Spontaneous regression of cerebral arteriovenous malformations: clinical and angiographic analysis with review of the literature

Abstract Spontaneous regression of cerebral arteriovenous malformation (AVM) is rare and poorly understood. We reviewed the clinical and angiographic findings in patients who had spontaneous regression of cerebral AVMs to determine whether common features were present. The clinical and angiographic findings of four cases from our series and 29 cases from the literature were retrospectively reviewed. The clinical and angiographic features analyzed were: age at diagnosis, initial presentation, venous drainage pattern, number of draining veins, location of the AVM, number of arterial feeders, clinical events during the interval period to thrombosis, and interval period to spontaneous thrombosis. Common clinical and angiographic features of spontaneous regression of cerebral AVMs are: intracranial hemorrhage as an initial presentation, small AVMs, and a single draining vein. Spontaneous regression of cerebral AVMs can not be predicted by clinical or angiographic features, therefore it should not be considered as an option in cerebral AVM management, despite its proven occurrence.

Keywords Brain · Arteriovenous malformations · Thrombosis · Spontaneous regression · Angiography

Introduction

Cerebral arteriovenous malformations (AVMs) are dynamic lesions. Their arterial supply and/or venous drainage may change [1]. Furthermore, they can increase in size [2, 3, 4] or progress to complete thrombosis spontaneously [2, 3]. Spontaneous regression or thrombosis of cerebral AVMs is one of the most poorly understood phenomena of cerebral AVM. It has been reported infrequently, however, the predisposing factors and angiographic characteristics are not known [2, 3, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18]. In this study, we report the clinical and angiographic findings in four patients who had spontaneous angiographic regression of cerebral AVMs and 29 cases in the literature. Additionally, we discuss the possible underlying pathophysiology.

Materials and methods

A retrospective review of the cerebral AVM database of the Toronto Vascular Malformation Study Group (n = 797) revealed four patients (M:F = 3:1) with spontaneous complete angiographic regression. The inclusion criteria for the patients were: angiographic confirmation of the cerebral AVM on initial presentation, no history of therapeutic interventions including endovascular treatment, surgery or radiation and no evidence of arteriovenous shunts on follow-up angiogram.

Clinical features studied included: age at diagnosis, initial mode of presentation, interval to thrombosis, and hemorrhagic events before thrombosis. The size and location of the cerebral AVM were determined on initial CT or MR images. The size of the cerebral AVM was categorized into small (< 3 cm), medium (3–6 cm) and large (> 6 cm) according to its maximum nidus diameter. The number of arterial feeders, number of veins and venous drainage patterns (deep, superficial or combined) were analyzed, based on the initial angiography. We also reviewed 29 cases from the literature that were reported to have spontaneously regressed.
Results

Clinical data analysis

The mean ages of our cases at diagnosis and of the reported cases were 37 and 31 years, respectively. Three of our patients (75%) presented with intracranial hemorrhage with neurological deficits (Fig. 1), and one presented with headache without neurological deficit (25%) (Fig. 2). In the literature, the initial presentations were described in 25 cases. They included seizure (20%), intracranial hemorrhage with neurological deficit (52%), headache without neurological deficit (16%), loss of consciousness (4%), increasing head circumference (4%) and clinically normal (4%). The interval to thrombosis in our patients was 3 months to 18 years (median = 2.4 years). In the literature, it was from 42 days to 26 years (median = 3 years). In our series there were no additional hemorrhagic and/or non-hemorrhagic neurological events during the interval periods. In the literature, 60% of patients had neurological events between the presentation and spontaneous regression. Among them, one-third of the patients had hemorrhagic events with neurological deterioration before thrombosis, and two-thirds had non-hemorrhagic neurological events such as seizure, hemiparesis, weakness or sensory changes.

Radiological data analysis

In the literature, there were 13 small (50%), ten medium (38%) and three large (12%) sized AVMs that spontaneously regressed. In our series, three cases were small and one was medium in size (Fig. 2). The locations of the AVMs were variable in both the literature and in our series. A single feeding artery was demonstrated in one of our cases (25%) and seven of the 20 cases (33%) from the literature. Seventy five percent (n=3) of our cases and 65% (n=13) in the literature showed more than three arterial feeders. A single draining vein was present in 75% of our cases (three of four) and in 74% in the literature (11 of 15). The clinical and angiographic characteristics of our cases and the literature group are summarized in Tables 1 and 2.

![Fig. 1a–e. Case 1. A 36-year-old man with AVM presented with intracranial hemorrhage 2.5 years ago. a Initial CT shows extensive hematoma on left basal ganglia. b Initial arterial phase angiogram demonstrates arteriovenous shunts with intramidal pseudoaneurysm, and venous phase shows single draining vein to cavernous sinus. Additionally, superior ophthalmic vein is engorged. d, e On follow-up angiogram there is no angiographic evidence of arteriovenous shunts.](image-url)