Arteriovenous Malformation Surrounding the Trigeminal Nerve
—Case Report—

Boris KRISCHEK, Sachiko YAMAGUCHI, Ulrich SURE, Ludwig BENES, Siegfried BIEN*, and Helmut BERTALANFFY

Departments of Neurosurgery and *Neuroradiology, Philipps University Hospital Marburg, Marburg, Germany

Abstract

A 57-year-old man presented with subarachnoid hemorrhage due to the rupture of an arteriovenous malformation (AVM) located at the base of the root of the right trigeminal nerve. In contrast to previous similar cases, his history included no evidence of trigeminal neuralgia or sensory loss. Right vertebral artery angiography revealed a doubled superior cerebellar artery feeding the angioma nidus. The patient refused radiotherapy and preferred surgical treatment. Intraoperatively, a close relationship between arterial feeders and rootlets of the trigeminal nerve was observed. Complete removal of the malformation was achieved and confirmed angiographically. The postoperative course was complicated by subdural hygroma that required repeated drainage and eventually a shunting procedure. This case demonstrates that microsurgical treatment of a trigeminal AVM is feasible. However, stereotactic radiosurgery may be the preferred treatment option considering the potential for postoperative complications.

Key words: arteriovenous malformation, subarachnoid hemorrhage, subdural hygroma, trigeminal nerve

Introduction

Trigeminal nerve arteriovenous malformations (AVMs) are known to cause trigeminal neuralgia. Moreover, the close relationship between cranial nerves and symptomatic AVMs usually results in nerve paresis. We describe a case of trigeminal nerve AVM that manifested as subarachnoid hemorrhage (SAH) without preceding symptoms.

Case Report

A 57-year-old man complained of sudden onset of headache and nausea. Cranial computed tomography (CT) revealed SAH (Fig. 1). He suffered no further neurological symptoms and none were noted in his history. Angiography revealed an angioma nidus on the right side of the brain stem supplied by a doubled superior cerebellar artery (Fig. 2).

The patient refused radiosurgery with a linear accelerator. Endovascular occlusion failed due to the small caliber of the vessel. Therefore, the vascular malformation was treated by microsurgery. A
right subtemporal approach was chosen because proximal control of the superior cerebellar arteries was considered necessary. The draining vein of the angioma was identified at the base of the root of the trigeminal nerve (Fig. 3). Inspection and intraoperative Doppler sonography examination revealed several pathological vessels within and around the trigeminal nerve reaching the gasserian ganglion. Some fibers of the trigeminal nerve had to be sacrificed during dissection of the AVM.

Postoperatively, the patient had partial sensory loss in the first and second branches of the right trigeminal nerve. Postoperative angiography demonstrated complete removal of the AVM and only a single right superior cerebellar artery was detected (Fig. 4).

Three months after the initial operation the patient presented at our department complaining of headache, gait ataxia, and left hemiparesis. Magnetic resonance imaging showed subdural hygroma of the right hemisphere (Fig. 5 left). Two separate burr hole procedures were performed, the latter with insertion of a temporary subdural drainage. The patient was discharged without neurological deficits.

Two months later, the patient was readmitted with the same complaints. CT showed a recurrent subdural hygroma. A subdural-peritoneal shunt was inserted. Postoperatively, the patient did well and the subdural hygroma completely disappeared on

Fig. 2 Anteroposterior (left) and lateral (right) left vertebral artery digital subtraction angiograms (arterial phase) demonstrating an angioma nidus (arrow) supplied by a doubled superior cerebellar artery (arrowhead).

Fig. 3 left: Intraoperative photograph showing the close relationship between the base of the trigeminal nerve (arrow) and the arteriovenous malformation. right: Illustration (by S. Yamaguchi) depicting the proximal part of the trigeminal nerve (arrow) and the malformation (arrowhead).

Fig. 4 Postoperative anteroposterior (left) and lateral (right) left vertebral artery digital subtraction angiograms showing no residual arteriovenous malformation and only one right superior cerebellar artery (arrowhead).
CT (Fig. 5 right). He has remained free of complaints apart from the partial facial hypesthesia for the past 2 years.

**Discussion**

SAH usually results from rupture of an intracranial aneurysm. Thus, the first differential diagnosis after the initial CT in our case was aneurysmal SAH. Surprisingly, angiography revealed the correct source of hemorrhage as a trigeminal AVM. AVMs usually bleed intraparenchymally, but may also cause SAH.24)

Trigeminal nerve AVM has caused long-lasting trigeminal neuralgia in all previous cases. Our patient had no trigeminal neuralgia, but presented with acute SAH. Trigeminal neuralgia may be caused by the continuous pulsation of the malformation closely related to the cranial nerve.5,7,14,15) The reason for the absence of neuralgia in our case might have been the equal distribution of blood flow through several vessels in the vicinity or located within the trigeminal root such as the doubled superior cerebellar artery in addition to the pathological feeding arteries (Fig. 2). We assume that the actual vessel pulsation may not have been sufficient to cause trigeminal neuralgia. Contrary to this hypothesis, a previous patient with a doubled superior cerebellar artery feeding the trigeminal AVM presented with neuralgia.15) Postoperative angiography in our case strongly suggested that one of the branches was primarily the main feeding artery, as it could not be demonstrated after the surgical procedure (Fig. 4).

Although the neurovascular compression seen during microvascular decompression surgery occurs mainly in the trigeminal root entry zone (REZ) and the midthird of the trigeminal nerve,5,10) there is a correlation between the length of the central nervous system segment and the incidence of vascular compression syndromes,17) presumably because the alignment difference of the myelin sheaths and separation of myelin forming cells in the peripheral nervous system segment are more resistant to compression than in the central nervous system segment. Five cases of micro-AVM embedded within the trigeminal REZ caused trigeminal neuralgia.11) In contrast to these vascular malformations, in our case the nerve compression by the AVM was remote from the REZ (Fig. 3). This is probably why the patient did not complain of previous symptoms.8,20) In a recent very similar case to ours, the reason for the missing trigeminal neuralgia was possibly the aberrant formation in the sprouting meshwork of primordial vessels in the trigeminal root during embryogenesis (weeks 5 to 6 of gestation).13)

As an alternative treatment modality, radiosurgery was primarily offered to our patient, who nevertheless preferred surgical therapy. Unfortunately, the postoperative course in our patient was complicated by subdural hygroma that required repeated surgical therapy. Eventually, subdural-peritoneal shunting of the hygroma became necessary. Possibly, such complications would not have occurred had the AVM only been treated by radiosurgery.2,16,21) On the other hand, these complications did not cause any permanent morbidity. When choosing radiosurgery as therapeutic option, stereotactic planning may be difficult for lesions associated with SAH located close to the brainstem.2,3,21) AVMs cannot be obliterated immediately after radiosurgery23) since the mechanism of radiation-induced blood vessel obliteration is gradual occlusion of the vessels, which remain in situ. Moreover, radiosurgery may not prevent significant rebleeding after this treatment. A study of patients with brain stem AVMs treated by radiosurgery found five of 30 patients had hemorrhage from the AVM after irradiation, including four deaths.9) In all five cases the AVMs had not been obliterated completely after radiosurgery.

Our case and others demonstrate that microsurgical removal of trigeminal nerve AVM is feasible and can have a good long-term outcome. However, additional surgical procedures may be required due to postoperative complications as seen in the present case. Radiosurgery may be an alternative treatment option for small AVMs of the trigeminal root but post-irradiation complications are significant, so which treatment option is preferable in...
such cases remains debatable.

References


24) Yasargil MG: Microsurgery IIIA. Stuttgart, New York, Thieme Verlag, 1987, 408 pp

Address reprint requests to: B. Krischek, M.D., Department of Neurosurgery, Philipps University Hospital Marburg, Baldingerstrasse, D-35033 Marburg, Germany.
e-mail: krischek@med.uni-marburg.de