Cerebral microarteriovenous malformation

In 1961 Margolis et al. proved through autopsy sectioning of 4 fatal intracranial hematomas that microarteriovenous malformations could act as sources of hemorrhage [1].

In the early literature, multiple reports documented tiny vascular lesions pathologically compatible with arteriovenous malformations (AVMs) in the walls of hemorrhage cavities [2] [3] [4] [5].

In 1987 Yaşargil defined cerebral microarteriovenous malformations as a subgroup of pial arteriovenous malformation characterizad by a nidus of 1 cm or smaller [6].

Epidemiology

Micro-arteriovenous malformation account for < 8% of surgically treated arteriovenous malformations (AVMs), and most reports of micro-AVMs as defined by Yaşargil are either isolated cases or cases included in series of angiographically occult arteriovenous malformations. In the literature Alén et al., have found only 4 small series of patients suffering from this pathological entity [7].

Willinsky et al. found an approximately 10% incidence in their series of brain AVMs [8] [9] , and Stiver and Ogilvy found in a retrospective review that 8% of the AVMs surgically treated at the Massachusetts General Hospital were micro-AVMs [10]

In the study of Alén et al., in which they used inclusion criteria and a study population similar to those of previous studies, revealed a similar incidence of micro- AVMs at 10.7% [11].

Most are diagnosed in patients in their 3rd or 4th decade, and there is no sex predilection [12].

Clinical features

Intracerebral hemorrhage is the main clinical manifestation of micro-AVMs [13].

In the series of Alén et al., all patients, except 2 in whom the AVM was found incidentally, presented with intracranial hemorrhage. In 1 patient, a previous cerebral hemorrhage had not been visualized with angiography 3 years earlier and was diagnosed with a micro-AVM after a second hemorrhage. In previous studies, all patients presented with spontaneous intracranial hemorrhage, although in some patients neurological symptoms preceded the hemorrhagic ictus [14] [15] [16] [17].

This dominant presentation with hemorrhage contrasts with that for larger AVMs, in which 65%–70% of cases present with hemorrhage.

Higher pressures in the feeding pedicles of small AVMs and a greater tendency for hemorrhaging in low- flow shunting compared with high-flow shunting AVMs have been reported [18] [19].

Diagnosis

It is beneficial to find a tiny nidus of dense vessels located on hematoma wall on enhanced thin layer CT scanning for a clear diagnosis and to detect any abnormal feeding artery or venous drainage for an indirect diagnostic evidence [20].

Micro-arteriovenous malformation are visible on angiography, sometimes just as an abnormal arteriole without draining veins or as an abnormal draining vein with the feeding vessels remaining undetectable. Other times a tiny lesion with the classic appearance of pathological arterioles and pathological draining veins is visible on angiography.

Superselective angiography has increased the ability to reach a diagnosis of micro- AVM when regular DSA is questionable or negative, allowing for more precise localization of the lesion and characterization of the angioarchitecture [21].

Differential diagnosis

Yasargil differentiated the cerebral microarteriovenous malformation from occult arteriovenous malformations and cryptic arteriovenous malformations [22].

Treatment

Resection is the main method of treatment for micro-AVMs [23].

Immediate complete obliteration of a micro-AVM with a high permanent cure and low morbidity rates was accomplished using endovascular treatment. Early embolization after bleeding should be considered as an alternative to resection [24].

Although microsurgical resection and endovascular embolization may both be considered in ruptured superficial micro-AVMs, the results of Overdevest et al., although based on a small series – showed that endovascular treatment was associated with a lower success rate (25% vs 80%) and a higher procedural complication rate (22% vs 0%) than microsurgical resection. In there experience the higher success rate of microsurgery benefits from the use of intra operative ICG-VA. This results suggest that microsurgery, using ICG-VA, should be considered the primary choice for the treatment of superficial micro-AVMs. In case an initial endovascular treatment attempt is offered, procedural risks should be kept at a minimum, as subsequent microsurgery can be performed safely and effectively [25].

Outcome

In the majority of cases, clinical outcome is determined primarily by the degree of recovery from the hemorrhage that brings the diagnosis of a micro-AVM to attention [26].

The majority of patients described in the different studies experienced improvement after hemorrhage, and very few permanent neurological deficits were related to the treatment procedure in the follow-up [27) 28) 29) 30) 31)].

In the series of Alén et al., no deaths occurred, and outcome was excellent for 18 patients (64%). Eighteen percent recovered with moderate disability, and 18% had severe disability. No complication was attributable to the treatment, but up to 57% (16) of the cases initially presented with neurological deficit. In general, the long-term neurological deficit is related to the severity of the initial hemorrhage [32)].

Case series

Eight consecutive patients with superficial micro-AVMs are presented. All patients received an initial embolization attempt with either nBCA or ONYX. If complete obliteration was not obtained, either a second embolization or surgical resection was offered. At surgery, indocyanine green video angiography (ICG-VA) was used in all cases. Effectiveness and safety of all procedures were evaluated retrospectively. Functional outcome at 6 months was assessed by the modified Ranking Score (mRS).

Patients had a mean age of 52±17 years and all presented with hemorrhage. The mean nidus size was 4 mm, and was localized supratentorially in 5 cases and infratentorially in 3. Initial embolization was successful in 2 patients (25%). One patient underwent a second, unsuccessful, embolization attempt and 1 patient did not receive further treatment. Consequently, five patients underwent surgery, which was successful in four (80%). The unsuccessful case was successfully reoperated. The only two procedural complications were related to superselective embolization, but neither caused clinical sequelae. Mean clinical follow-up was 29 months (range, 4–75mo), with mRS 0 in 2, mRS 1 in 4 and mRS 3 in 2 cases.

In a current case series, embolization of superficial micro-AVMs was associated with a lower success rate (25% vs 80%) than microsurgery and a higher procedural complication rate (minor complications: 22% vs 0%) [33)].

The clinical data, radiological features, treatment, and follow-up results for a consecutive series of 13 cases with micro-AVMs were retrospectively analyzed.

All 13 patients presented with intracerebral hemorrhage. Ten cases were confirmed by enhanced thin layer CT scanning and CTA, and the other 3 cases were confirmed by DSA. Treatment consisted of surgical removal in 10 cases, endovascular embolization in 1, and radiosurgery in 2. The modified GOS score was achieved in the third month after discharge: 10 cases were rated with 5 points (good recovery), 1 case was rated with 4 points (mild disability), and 2 cases were rated with 3 points (severe disability). During follow-up, No case of rebleeding was reported.

Intracerebral hemorrhage is the main clinical manifestation of micro-AVMs. It is beneficial to find a tiny nidus of dense vessels located on hematoma wall on enhanced thin layer CT scanning for a clear diagnosis and to detect any abnormal feeding artery or venous drainage for an indirect diagnostic evidence. Resection is the main method of treatment for micro-AVMs [34)].

Between 1997 and 2006, 25 patients (12 females and 13 males) with 26 micro-AVMs were treated. Twenty-four patients presented with intracerebral hematoma and 1 with subarachnoid hemorrhage only. All patients underwent CT on admission, diagnostic cerebral angiography, and 1 session of endovascular treatment during the acute phase.

Procedure-related complications occurred in 3 patients (12%), which caused temporary hemiparesis in 1 (4%) and no clinical sequelae in 2 patients (8%). Complete nidus obliteration was achieved at the end of the embolization in 22 (84.6%) of 26 lesions. Two recurrences were evident on follow-up angiography 6 months postembolization, resulting in a complete obliteration rate of 77% (20 of 26 lesions) after a single treatment. Late angiography was performed in 12 patients, and no further recurrences were identified.

Immediate complete obliteration of a micro-AVM with a high permanent cure and low morbidity rates was accomplished using endovascular treatment. Early embolization after bleeding should be considered as an alternative to resection [35].

The clinical presentation, diagnostic features, principles of endovascular or surgical treatment, and outcomes for a consecutive series of 10 patients (five male, five female; mean age, 48.8 years; age range, 31-65 years) with angiographically demonstrated cerebral micro-AVMs were retrospectively analyzed.

All patients presented with a cerebral hematoma (supratentorial in eight, infratentorial in two, intraventricular in one, subarachnoid in one; mean volume, 11.6 cm(3)), which was superficially situated in nine patients. Neurologic deficits were observed in nine patients, and three patients had seizures. The mean delay between the onset of symptoms and diagnosis was 129.8 days (range, 6 days to 1 year). Superselective angiography was performed in seven patients and followed by successful acrylic embolization of the lesion in five. Five patients underwent surgical intervention, which led to definitive resection. Although long-term neurologic problems were present in eight patients, they were able to return to their previous activities and employment.

The diagnosis of cerebral micro-AVMs requires a high index of suspicion, especially in young adults with atypical hemorrhaging. Single-shot embolization of micro-AVMs may be a safe alternative to the established surgical therapy in select cases. Outcomes depend mostly on the clinical conditions at admission [36].

Twelve patients with micro-AVMs that had been treated by surgical resection were retrospectively analyzed. The mean follow-up monitoring period was 35 months (range, 2-76 mo). Outcomes, as assessed in follow-up visits and telephone interviews (using a questionnaire), were classified according to the Glasgow Outcome Scale.

All 12 patients presented with intracranial hemorrhage, which was intraparenchymal and superficially situated in 10 patients (83%) and intraventricular in 2 patients (17%). Hemorrhages were large (mean volume, 23 ml3; range, 1-58 ml3) and were associated with neurological deficits for 10 of 12 patients (83%). The identification of an arterialized draining vein during surgery and stereotactic angiography greatly facilitated surgical localization of the lesions. One patient (8%) developed a mild permanent deficit as a result of surgery. Although Glasgow Outcome Scale scores were excellent for all except

one patient, nine patients (75%) experienced long-term neurological problems.

Micro-AVMs typically present with large hemorrhages and are associated with significant neurological deficits. If a superficial clot is present, surgical resection of the lesion is strongly advocated. The ultimate clinical outcomes are determined primarily by deficits present after the initial hemorrhaging episodes [37].

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13 patients had small (less than 1 cm) parenchymal arteriovenous malformations (mAVMs) with small nidus or fistula and a single normal-sized feeding artery and draining vein. All 13 patients presented with intracerebral haematomas (ICHs). The average age in this group was 31 years with no sex dominance; 8 patients had no antecedent symptoms. In 11 patients the small AMV could be demonstrated angiographically, with the remaining 2 malformations evident at surgery. In addition, all these mAVMs, being superficial (95% cortical), were surgically removable with no perioperative morbidity. They were not accessible by endovascular approach. This population group narrows the concept of occult vascular lesions if high quality angiographic studies are performed. mAVMs are by nature CT and MRI occult [38].

Case reports

A patient who presented with superior quadrianopsia due to an occipital micro AVM that bled into the optic radiation. Onyx embolization was attempted. However, early follow-up angiogram revealed recanalization and recurrence of the AVM. He was then taken to the hybrid operative room, where a complete resection was achieved confirmed by intraoperative angiogram. He made a complete recovery with no new neurologic deficit and stable visual field deficit. This case demonstrates treatment strategy, surgical planning, and technical nuances in microsurgical resection of micro AVMs located in an eloquent area. Management of a ruptured microarteriovenous malformation (microAVM) localized in an eloquent brain region is challenging. The major difficulties are those related to localizing and defining the micronidus in order to achieve complete resection and definitive cure while preserving function. The best and definitive treatment for AVMs is either surgical resection or radiosurgery. However, in thee institute a small subset of microAVMs might be cured by endovascular embolization in a single session. In the case presented here, a single feeder was demonstrated and microcatheter navigation toward a good working position seemed feasible; thus they decided to try first an endovascular approach [39].

A 39- year-old patient who presented with classic symptoms of trigeminal neuralgia (TN), but was found on imaging studies to harbour a small intrinsic vascular malformation within the nerve. Based on size and drainage, the arteriovenous malformation (AVM) was Spetzler-Martin Grade III and no previous history of bleeding was reported. The patient had failed a trial of carbamazepine, and no surgical procedures had been performed. A decrease in symptoms was reported by the 6-month follow-up. A review of the literature on microAVM-induced TN is provided as well as a discussion of management [40].

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