Unusual Intramedullary Vascular Lesion: Report of Two Cases

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Abstract

OBJECTIVE AND IMPORTANCE:

Spinal arteriovenous malformations have been divided by location into dural (Type I), intramedullary glomus (Type II), juvenile (Type III), and perimedullary direct arteriovenous fistulae (Type IV). We report two cases of an unusual intramedullary proliferation of hyalinized capillaries that do not fit into any of these categories.

CLINICAL PRESENTATION:

A 27-year-old woman and a 62-year-old man presented with subacute progressive caudal myelopathy. Magnetic resonance imaging revealed focal spinal cord enlargement, high signal on T2-weighted images, and patchy enhancement with gadolinium consistent with tumor. No serpentine flow voids were visualized on the surface of the spinal cord. Spinal angiography revealed nothing abnormal. No abnormal vasculature was grossly visible on open biopsy. Histological examination of the tissue specimens revealed a proliferation of capillary-sized vessels with varying degrees of vascular wall changes ranging from endothelial hyperplasia to concentric hyalinization, suggesting ongoing evolution of the lesion. Surrounding neural tissue demonstrated ischemic changes characterized by myelin and axonal loss and astrocytosis but no necrosis.

INTERVENTION:

Patients were treated with chronic anticoagulation, which seemed to slow, but not halt, symptomatic disease progression.

CONCLUSION:

Although the pathological substrate seems to be an acquired intramedullary vascular lesion characterized primarily by capillary proliferation, the cause of this lesion is unknown. This disease differs from Foix-Alajouanine syndrome and subacute necrotizing myelopathy by an absence of abnormal surface vessels and a lack of intramedullary necrosis. The histological findings are reminiscent of the process that occurs in the kidney and various end organs from long-standing mild to moderate elevations in blood pressure or chronic diabetes. Tissue ischemia may result from luminal obstruction by severe hyalinization and thrombosis. Because the natural history of this disease is unknown, it is unclear whether anticoagulation slowed disease progression.

Key words: Arteriovenous malformations, Capillary, Hyaline, Myelopathy, Spinal cord

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Spinal arteriovenous malformations (AVMs) have been divided by location into dural (Type I) and intradural subtypes. Intradural AVMs are further subcategorized into intramedullary glomus (Type II), juvenile (Type III), and perimedullary direct arteriovenous (AV) fistulae (Type IV) (16, 19, 31, 33, 37, 40). Telangiectasias and cavernous malformations may also appear in intramedullary and extramedullary locations (16)d

In 1995, Montine et al. (29) reported three elderly patients who presented with a progressive myelopathy. Preoperative magnetic resonance imaging (MRI) and computed tomographic myelography suggested a spinal cord neoplasm in the region of the conus. Histological examination of the tissue specimens,

however, revealed abundant hyalinized capillary-sized vessels, reactive astrocytosis, perivascular hemosiderin, and occasional intraluminal thrombus. These findings are similar to the intraparenchymal changes that follow venous congestion associated with dural or perimedullary AV fistulae (Type I or Type IV) (2, 21, 31, 40). In the cases reported by Montine et al.(29), however, no abnormal vessels were visualized on the surface of the spinal cord during surgery. They concluded that this"isolated intramedullary vascular malformation" may be a separate pathological entity that must be considered in the differential diagnosis of a progressive disorder of the spinal cord in the elderly patient. No mention was made, however, of the

clinical course, pathophysiology, or possible therapeutic interventions for this disease.

We present two additional cases of this unusual spinal vascular lesion with clinical follow-up after treatment with long-term anticoagulation. We propose that although previously categorized as a type of vascular malformation, the pathophysiology of this lesion is still unknown. The first case is novel because it involves a young woman. A spinal angiogram obtained in the second case disclosed nothing abnormal, providing further evidence of the absence of extramedullary spinal cord AVMs.

## CASE REPORTS

Patient 1

History

The patient was a 62-year-old man with a 10-year history of progressive weakness and numbness in the lower extremities ascending to the L1 level and a 3-year history of sexual and bladder dysfunction. There were no visual complaints. Physical examination revealed a slight weakness of the distal lower extremities, with marked loss of pinprick, vibration, and proprioception. The results of the neurological examination of the upper extremities were normal. The results of the laboratory evaluation were within normal limits, including complete blood count, chemistry, sedimentation rate, antinuclear antibodies, anti-DNA, Lyme titers, Schilling test, serum protein electrophoresis, Venereal Disease Research Laboratory test, and human T-cell lymphoma virus-1 test. The results of a cerebrospinal fluid (CSF) examination were unremarkable, including immunoglobulins, oligoclonal bands, and Lyme titers.

Thoracolumbar MRI 3 years before presentation to The Neurological Institute of New York were significant for increased signal in the lower thoracic spinal cord and conus on T2-weighted images, extending from T9 to L1, with linear, heterogeneous gadolinium enhancement. Myelography revealed nothing remarkable. No enlargement of the spinal cord was detected. High-dose steroids only exacerbated the patient's condition. At our institution, a spinal angiogram was obtained that included bilateral injection of the intercostal arteries from T7 to T12 and bilateral injection of the lumbar arteries from L1 to L3. No abnormal vessels were identified nor was there obvious stasis in the anterior spinal artery. Repeat MRI at the same time revealed enlargement of the conus with diffuse, heterogeneous enhancement(Fig. 1, A-C).

At surgery, numerous small vessels were observed over the conus and cauda; however, they were neither engorged nor dilated. The parenchyma of the conus was diffusely xanthochromic, rubbery, and relatively avascular. Multiple biopsies were obtained.

FIGURE 1.

A T1-weighted thoracolumbar MRI showing decreased signal and enlargement of the spinal cord at T9-L1. *B*, patchy enhancement with gadolinium. *C*, T2-weighted image showing high signal within the lesion.
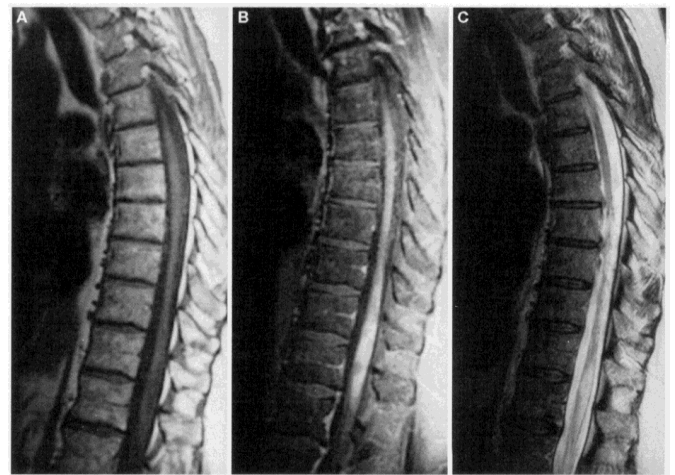
Histological findings

The resection specimen consisted of several small pieces of soft, white tissue measuring $0.4 \times 0.3 \times 0.2$ cm in aggregate. Tissue was fixed in 10% buffered formalin solution and embedded in paraffin. Sections were subjected to hematoxylin and eosin, reticulin, Masson's trichrome, Congo Red, von Kossa, Luxol fast blue-periodic acid Schiff, and Bielschowsky stains.

Microscopic examination revealed white matter displaying extensive sclerotic changes associated with numerous abnormal capillary-sized vessels. The vessels were dispersed singly and in back-to-back clusters reminiscent of capillary telangiectasias. The vessels had thick, hyalinized walls and inconspicuous endothelial lining (Fig. 2, A and B). A minimal amount of hemosiderin was identifiable surrounding some of the small vessels, and Rosenthal fibers were observed focally. The intervening neuropil was gliotic, containing scattered gemistocytic astrocytes. Disruption and loss of myelinated axons were demonstrated by special stains. No evidence of inflammation or vasculitis was observed.

Clinical management and follow-up

The patient was discharged, and a regimen of Coumadin was begun, which was discontinued after 6 months by the patient's primary care physician to reduce the risk associated with the patient's occasional falls. During the 6 months of anticoagulation, no further clinical decline was noted. Two years after the biopsy, after cessation of Coumadin, the patient reports subtle progression of his motor and sensory symptoms.

FIGURE 2.

A, spinal cord biopsy from Patient 1 demonstrating abnormal vascular structures in the neural tissue stained with Masson's trichrome (original magnification, ×20). *B*, Higher power view of single hyalinized vessels associated with adjacent cluster of abnormal proliferating capillaries. The hyalinized lesion represents a chronic change, whereas the capillary clusters show endothelial hyperplasia reflecting a more acute reaction (original magnification,×100). *C*, spinal cord biopsy from Patient 2 with tortuous vessels, some hyalinized, of varying sizes highlighted by Masson's trichrome(original magnification, ×20). *D*, congeries of vessels varying in wall thickness and lumen size (original magnification,×100).
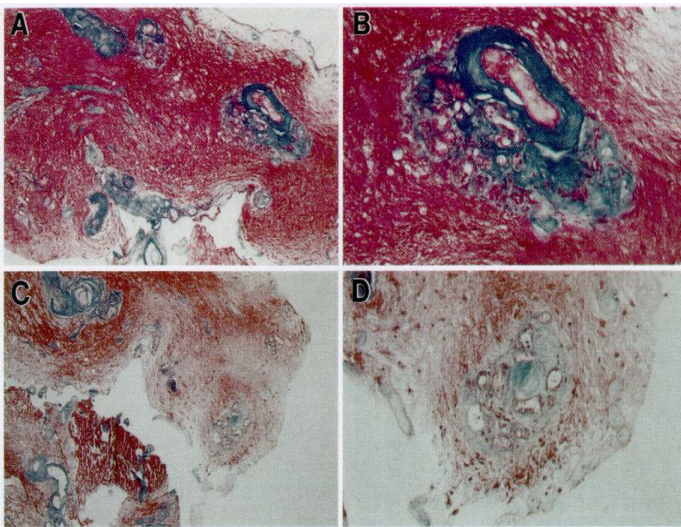

Patient 2

History

The patient was a 27-year-old woman with no significant past medical history who presented with 16 months of progressive weakness and numbness in the lower extremities ascending to a T10 level and mild stress incontinence. She denied any back pain. At physical examination, she exhibited a spastic paraparesis, decreased pinprick sensation in the lower extremities with some sacral sparing, and preserved vibration and position sense. The results of a neurological examination of the upper extremities were normal. High-dose steroids had been administered but seemed to worsen her condition.

The results of a laboratory evaluation were normal, including complete blood count, chemistry, sedimentation rate, antinuclear antibodies, anti-DNA, Lyme titers, Schilling test, serum protein electrophoresis, Venereal Disease Research Laboratory test, and human T-cell lymphoma virus-1 test. The results of a CSF examination were unremarkable, including immunoglobulins, oligoclonal bands, and Lyme titers. MRI of the head revealed nothing abnormal. Thoracic MRI revealed two intramedullary lesions, at the level of T6-T7 and T8-T9. The lower lesion was associated with a slight enlargement of the spinal cord. Both lesions exhibited high signal on T2-weighted sequences and patchy enhancement with gadolinium (Fig. 3, A-C). No abnormal vessels or flow voids were identified. The patient underwent an open biopsy.

FIGURE 3.

A, T1-weighted thoracic MRI showing enlargement of the spinal cord at T8-T9. *B*, patchy gadolinium enhancement at T6-T7 and T8-T9. *C*, T2-weighted image showing high signal in both lesions.
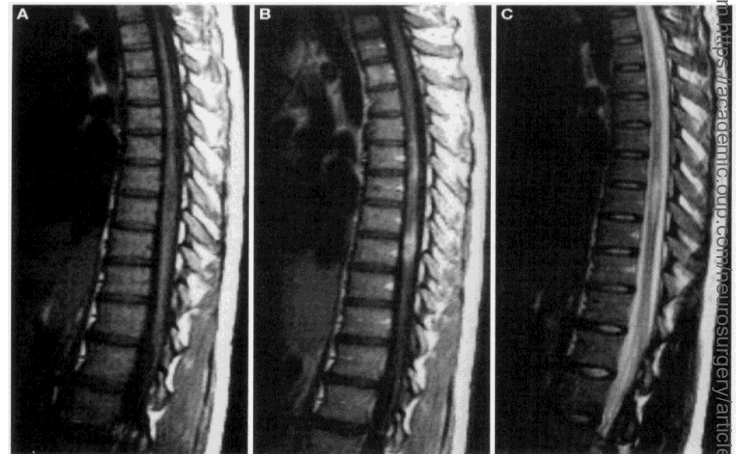
At surgery, the spinal cord was not hypervascular, although several small serpentine vessels were observed on the surface. The spinal cord seemed slightly xanthochromic with cystic degeneration in the deeper tissues. Several biopsies were obtained.

Histological findings

The biopsy specimen consisted of several small pieces of soft, white tissue measuring $0.2 \times 0.1 \times 0.1$ cm in aggregate. Tissue was fixed in 10% buffered formalin solution and embedded in paraffin. Sections were subjected to hematoxylin and eosin, reticulin, Masson's trichrome, Congo Red, von Kossa, Luxol fast blue-periodic acid Schiff, and Bielschowsky stains.

Microscopy revealed small fragments of neural tissue containing increased numbers of abnormal capillary-sized blood vessels predominantly arranged in clusters (Fig. 2, C and D). In some of the clusters, prominent hyalinization of thickened vessel walls was observed with near obliteration of lumina. Other clusters demonstrated minimal hyalinization of capillary vessel walls; however, hyperplasia and proliferation of endothelial cells were prominent. Intervening neural tissue was gliotic with secondary loss of both myelin and axons, as demonstrated by Luxol fast blue-periodic acid Schiff and Bielschowsky stain. Although rare inflammatory cells were identifiable, no evidence of vasculitis or fibrinoid necrosis was noted. Hemosiderin and Rosenthal fibers were not observed.

Clinical management and follow-up

Immediately after the biopsy, the patient experienced a slight deterioration in her neurological condition. She was placed on long-term Coumadin and, 9 months after her biopsy, has returned to her preoperative status.

DISCUSSION

We report two patients that presented with progressive caudal spinal cord myelopathy, one a 27-year-old woman and the other a 62-year-old man. MRI examination was most consistent with neoplasm, as follows: focal enlargement of the spinal cord, low signal on T1-weighted and high signal on T2-weighted images, and patchy enhancement with gadolinium. CSF profiles were within normal limits. In one patient, myelography and spinal angiography revealed nothing remarkable. At open biopsy, no abnormal vessels were identified on the surface of the spinal cord. Histological examination, however, revealed abundant hyalinized capillary-sized vessels, located uniquely within the parenchyma of the spinal cord, associated with secondary demyelination, axonal loss, gliosis, and occasional perivascular hemosiderin and intraluminal thrombus. These pathological findings are similar to the intramedullary changes that accompany an extramedullary AV fistula (Type I or Type IV) (2, 21, 31, 40). Three identical cases were presented by Montine et al. (29) and labeled "isolated intramedullary spinal vascular malformations." They noted that this pathological entity did not seem to fall into the classic definition of spinal cord AVMs and emphasized the importance of considering this unusual type of vascular malformations in the differential diagnosis of elderly patients with caudal spinal cord lesions. They did not, however, discuss the pathophysiology of this entity, nor did they provide any clinical follow-up. We placed our patients on long-term anticoagulation to prevent any thrombotic events, which may have stabilized disease progression. One of our patients has not demonstrated symptomatic deterioration after 9 months. The other patient remained stable during 6 months of anticoagulation therapy, but has progressed slightly 18 months after stopping Coumadin. We propose that rather than being a type of isolated intramedullary vascular malformation, this lesion is a unique process, the cause of which is unknown.

Vascular malformations of the central nervous system are divided into four types, i.e., AVMs, capillary telangiectasias, venous malformations, and cavernous malformations, depending on the composition of the vascular wall, the presence or absence of intervening neural parenchyma, and the state(normal or gliotic) of the intervening tissue (26). Whereas AVMs consist of abnormal AV hybrids having irregularly hyalinized walls, classically devoid of a capillary bed, with gliotic intervening neural tissue, cavernous malformations consist of back-to-back thin-walled dilated sinusoidal vessels without intervening neural tissue; telangiectasias are dilated capillaries with relatively normal intervening parenchyma. The lesion described here bears some vague histological resemblance to AVMs inasmuch as the involved vessels are frequently hyalinized and the intervening

tissue is gliotic. However, unlike AVMs, the vessels are capillary-sized, and there is neither a fistula nor dilated veins. Unlike a telangiectasia, these capillaries are not dilated, there is significant hyalinization, and, in some areas, active vessel proliferation and back-to-back clustering.

Spinal cord AVMs are further divided into four subcategories(11, 12, 16, 33, 34, 37, 40). Type I dural AV fistulae consist of one or several arterial feeders originating from a dural branch of the radiculomedullary arteries, draining into the surface venous plexus along the dorsal aspect of the spinal cord. Type II AVMs, or glomus malformations, are fed by multiple branches from the anterior spinal artery, have an intraparenchymal nidus, and drain into the coronal venous plexus surrounding the spinal cord. Type III AVMs, or juvenile malformations, are large, complex lesions with multiple feeders from many levels. They demonstrate intra- and extramedullary, and even extraspinal, extension. Type IV AVMs are intradural, perimedullary AV fistulae, usually located on the anterior aspect of the spinal cord, fed by the anterior spinal artery, and draining into an enlarged venous outflow tract. The vascular lesion presented in this article does not fall into any of the above categories, therefore it either represents a new pathological entity or an intramedullary reaction to another, anatomically separate, disease process.

In 1926, Foix and Alajouanine (5) reported two patients presenting with a progressive myelopathy associated with spinal cord necrosis and dilated tortuous vessels on the surface of the spinal cord. Since this original description, there have been numerous reports of similar cases of Foix-Alajouanine syndrome (FAS) (15, 16). Currently, most authors think that FAS is caused by a Type I dural AV fistula, a pathological entity not recognized at the time (4, 8). The intramedullary pathological changes that accompany FAS have been well described and consist of demyelination, myelomalacia, and necrosis associated with a proliferation of hyalinized capillary-sized vessels and occasional intraluminal thrombosis and luminal calcification(2, 8, 13, 21, 31, 40). These findings are thought to represent a reaction to venous hypertension, stasis, ischemia, and, ultimately, venous thrombosis. Several authors have commented regarding the ease with which these intramedullary reactive changes might be confused with a primary vascular malformation(16, 40, 40).

The intramedullary pathological changes that accompany FAS, in association with progressive myelopathy, fall under the broad category of diseases labeled subacute necrotizing myelopathy (SNM). Multiple causes have been postulated for SNM besides extramedullary spinal cord AV fistulae. Possibilities include, venous infarction associated with a hypercoagulable state, multiple sclerosis, varicella zoster, syringomyelia, toxins, systemic lupus erythematosus, paraneoplastic syndrome, pulmonary tuberculosis, and idiopathic SNM (8, 15, 20). Because most of these patients do not undergo spinal angiography, definitive elimination of spinal AVMs (FAS) is difficult (8, 20, 28). The MRI appearance of SNM has also been well characterized, i.e., focal expansion of the spinal cord, decreased or isointense T1-weighted signal, increased T2-weighted signal, and rim-like enhancement(14, 23, 27, 28, 32).

The elderly age of onset, progressive deterioration, caudal location, and intramedullary pathological appearance of the cases presented by Montine et al. (29) and in our second case are reminiscent of the characteristics of a Type I dural AV fistulae (1, 33, 34, 38). Although no abnormal vessels were identified on open biopsy, the possibility still exists that a subtle or distant AV fistula was responsible, especially because no angiograms were obtained by Montine et al. (29). Our first case, however, occurred in a young woman, which is unusual for a dural AV fistula (1, 33, 34, 38). Our second patient underwent extensive spinal angiographic evaluation and no evidence of abnormal vasculature was identified. This raises the possibility of thrombosed spinal AVMs. We think that this is unlikely because previous reports of thrombosed spinal AVMs not apparent on angiography have noted stasis in the anterior spinal artery from venous congestion (32, 39). We did not observe this phenomenon. Similarly, the MRI failed to demonstrate any serpentine areas of low signal around regions of the spinal cord not exposed at surgery, which might represent either thrombosed or dilated veins (25, 27, 32).

Another possibility is that these cases represent either idiopathic SNM, or the early stages of FAS. In 1962, Greenfield and Turner(10) presented a case of acute necrotizing myelopathy in which the intramedullary vascular changes occurred in the absence of dilated, tortuous extramedullary vessels. They postulated that the intramedullary vascular proliferation, hyalinization, and necrosis were the primary lesions, whereas the alterations in the large superficial veins occurred later. It might be argued that our cases were biopsied at an earlier stage than the cases that required autopsy for pathological data described in the older literature. There are several reasons, however, for excluding these hypotheses. Acute necrotizing myelopathy presents from days to weeks, and autopsy ensues soon after. The chronic cases present from months to years, and most patients die within 1 to 2 years of presentation from infected bedsores and sepsis (5, 10, 15). Our patients underwent biopsies at 16 months and 10 years after disease onset. Therefore, if secondary changes were to occur in the extramedullary vessels, there was adequate time.

More importantly, there are a few critical histological differences between the disease we are describing and SNM or FAS. Although both entities exhibit demyelination, gliosis, abundant hyalinized vessels, and an absence of inflammatory cells, SNM and FAS are characterized by the presence of necrosis (8, 13, 16, 20, 21, 28, 40), which was notably absent in our cases. In addition, there seems to be a more exuberant proliferation and clustering of both hyalinized and nonhyalinized capillary-sized vessels in our cases, whereas the hyalinized vessels observed in SNM and FAS represent postischemic, reactive hyperplasia of individual vessels (2, 8, 13, 16, 21, 40). This active proliferation implies an ongoing disease process, which is why the spinal cord is enlarged and not shrunken on MRI. The absence of necrosis on our pathological specimens may be just a sampling error. However, descriptions of SNM and FAS specify that the vascular proliferation is more prominent surrounding areas with the most severe necrosis (8, 15). On MRI, the lesion in SNM is hypointense on T1-weighted and hyperintense on T2-weighted images, not unlike the cases we are presenting, yet the enhancement is minimal and described as "rim-like" (28, 28, 32). In our cases, and those described by Montine et al. (29), the lesions enhance centrally in a patchy, inhomogeneous manner, not unlike a neoplasm.

The course of this disease is characterized by slow progression; however, the pathophysiology is unknown. Although small amounts of perivascular hemosiderin were present in the histological specimens, there was not sufficient quantities to indicate significant hemorrhage. Similarly, although no gradient-echo sequences were obtained, MRI examination did not reveal the signal characteristics of recent hemorrhage nor was there blood grossly visible on open biopsy. Therefore, we do not think that hemorrhage contributed significantly to clinical deterioration.

Another possible cause for the symptomatic progression is "arterial steal"(18, 22, 36). For this phenomenon to occur, there must be a common blood supply, distal to the aorta, which is shared by both the spinal cord and the malformations (2, 18). Although this is rare in most spinal cord AVMs(18), these intramedullary vascular lesions are distal, deep within the parenchyma, and most likely share a common blood supply with the surrounding spinal cord. However, the high vascular resistance of a capillary bed with concentric hyalinization would divert little blood from the surrounding spinal cord.

A more likely possibility is ischemia from concentric hyalinization and luminal obliteration of the capillaries. Thrombosis may occasionally appear in severely narrowed vessels. The hyalinization is similar to the process that occurs in the kidney and various end organs from long-standing mild to moderate elevations in blood pressure or chronic diabetes. Chronic damage to small vessels permits the transudation of plasma proteins through leaky endothelia. Slow spontaneous activation of the alternative complement pathway ensues and C3, factor H, and immunoglobulin M bind to the hyaluronic acid in the endothelial wall (9). Accumulation of these plasma proteins, as well as thickening of the basement membrane, leads to hyalinization and tissue ischemia. With more severe cases of malignant hypertension, fibrinoid necrosis and hyperplastic arteriolitis ensue(3, 9, 24, 30). In FAS and SNM, fibrinoid necrosis appears in the severely damaged vessels in the necrotic areas. Radiation myelopathy induces similar changes in the spinal cord from damage to endothelial cells, although the demyelination and necrosis of the surrounding parenchyma may be caused as much by the direct effects of radiation, as ischemia from luminal obliteration(17).

The intramedullary pathological changes in FAS or SNM begin with an insult to vascular endothelia, caused either by venous hypertension secondary to a disease of the superficial venous channels or an intraparenchymal toxic/metabolic/infectious insult. The end result is fibrinoid necrosis of the intra- and extramedullary vasculature and thrombotic occlusion. The resulting ischemic/necrotic tissue induces the proliferation of a reactive neovascularity in adjacent tissue. These newly formed capillaries permit passage of plasma proteins, resulting in hyalinization(35). In the disease we describe, we hypothesize that the primary abnormality is the proliferation of capillaries with

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leaky endothelia. Transudation of plasma proteins induces hyalinization. Progressive luminal obliteration results in ischemia. Intermittent thrombotic events exacerbate the ischemia. This would explain why anticoagulation might slow, but not halt the progression of this disease. However, because the natural history of this disease is unknown, we cannot definitively say whether chronic anticoagulation had any effect.

The cause of the vascular proliferation remains obscure. It is unlikely that the lesion is congenital, because most patients present late in life. Angiogenesis has been linked with ischemia, inflammation, and neoplastic release of numerous growth and inhibitory factors (6, 7). However, evidence of any ischemic/toxic/infectious/neoplastic insult was lacking in our cases. Our tissue samples consisted of formalin-fixed, paraffin-embedded tissues, but if future cases undergo appropriate immunological analysis using frozen tissues and electron microscopic examination using glutaraldehyde fixed materials, the cause of this unusual lesion may be forthcoming. Although avoiding a biopsy altogether might be preferential, the clinical presentation and radiographic appearance of this lesion mimics an intramedullary tumor so well as to make preoperative identification nearly impossible. If a lesion resembling those described in this report is encountered, after appropriate expectant management, biopsy needs to be seriously considered, because it did not seem to hasten disease progression. Future therapeutics presently unavailable may someday offer treatment for this unusual lesion.

REFERENCES

1. Aminoff MJ, Logue V Clinical features of spinal vascular malformations. Brain 97:197–210, 1974.

2. Aminoff MJ, Barnard RO, Logue V The pathophysiology of spinal vascular malformations. J Neurol Sci 23:255–263, 1974.

3. Bollinger A, Frey J, Jager K, Furrer J, Seglias J, Siegenthaler W Patterns of diffusion through skin capillaries in patients with long-term diabetes. N Engl J Med 307:1305–1310, 1982.

4. Criscuolo GR, Oldfield EH, Doppman JL Reversible acute and subacute myelopathy in patients with dural arteriovenous fistulas. J Neurosurg 70:354–359, 1989.

5. Foix C, Alajouanine T La myélite nécrotique subaigue. Rev Neurol 2:1–42, 1926.

6. Folkman J Clinical applications of research on angiogenesis. N Engl J Med 333:1757–1763, 1995.

7. Folkman J, Shing Y Angiogenesis. J Biol Chem 267:10931–10934, 1992.

8. Folliss AGH, Netsky MG Progressive necrotic myelopathy, in Vinken PJ, Bruyn GW (eds) Handbook of Clinical Neurology. Amsterdam, North-Holland, 1970, pp 452–468.

9. Gamble CN The pathogenesis of hyaline arteriolosclerosis. Am J Pathol 122:410–420, 1986.

10. Greenfield JG, Turner JWA Acute and subacute necrotic myelitis. Brain 62:227–252, 1939.

11. Guegen B, Merland JJ, Riche MC, Rey A Vascular malformations of the spinal cord: Intrathecal perimedullary arteriovenous fistulas fed by medullary arteries. Neurology 37:969–979, 1987.

12. Heros RC, Debrun GM, Ojemann RO, Lasjaunias PL, Naessens PJ Direct spinal arteriovenous fistula: A new type of spinal AVM. J Neurosurg 64:134–139, 1986.

13. Hughes JT Disorders of the spine and spinal cord, in Adams JH, Duchen LW (eds) Greenfield's Neuropathology. New York, Oxford University Press, 1992, pp 1083–1116.

14. Isu T, Iwasaki Y, Akino M, Koyanagi I, Abe H Magnetic resonance imaging in cases of spinal dural arteriovenous malformation. Neurosurgery 24:919–923, 1989.

15. Jaffe D, Freeman W Spinal necrosis and softening of obscure origin. Arch Neurol Psychiatry 49:683–707, 1943.

16. Jellinger K Vascular malformations of the central nervous system: A morphological overview. Neurosurg Rev 9:177–216, 1986.

17. Jellinger K, Sturm KW Delayed radiation myelopathy in man: Report of twelve necropsy cases. J Neurol Sci 14:389–408, 1971.

18. Kaufman HK, Ommaya AK, Di Chiro G, Doppman JL Compression vs "steal:" The pathogenesis of symptoms of arteriovenous malformations of the spinal cord. Arch Neurol 23:173–178, 1970.

19. Kendall BE, Logue V Spinal epidural angiomatous malformations draining into intrathecal veins. Neuroradiology 13:181–189, 1977.

20. Kim RC Necrotizing myelopathy. AJNR Am J Neuroradiol 12:1084–1086, 1991.

21. Koeppen AH, Barron KD, Cox JF Foix-Alajouanine syndrome. Acta Neuropathol (Berl) 29:187–197, 1974.

22. Krayenbuhl H, Yaşargil MG, McClintock HG Treatment of spinal cord vascular malformations by surgical excision. J Neurosurg 30:427–435, 1969.

23. Lahanis S, Vlahos L, Gouliamos A, Papavasilou C Arteriovenous malformation of the spinal cord mimicking tumor. Neuroradiology 35: 598–599, 1993.

24. Leitschuh M, Chobanian A Vascular changes in hypertension. Med Clin North Am 71:827–841, 1987.

25. Masaryk TJ, Ross JSM Radiculomeningeal vascular malformations of the spine: MR imaging. Radiology 164:845–849, 1987.

26. McCormick WF The pathology of vascular("arteriovenous") malformations. J Neurosurg 24:807–816, 1966.

27. Minami S, Sagoh T, Nishimura K, Yamashita K Spinal arteriovenous malformation: MR imaging. Radiology 169:109–115, 1988.

28. Mirich DR, Kucharcyzk W, Keller MA, Deck J Subacute necrotizing myelopathy: MR imaging in four pathologically proved cases. AJNR Am J Neuroradiol 12:1077–1083, 1992.

29. Montine TJ, O'Keane JC, Eskin TA, Giangaspero F, Gray L, Friedman AH, Burger PC Vascular malformations presenting as spinal cord neoplasms: Case report. Neurosurgery 36:194–198, 1995.

30. Nag S, Robertson DM, Dinsdale HB Intracerebral arteriolar permeability to lanthanum. Am J Pathol 107:336–341, 1982.

31. Oldfield EH Venous congestion of the spinal cord: A reversible mechanism of myelopathy, in Holtzman RNN, Stein

BM (eds)Surgery of the Spinal Cord: Potential for Regeneration and Recovery. New York, Springer-Verlag, 1992, pp 277–300.

32.Renowden SA, Molyneux AJ Case report: Spontaneous thrombosis of a spinal dural AVM (Foix-Alajouanine syndrome)-Magnetic resonance appearance. Clin Radiol 47:134–146, 1993.

33.Rosenblum BR Current concepts of spinal arteriovenous malformations, in Rosenblum, BR (ed) Cerebral and Spinal Arteriovenous Malformations. Philadelphia, Hanley & Belfus, 1988, pp 43–60.

34.Rosenblum B, Oldfield EH, Doppman JL, Di Chiro G Spinal arteriovenous malformations: A comparison of dural arteriovenous fistulas and intradural AVMs in 81 patients. J Neurosurg 67:795–802, 1987.

35.Schoefl GI Studies of inflammation: III-Growing capillaries: Their structure and permeability. Virch Arch Pathol Anat 337:97–125, 1963.

36.Shepherd R Some new concepts in intradural spinal angioma. Riv Patol Nerv Ment 86:276–283, 1965.

37.Symon L, Kuyama H, Kendall B Dural arteriovenous malformation of the spine: Clinical features and surgical results in 55 cases. J Neurosurg 60:238–247, 1984.

38.Tobin WD, Layton DD The diagnosis and natural history of spinal cord arteriovenous malformations. Mayo Clin Proc 51:637–646, 1976.

39.Wrobel CJ, Oldfield EH, Di Chiro G, Tarlov EC, Baker RA, Doppman JL Myelopathy due to intracranial dural arteriovenous fistulas draining into spinal medullary veins: Report of three cases. J Neurosurg 69:934–939, 1988.

40.Yaşargil MG, Symon L, Teddy PJ Arteriovenous malformations of the spinal cord, in Symon L, Brihaye J, Guidetti B, Loew F, Miller JD, Pasztor E, Pertioset B, Yaşargil MG (eds) Advances and Technical Standards in Neurosurgery. Vienna, Springer-Verlag, 1984, pp 62–99.

COMMENTS

The authors report two interesting patients and add this experience to a previous series published in 1995. These patients presented with progressive myelopathy and developed relatively caudal spinal cord abnormalities that, on biopsy, proved to be an unusual and apparently novel type of vascular lesion. These lesions were within the substance of the spinal cord and consisted of hyalinized capillary-sized vessels, secondary demyelination, gliosis, axonal loss, perivascular hemosiderin, and intraluminal thrombosis. The authors' discussion makes a compelling argument that this is a vascular lesion. They hypothesize that a proliferation of capillaries with leaky endothelia lead to transudation of protein from the plasma, which leads to hyalinization and luminal obliteration and ischemia.

Unfortunately, as the authors point out, the cause, pathophysiological features, and natural history of this condition are unknown. Similarly, the treatment options for the physician are unclear. Imaging characteristics do not seem to be distinguishable from certain types of intramedullary tumor, and, therefore, in the face of progressive symptoms, biopsy seems warranted. Although anticoagulation may have stabilized the course of the condition in a patient in this series, significant experience would be required to determine whether this is an effective therapy.

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The authors recognize that the intramedullary vascular lesions reported are not lesions that can be classified as the more commonly recognized arteriovenous malformations. It is not surprising that these lesions were not frequently recognized before the more common use of magnetic resonance imaging, because often the spinal cord may not be enlarged and myelography would reveal nothing remarkable. Even where clinical examination presented a high index of suspicion for an intramedullary lesion, angiography after myelography may also not demonstrate the particular lesion. Furthermore, the surgical findings are nonspecific, and the appearance of the spinal cord may be either normal or demonstrate signs of previous hemorrhage with xanthochromia on the surface and, occasionally, some suggestion of excessive surface vasculature. Invariably, no specific mass lesion is identified, and the surgical procedure needs to be confined to simply obtaining small biopsy specimens, as was performed in these two cases.

It is possible that these abnormalities are extremely rare because of the frequency of magnetic resonance imaging scanning and the relative rarity of this reported surgical finding. Diagnosis is actually dependent on biopsy, which, as mentioned, is likely to demonstrate hyalinized capillary-sized vessels with intervening gliotic tissue. Although the authors present some considerations for the cause of this entity, the true cause or causes are unknown. Similarly, there is some reason to expect that treatment with Coumadin may be of benefit. The rarity of this abnormality and a lack of good understanding of the natural history of this rare lesion do not allow us to know how beneficial anticoagulation therapy is. Nevertheless, the infrequency of true hemorrhages in these lesions suggests that the risk of hemorrhage-induced complications secondary to anticoagulation therapy would be low.

In summary, it is important for the clinician and the neuroradiologist to be aware of these lesions because these abnormalities may occasionally be observed on spine magnetic resonance imaging. Even an incidental detection of this entity might be important for a meaningful differential diagnosis.

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