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CASE REPORT

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Brain Arteriovenous Malformation and Its Implication in Forensic Pathology*

ABSTRACT: Despite the fact that brain arteriovenous malformations (BAVMs) are a possible cause of sudden and unexplained death, very few papers have discussed their importance in the forensic context. BAVMs consist of tangled masses of tortuous arteries and veins devoid of intervening capillaries that frequently extend from brain parenchyma into the subarachnoid space. Apart from BAVMs, three major groups of vascular malformations of the brain are known: cavernous hemangioma, venous angioma, and capillary telangiectasia. BAVMs and cavernous hemangioma often cause hemorrhages, while venous angioma and capillary telangiectasia are typically asymptomatic. Presented here is the case of a 14-year-old girl who died from a ruptured BAVM. The present case is a reminder that the forensic pathologist should be able to recognize BAVMs and to differentiate it from other types of vascular malformations. Although rare, it is a cause of sudden death not to be overlooked, especially in children.

KEYWORDS: forensic science, arteriovenous malformations, sudden death

Sudden death has been defined as a natural unexpected death occurring instantaneously or, most of the time, within 1 h after the onset of symptoms (1,2). The type of conditions causing these sudden and unexpected deaths varies markedly depending on the age group. In the pediatric population, the presence of a congenital malformation, of a sequela of prematurity, and of an infection represent the most important causes of sudden death in early infancy, while in later childhood, chronic respiratory disease, infection, cerebral palsy, and epilepsy have to be considered (3,4).

Central nervous system causes of sudden and unexpected death in childhood are unusual and most often due to epilepsy, infection, or hemorrhage (4). In a very interesting study, Byard et al. (5) reviewed all of the autopsy cases in a children's hospital over a 27-year period and found a total of 10 cases of sudden and unexpected death due to hemorrhage from occult central nervous system lesions. Six of the latter were related to vascular malformations, while the remaining four were caused by primary brain tumors.

We here report the case of a sudden and unexpected death from rupture of a cerebral arteriovenous malformation in a young teenager.

Case Report

A 14-year-old girl was found dead on her bed. The girl was previously in good health, except for mild asthma. The eldest of nine children, she lived in a marginal religious community. Her parents were members of a group that allowed child beating and rejected vaccination and modern medicine.

On the morning before her death, the girl was feeling fine and went to school, as usual. Around noon, she started feeling sick, complaining of nausea and headache. She vomited twice. Her body temperature was apparently normal. Around 18:00 hours, she went to lie down on her bed where she was later found dead the next morning, her legs hanging on the side.

The girl's body was sent to a hospital pathology unit for autopsy. The pathologist was alerted by a bluish discoloration on the left side of the face and by a "traumatic hemorrhagic lesion" in the left eye. Considering the tolerance toward child beating in the girl's community, the coroner and police investigators were particularly alarmed by these possible traumatic lesions and the body was transferred to the provincial forensic laboratory for autopsy.

External exam of the 60 lb (27 kg) and 4 ft 5 in. (1.37 m) girl revealed nothing worthy of note. The bluish discoloration on the left side of the face turned out to be postmortem lividity, while the "lesion in the eye" was only a *tache noire* (i.e., blackish postmortem discoloration of the conjunctiva). There was absolutely no evidence of traumatic lesion at external and internal exam. The brain, however, showed massive edema (1580 g). On brain cut sections, an intracerebral hemorrhage with secondary necrosis was found around the left lateral ventricle, extending inside the ventricle with widening of the latter. Microscopically, the lesion was composed of different caliber thick-walled vascular channels surrounded by intervening reactive cerebral parenchyma, with gliosis and hemosiderin deposits (Fig. 1). The abnormal vessels were focally extending into the subarachnoid space. The rest of the autopsy was noncontributive, except for mild lung congestion. Toxicological analysis only revealed a therapeutic dose of acetaminophen.

Death was attributed to cerebral arteriovenous rupture, and manner of death was ruled natural.

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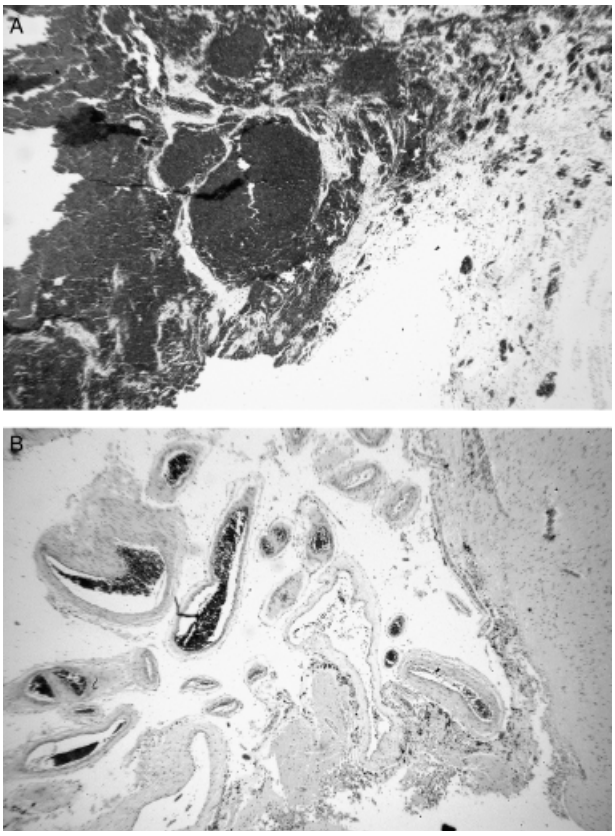

FIG. 1—*(A) Under the microscope, the initial lesion is largely destroyed by the hemorrhage (× 25). (B) Focally, residual abnormal vessels are seen: different caliber channels of different thickness surrounded by intervening reactive cerebral parenchyma, with gliosis and hemosiderin deposits (× 25).*

Discussion

Brain arteriovenous malformations (BAVMs) are congenital vascular lesions that seem to affect less than 0.7% of the population (6–8). About 20% of these BAVMs are detected in patients of <21 years of age, including 8% in children of 10 years of age or less (7,9). BAVMs discovered in childhood tend to be larger in size and are more likely to present initially with intracranial hemorrhage than those becoming symptomatic during adult life (7,9–11).

BAVMs should be differentiated from other types of brain vascular malformations: cavernous hemangioma, venous angioma, and capillary telangiectasia (Table 1) (12,13). While BAVMs and cavernous hemangioma are clinically significant causes of brain hemorrhage, venous angioma and capillary telangiectasia are mainly incidental findings.

In the forensic setting, very few articles have focused on BAVMs. Karhunen et al. (14) have reported that BAVMs were detected in 0.06% of medicolegal autopsies in Helsinki (five cases in 8038 consecutive autopsies, including the case of an 8-year-old boy with a large cerebellar hemorrhage). Apart from this study, two forensic papers by Rosen (15) and by Azzopardi (16) reported a total of five sudden deaths from cerebellar arteriovenous malformation rupture in children aged from 5 to 10 years old. Also worth mentioning is a study by Byard et al. that reviewed all sudden childhood deaths (under the age of 17 years) occurring over a 35-year period during sporting activities at the Forensic Science Center in Adelaide, South Australia. Among the 12 cases described in this study figures the case of an 8-year-old boy found unconscious in a swimming pool. Death was due to subarachnoid hemorrhage caused by a previously unsuspected cerebral arteriovenous malformation (17).

The present case is a reminder that BAVMs are a possible cause of sudden and unexpected deaths in children and therefore, forensic pathologists should be able to recognize this lesion and differentiate it from the other types of vascular malformation.

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References

1. Goldstein S. The necessity of a uniform definition of sudden coronary death: witnessed death within 1 hour of the onset of acute symptoms. Am Heart J 1982;103(1):156–9.
2. DiMaio VJ, DiMaio D. Deaths due to natural disease. In: Geberth VJ, series editor. Forensic pathology. 2nd ed. Boca Raton, FL: CRC Press, 2001:43–89.
3. Norman MG, Taylor GP, Clarke LA. Sudden unexpected, natural death in childhood. Pediatr Pathol 1990;10(5):769–84.
4. Siboni A, Simonsen J. Sudden unexpected natural death in young persons. Forensic Sci Int 1986;31(3):159–66.
5. Byard RW, Bourne AJ, Hanieh A. Sudden and unexpected death due to hemorrhage form occult central nervous system lesions. Pediatr Neurosurg 1991–1992;17(2):88–94.
6. Fleetwood IG, Steinberg GK. Arteriovenous malformations. Lancet 2002;359(9309):863–73.
7. Mohr JP. Brain arteriovenous malformations: children and adults. Stroke 2005;36(10):2060–1.

TABLE 1—*Brain vascular malformations.*

Vascular Malformation	Clinical Significance	Macroscopy	Microscopy
BAVMs	Significant cause of brain hemorrhage and subarachnoid hemorrhage	Network of wormlike vascular channels of variable calibers and thickness	Contain arteries, veins, and abnormal vessels with thin walls and a prominent internal elastic lamina or thick walls and nonelastic tissue Surrounding and intervening reactive brain parenchyma, with gliosis, old hemorrhage, calcification
Cavernous hemangioma	Significant cause of hemorrhage	Resembles small hematoma within brain parenchyma	Microscopically, tightly packed collection of hyalinized vessels lacking intervening brain parenchyma
Venous angioma	Rarely bleeds (except in the cerebellum)	Resembles petechial hemorrhages	Composed of thin-walled, dilated vascular channels lying within otherwise normal brain parenchyma
Capillary telangiectasia	Incidental finding, virtually never hemorrhagic	Bluish discoloration, most frequently in the pons	Collection of small-caliber, very thin-walled channels surrounded and separated by normal brain

BAVMs, brain arteriovenous malformations.

8. Al-Shahi R, Fang JS, Lewis SC, Warlow CP. Prevalence of adults with brain arteriovenous malformations: a community based study in Scotland using capture–recapture analysis. J Neurol Neurosurg Psychiatry 2002;73(5):547–51.
9. Stapf C, Khaw AV, Sciacca RR, Hofmeister C, Schumacher HC, Pile-Spellman J, et al. Effect of age on clinical and morphological characteristics in patients with brain arteriovenous malformation. Stroke 2003;34(11):2664–9.
10. Wilkins RH. Natural history of intracranial vascular malformations: a review. Neurosurgery 1985;16(3):421–30.
11. Fullerton HJ, Achrol AS, Johnston SC, McCulloch CE, Higashida RT, Lanton MT, et al. UCSF BAVM Study Project. Long-term hemorrhage risk in children versus adults with brain arteriovenous malformations. Stroke 2005;36(10):2099–104.
12. Ellison D, Love S, Chimelli L, Harding BN, Lowe J, Vinters HV. Hemorrhage. In: Ellison D, Love S, editors. Neuropathology: a reference text of CNS pathology. 2nd ed. London: Mosby International Ltd, 2004:209–37.
13. Frosch MP, Anthony DC, De Girolam U. The central nervous system. In: Kumar V, Abbas AK, Fausto N, editors. Robbins and Cotran pathologic basis of disease. 7th ed. Philadelphia: Elsevier Saunders, 2005:1347–419.
14. Karhunen PJ, Penttilä A, Erkinjuntti T. Arteriovenous malformation of the brain: imaging in post-mortem angiography. Forensic Sci Int 1990;48(1):9–19.
15. Rosen RS, Armbrustmacher V, Sampson BA. Spontaneous cerebellar hemorrhage in children. J Forensic Sci 2003;48(1):177–9.
16. Azzopardi A, Grech JL. Arteriovenous malformation of the cerebellum: a case report and its medicolegal implications. Forensic Sci 1974;4(1):75–82.
17. Byard RW, James RA, Gilbert JD. Childhood sporting deaths. Am J Forensic Med Pathol 2002;23(4):364–7.

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