannoma was found to involve the dermal venous system. However, an intraarterial schwannoma, as in the present case, has never been reported.

An intracranial schwannoma arises from the sheath of the cranial nerves, except for the olfactory and optic nerves and is a benign nerve sheath neoplasm originating from Schwann cells. Thus, an intracranial schwannoma that does not arise from the cranial nerve is exceedingly rare. Only a few cases of schwannomas arising in the brain parenchyma or olfactory nerve have been reported.5,8,13,15 Aberrant Schwann cells in the brain parenchyma and mesenchymal pial cells transformed into ectodermal Schwann cells are the 2 prevailing theories explaining the possible origin of these tumors.

An intraarterial schwannoma would seem to arise...
FIG. 3. Photomicrographs showing the intraarterial schwannoma. A: The arterial wall (arrows) is ruptured, and the tumor (dotted line) appears to penetrate into the arterial wall. B: Higher-magnification image revealing a cellular lesion of spindle cells with wavy nuclei and ample pink cytoplasm in the arterial wall. C: The tumor cells show diffuse nuclear and cytoplasmic reactivity for S100 protein. D: Smooth muscle actin tested positive in the smooth muscle of the vascular wall, yet tested negative in tumor cells. H & E (A and B), original magnification ×100 (A) and ×400 (B–D).

from Schwann cells that exist within the perivascular nerve plexuses around large arteries in the subarachnoid space. Rich cerebral vascular innervation has already been investigated in animals and humans. In particular, Cuevas et al. studied MCA innervation using transmission electron microscopy and observed nerve fibers surrounded by Schwann cell processes in the tunica adventitia and outermost layer of the tunica media.

An intraarterial schwannoma presenting with an SAH can pose diagnostic problems. In such cases, angiographic evaluation is initially performed to find a vascular lesion as the cause of the SAH, and a small bulge in the proximal cerebral arteries can lead to a misdiagnosis of a dissecting aneurysm due to its higher incidence. However, if an intraarterial neoplasm is considered in the differential diagnosis, enhanced MR images can be helpful.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Park. Acquisition of data: both authors. Analysis and interpretation of data: Lee. Drafting the article: Park. Approved the final version of the manuscript on behalf of both authors: Park. Study supervision: Park.

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