Multiplicity of dural arteriovenous fistulas

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Object. Dural arteriovenous fistulas (AVFs) are a well-known pathoanatomical and clinical entity. Excluding bilateral involvement of the cavernous sinus, multiple dural AVFs are rare, with isolated reports in the literature. The additional risk associated with multiplicity is unknown, although it has been claimed that there is a greater risk of hemorrhage at presentation. In a group of 284 patients with dural AVFs consecutively treated at a single center, the occurrence of multiplicity is investigated and its risk factors for hemorrhage are identified.

Methods. Among the 284 patients with both cranial and spinal dural AVFs, 20 patients with multiple fistulas were found. Nineteen (8.1%) of 235 patients with cranial AVFs had multiple cranial fistulas, and one (2%) of 49 patients with spinal AVFs harbored two spinal fistulas. Twelve patients were found to have a lesion at two separate sites, seven patients had them at three locations, and one patient had four fistulas, each at a different site.

In the subgroup with multiple AVFs the percentage of hemorrhage at presentation was three times higher than in the entire group (p = 0.01). Cortical venous drainage in cranial fistulas was present in 84% of patients with multiple lesions compared with 46% of patients with solitary lesions (p < 0.005).

Conclusions. Multiple dural AVFs are not rare. In this group of 284 patients it was found in 8.1% of all patients with cranial dural AVFs. Multiplicity was associated with a higher percentage of cortical venous drainage, a pattern of drainage reportedly yielding a higher risk for hemorrhage.

Key Words • dural arteriovenous fistula • multiple lesions • Borden classification • hemorrhage

Clinical Material and Methods

Source of Cases

The University of Toronto Brain Vascular Malformation Study Group retrospectively acquired data in patients who had received a cerebrovascular diagnosis during the period from 1984 to 1989, and has been gathering information prospectively since then. The continually updated database now contains information on 925 pial AVMs, 350 cavernous malformations, and 284 dural AVFs. The group of patients with dural AVFs of the adult type was investigated in detail. This consecutive group, which consists of 235 patients with cranial fistulas and 49 with spinal fistulas, was diagnosed between June 1984 and March 2001. From both the cranial and the spinal population, 20 patients were found to have two or more fistulas (Table 1).

Multiplicity of dural AVFs was defined as the occurrence of two or more fistulas that are anatomically separate within one patient. Therefore, to be considered multiple, the locations of the fistulas, the arterial feeding vessels, and the venous drainage had to be distinct. Simultaneous existence was not obligatory; multiplicity was also considered present if two or more fistulas were metachronously diagnosed. Bilateral cavernous sinus dural AVFs were excluded, because both fistulas are involved in the same location.

In all patients one or more angiograms were available, and these were assessed to verify multiplicity. The signs
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and symptoms at clinical presentation were noted and the pattern of venous drainage was graded according to the Borden classification.3 The chi-square test was used to determine statistical significance.

Summary of Cases

The 20 patients included in the study presented with dural AVFs at the Toronto Western Hospital between March 1986 and February 2000. There were four children and 16 adults, with males predominant; there were 14 males compared with six females. The children presented at a mean age of 2.6 ± 2 years (standard deviation) and the mean age in the adult subgroup was 58.3 ± 11.8 years (standard deviation).

Multiple cranial dural AVFs were found in 19 patients; these lesions all existed simultaneously. The two spinal dural AVFs in one patient were metachronously diagnosed when the individual failed to improve after curative treatment of the fistula that was responsible for the initial presentation. Twelve patients (60%) had a double lesion, seven patients (35%) harbored a fistula at three separate sites, and one patient (5%) was diagnosed with four concurrent lesions. In most cases the clinical presentation was aggressive: in the group with multiple cranial fistulas, seven (37%) presented with a hemorrhage and nine (47%) with a neurological deficit. One child (5%) presented neonatally with congestive heart failure. Only two patients (11%), both adults, presented with a benign history of bruit. The patient with the spinal dural AVFs presented with a myelopathy caused by venous congestion of the spinal cord. Disregarding the patient with the double spinal dural AVFs, cortical venous reflux (Borden Type II or III) was present in 16 (84%) of 19 patients.

The mean follow up in all but one patient (95%) was 4.7 years (range 0.8–14.2 years). All 17 patients with a Borden Type II or III fistula were considered to harbor a high risk for hemorrhage or neurological deficit. Fifteen (88%) were treated using endovascular embolization (five patients, 29%), surgical disconnection of the shunting vein (four patients, 24%), or a combination of both methods (six patients, 35%). One patient (6%) refused therapy and died shortly thereafter of a cerebellar hemorrhage, and one other patient (6%) was lost to follow-up review before treatment. The fistulas with a Borden Type I classification were considered to be benign and were followed in the outpatient clinic on a yearly basis.

Discussion

Dural AVFs are generally accepted to be acquired lesions2,15 that consist of abnormal arteriovenous shunts within the leaflets of the dura.10 Usually they develop within the wall of a dural sinus, but other sites not directly related to a dural sinus have been demonstrated as well.3,5 The exact pathogenesis is still unclear, although in a rat model sinus thrombosis and venous hypertension have been reported as major provoking factors.1,19 In specimens obtained in humans, Uranishi, et al.26 have demonstrated sinus thrombosis histologically in 100% of surgically treated cranial dural AVFs (nine cases) and underlined the possible impact of angiogenetic growth factors.

The scarcity of reports in the literature concerning multiple dural AVFs, which are mostly illustrated in case histories15,14,16,15 and small series,21 has imposed the idea that multiplicity is rare. Barnwell, et al.,2 suggested a frequency of 7% in their series of 105 patients, six of whom had two lesions, and one of whom harbored three separate fistulas. The existence of more than three fistulas has not been described in any of the earlier reports. In our series two, three, and even four lesions are demonstrated in 19 (8.1%) of 235 cranial dural AVFs, which confirms the suggestion by Barnwell, et al., and indicates that multiplicity is not at all rare. The spinal case, one (2%) of 49 patients, hereby proves that multiplicity is not limited to the cranial com-

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part. To eliminate the potential risk of missing a fistula with cortical venous reflux, a complete angiographic evaluation should be performed, both at clinical presentation and when there is a change in the symptomatology.

Barnwell, et al., 2 also postulated, based on their series of seven patients, that multiple lesions are related to a more aggressive clinical presentation and entail a higher risk of hemorrhage. Based on our observations, their assumption proves to be right. Disregarding the child with congestive heart failure, which was related to a significant hemodynamic shunt, a benign clinical presentation in our multiple AVF group was found in only two cases (11%); 89% of patients presented with a hemorrhage or a neurological deficit. Hemorrhage as a presenting sign was three times more frequent in the group with multiple cranial lesions (seven of 19 cases) than in patients with solitary cranial fistulas (27 of 216 cases; p = 0.01). In our opinion this aggressive behavior at presentation is attributable to the high rate of cortical venous reflux. Sixteen patients (84%) were demonstrated to have cortical venous reflux, which is a significantly higher percentage compared with the 46% (100 of 216 cases) with cortical venous reflux in solitary cranial fistulas (p < 0.005). As reported by our group and by others, cortical venous reflux is related to aggressive behavior and a high risk of hemorrhage in AVFs, both at presentation and in the natural history. 7-9 Consequently, treatment in this series was restricted to lesions that were Borden Type II or III. In accordance with the report by Davies, et al., 6 the fistulas without cortical venous reflux were closely watched instead of surgically treated, because they are considered to be benign.

Conclusions

Multiple dural AVFs are not rare. In our group of 284 patients it was found in 8.1% of those with cranial dural AVFs and in 2% of patients with spinal dural AVFs, underlining the necessity of a full angiographic evaluation at presentation. In addition, multiplicity was associated with a significantly higher percentage of cortical venous drainage, a pattern of drainage clearly reported to carry a higher risk of hemorrhage.

References


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