Child's Nerv Syst (1998) 14: 285–287

CASE REPORT

Hiroki Kurita
Tomio Sasaki
Ichiro Suzuki
Takaaki Kirino

Basal ganglia arteriovenous malformation presenting as "writer's cramp"

Received: 20 August 1997
Revised: 29 December 1997

H. Kurita (✉) · T. Sasaki · I. Suzuki
T. Kirino
Department of Neurosurgery,
Faculty of Medicine,
University of Tokyo,
7-3-1 Hongo, Bunkyo-ku,
Tokyo, 113 Japan
Tel.: +81-3-3815-5411
Fax: +81-3-5800-8655

Abstract A 12-year-old girl with a 3-year history of writer's cramp in association with a basal ganglia arteriovenous malformation (AVM) is reported. The lesion was localized to the left globus pallidus and putamen, extending to the adjacent white matter of the frontal lobe. Our experience confirms a common anatomical basis of symptomatic focal dystonia: disruption of the pathways within and adjacent to these structures. Appropriate imaging should be carried out in patients with unexplained movement disorders.

Key words Arteriovenous malformation (AVM) · Writer's cramp · Symptomatic focal dystonia · Basal ganglia

Introduction

Writer's cramp is a well-recognized condition defined as a focal task-specific dystonia characterized by difficulty in executing fine movement of the hand [8, 14, 18]. Although unilateral symptomatic dystonia associated with contralateral localized basal ganglia lesions has been documented [1, 2, 4, 7, 11, 13, 16, 19], vascular anomalies have only rarely been implicated as a cause of focal dystonia [3, 6–8, 17]. Here we report a patient with writer's cramp as the first manifestation of basal ganglia arteriovenous malformation (AVM).

Case report

A 12-year-old right-handed girl was the product of a normal pregnancy and delivery, and early development had been normal. There was no family history of movement disorders. She had first developed difficulty in writing at the age of 9 years, grasping the pen too firmly when she attempted to write. At that time, she did not experience weakness, sensory disturbance, or involuntary movements, and had no difficulty in using her right hand for other skilled tasks. The condition was thought to be a phobic reaction to writing, and the patient was treated conservatively. Subsequently, the symptom progressed gradually and she began to prefer the left hand for writing. At the age of 11 years, she was noted to have weakness of her right hand, occasionally associated with involuntary movement and trouble in performing fine tasks. At the age of 12, she experienced severe headache associated with nausea and vomiting and was evaluated at our institute. Results of neurological and physical examinations were normal, except for mild weakness of the right hand. No abnormality was found in the electroencephalogram (EEG) recording. Magnetic resonance imaging (MRI) demonstrated an unruptured left basal ganglia AVM localized in the left globus pallidus and putamen, extending to the adjacent white matter of the frontal lobe (Fig. 1A). Cerebral angiography revealed a large high-flow AVM fed mainly by the lenticulostriate arteries and draining into both the basal vein and the vein of Labbé (Fig. 1B). Because there was no evidence of previous hemorrhage, and the patient presented minimal neurological deficit, she was treated conservatively. She has now been observed for an additional year and remains neurologically stable.

Discussion

Our patient's condition was characterized initially as simple writer's cramp resulting in difficulty in performing only one specific task, and then progressed to dystonic writer's cramp, by which time she was experiencing difficulties with multiple manual tasks and associated involuntary muscle spasms [8, 14, 18].

Writer's cramp is a physical disorder that is rarely associated with other progressive diseases. Approximately 5% of patients with the disorder have a positive family history of a similar condition, 5–10% of patients report local hand/arm injury preceding the onset of the symptom, and 5% subsequently develop neurological disease, such as

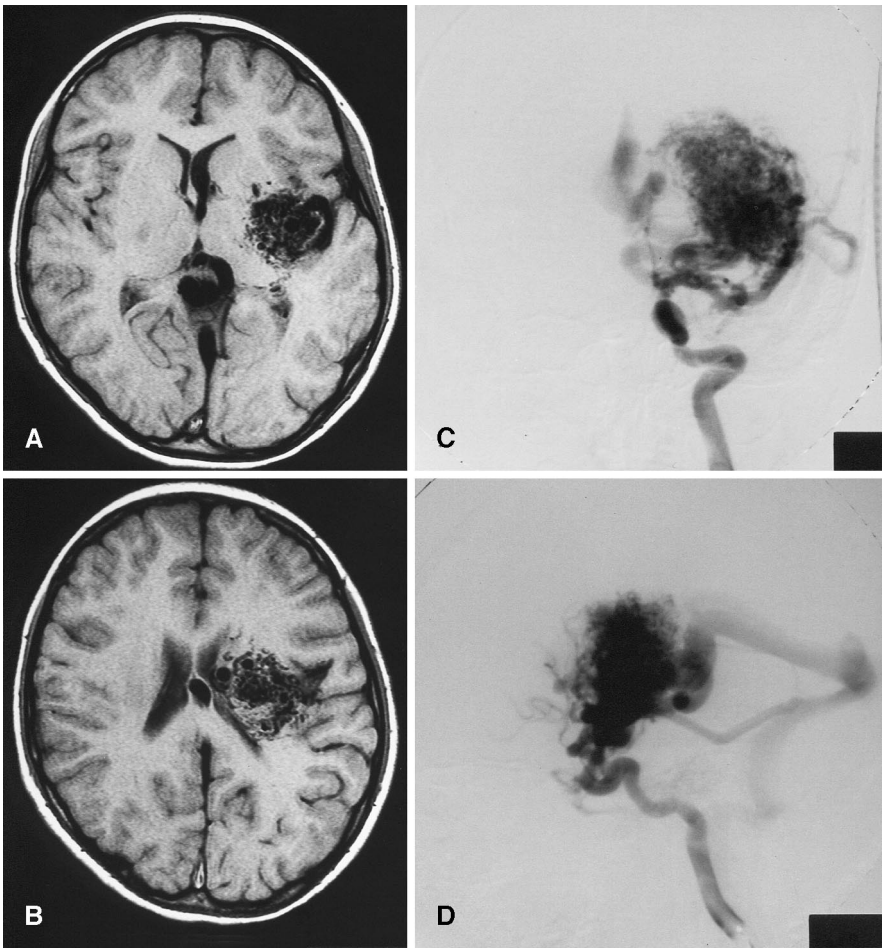

Fig. 1A–D A 12-year-old girl with a 3-year history of "writer's cramp." **A, B** T1-weighted MR images reveal an AVM localized in the left basal ganglia and adjacent white matter. **C, D** Left internal carotid angiograms demonstrate a high-flow AVM supplied mainly by the lenticulostriate arteries and draining into both the basal and cortical veins

Parkinson's disease, spinocerebellar degeneration, and multiple sclerosis [8, 14, 18]. Although unilateral dystonia is reported to be occasionally associated with structural disease, such as tumors [5, 8–11], vascular anomalies [3, 6–8, 17], infarcts [2, 4, 8, 16, 19], hemorrhages [8], and head trauma [1, 2, 4, 8], localized vascular lesion of the brain has not been reported as a cause of typical writer's cramp.

Cerebral AVMs rarely present as movement disorder [3, 6, 8, 17]. In our literature study, only 11 cases have been reported. These included 5 patients who presented with torticollis or retrocollis [6, 8, 17] and 6 patients with unilateral limb dystonia [3, 6, 8]. Marsden and Sheehy [8] reported 2 cases of hemidystonia ascribed to basal ganglia AVM, but did not describe the initial manifestation and clinical course. Lobo-Antunes et al. [6] reported 2 patients with localized dystonia of the arm due to contralateral basal ganglia-thalamic AVMs. One patient developed hemiparesis and involuntary movement of the arm after rupture of the AVM, and the other presented with tremor and progressive weakness of the arm preceding the development of athetoid posturing of the hand. Of 2 patients with AVM-induced dystonia documented by Friedman et al. [3], 1 initially presented with left hand clumsiness while playing the piano. Right basal ganglia AVM was disclosed 19 years later when he suffered spontaneous involuntary movement of the left hand. The other patient was diagnosed as having left parietal AVM 15 months after developing abnormal movement of the right arm.

Lesions involving the striatopallidothalamic connections with the premotor cortex are reported to have the most common anatomicophysiological correlation with unilateral hemidystonia [4, 8, 12, 15]. Although the mechanism of dystonia is not well understood, ischemia of the surrounding brain due to a vascular steal effect may have a causal role in the genesis of dystonia secondary to AVMs in these structures [3]. Our present case confirms the anatomical basis of focal dystonia and argues for neuroimaging in patients with "writer's cramp" that could be an initial manifestation of a focal vascular lesion.

References

1. Brett EM, Hoare RD, Sheehy MP, Marsden CD (1981) Progressive hemidystonia due to focal basal ganglia lesion after mild head trauma (letter). J Neurol Neurosurg Psychiatry 44:460
2. Demierre B, Rondot P (1983) Dystonia caused by ptamino-capsulo caudate vascular lesions. J Neurol Neurosurg Psychiatry 46:404–409
3. Friedman DI, Jankovic J, Rolak LA (1986) Arteriovenous malformation presenting as hemidystonia. Neurology 36:1590–1593
4. Fross RD, Martin WRW, Li D, Stuessl AJ, Adam MJ, Ruth T, Pate BD, Burton K, Calne D (1987) Lesions of putamen: their relevance to dystonia. Neurology 37:1125–1129
5. Krauss JK, Mohadjer M, Nobbe F, Scheremet R (1991) Hemidystonia due to a contralateral parieto-occipital metastasis: disappearance after removal of the mass lesion. Neurology 41:1519–1520
6. Labo-Antunes J, Yahr MD, Hilal SK (1974) Extrapyramidal dysfunction with cerebral arteriovenous malformations. J Neurol Neurosurg Psychiatry 37:259–268
7. Lorenzana L, Cabezudo JM, Porras LF, Polaina M, Rodriguez-Sanchez JA, Garcia-Yagüe LM (1992) Focal dystonia secondary to cavernous angioma of the basal ganglia: case report and review of the literature. Neurosurgery 31:1108–1112
8. Marsden CD, Sheehy MP (1990) Writer's cramp. Trends Neurosci 13:148–153
9. Meyrignac C, Keravel Y, Boulu P, Nguyen JP, Degos JD (1988) Crampe des ecrivains et meningiome frontal gauche. Rev Neurol (Paris) 144:378–380
10. Milanov I, Georgiev D (1995) Writer's cramp and tremor due to brain tumor. Can J Neurol Sci 22:59–61
11. Narbona J, Obeso JA, Tunon T, Martinez-Lage JM, Marsden CD (1984) Hemi-dystonia secondary to localized basal ganglia tumor. J Neurol Neurosurg Psychiatry 47:704–709
12. Obeso JA, Gimenez-Roldan S (1988) Clinicopathological correlation in symptomatic dystonia. In: Fahn S (ed) Dystonia, vol 2. (Advances in neurology, vol 50) Raven Press, New York, pp 113–122
13. Quinn N, Bydder G, Leenders N, Marsden CD (1985) Magnetic resonance imaging to detect deep basal ganglia lesions in hemidystonia that are missed by computed tomography. Lancet II 1007–1008
14. Rhoad RC, Stern PJ (1993) Writer's cramp – a focal dystonia: etiology, diagnosis, and treatment. J Hand Surg [A] 18:542–544
15. Roos RAC, Bruyn GW (1986) Symptomatic dystonias. In: Vinken PJ, Bruyn GW, Klawans HL (eds) Handbook of clinical neurology, vol 5. Elsevier, Amsterdam, pp 499–518
16. Russo LS (1983) Focal dystonia and lacunar infarction of the basal ganglia. Arch Neurol 40:61–62
17. Shaw KM, Hunter KR, Stern GM (1972) Medical treatment of spasmodic torticollis. Lancet I:1399
18. Sheehy MP, Marsden CD (1982) Writer's cramp – a focal dystonia. Brain 105:461–480
19. Traub M, Ridley A (1982) Focal dystonia in association with cerebral infarction. J Neurol Neurosurg Psychiatry 45:1073–1074