

Acta Medica Okayama

Volume 45, Issue 6

1991

Article 7

DECEMBER 1991

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Abstract

An autopsy case of spinal arteriovenous malformation (AVM) was reported. The patient was a 75-year-old male and his initial neurologic symptoms were paraplegia, paresthesia below the umbilical level and urination difficulty. Subsequently night delirium and parkinsonism also appeared. The clinical and pathological findings in this case are identical with those in the spinal AVM except for Parkinson's disease. In addition, the lateral funiculus of the spinal cord in the middle thoracic segment showed pallor: Under light microscopy, the funiculus was spongiform, with a thinner wall of the myelin sheath, enlargement of the axon and the perivascular infiltration of phagocytes without plasma exudation. The changes in the lateral funiculus seemed to indicate early congestive changes.

KEYWORDS: Foex-Alajouanine syndrome, mid-thoracic lesion, early congestive change

An Autopsy Case of Spinal Arteriovenous Malformation (Foix-Alajouanine Syndrome)

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An autopsy case of spinal arteriovenous malformation (AVM) was reported. The patient was a 75-year-old male and his initial neurologic symptoms were paraplegia, paresthesia below the umbilical level and urination difficulty. Subsequently night delirium and parkinsonism also appeared. The clinical and pathological findings in this case are identical with those in the spinal AVM except for Parkinson's disease. In addition, the lateral funiculus of the spinal cord in the middle thoracic segment showed pallor: Under light microscopy, the funiculus was spongiform, with a thinner wall of the myelin sheath, enlargement of the axon and the perivascular infiltration of phagocytes without plasma exudation. The changes in the lateral funiculus seemed to indicate early congestive changes.

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Foix-Alajouanine syndrome (F-A syndrome) is now considered to be due to an arteriovenous malformation (AVM) of the spinal cord (1-4), and recent progress in imaging techniques has resulted in an increase in the number of patients diagnosed as having this syndrome (7-11). However, patients whose nidi are not well visualized by imaging, or those showing atypical clinical findings may be erroneously diagnosed. We describe a patient who was followed up for cerebrospinal disorder of unknown diagnosis and was diagnosed as having spinal AVM at autopsy.

Case Report

The patient was a 75-year-old male. At the age of 70, the patient felt pain and weakness in both legs, and he was admitted to a hospital. After 2 months of hospitalization, waste of the legs improved slightly, but paresthesia (numbness) remained unchanged. At the age of 71, he was again admitted because of exacerbated gait disturbance preventing him from walking by himself, ascension of numbness to the abdominal level and dysuria. His consciousness was clear. The fundi and extraocular movement were normal. Swallowing was not smooth, and speech was unclear. There was no rigidity in the neck. The upper limbs showed no abnormality of the myotonus, muscle strength or sensorium. The

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legs were flaccid, hypotonic and paralytic, scoring 2-3 in the Daniels scale. Sensitivity to touch, temperature and vibration were decreased below the umbilical level. While the upper limbs showed normal tendon reflexes, there was no patellar reflex or Achilles tendon reflex in the lower limbs, with no pathologic reflex. Diffuse muscular atrophy was noted in both legs. There was no fasciculation. Sphincter disturbance was present,

causing prolonged urination and constipation. Urinalysis, routine hematology or blood biochemistry revealed no abnormality. Examination of the cerebrospinal fluid (CSF) revealed a normal pressure, 150-90 mm H₂O. The cell count was 7/mm³, but the protein content was markedly increased to 410 mg/dl, being albuminocytologic dissociation. Myelography revealed a slow passage of the contrast material without any space-

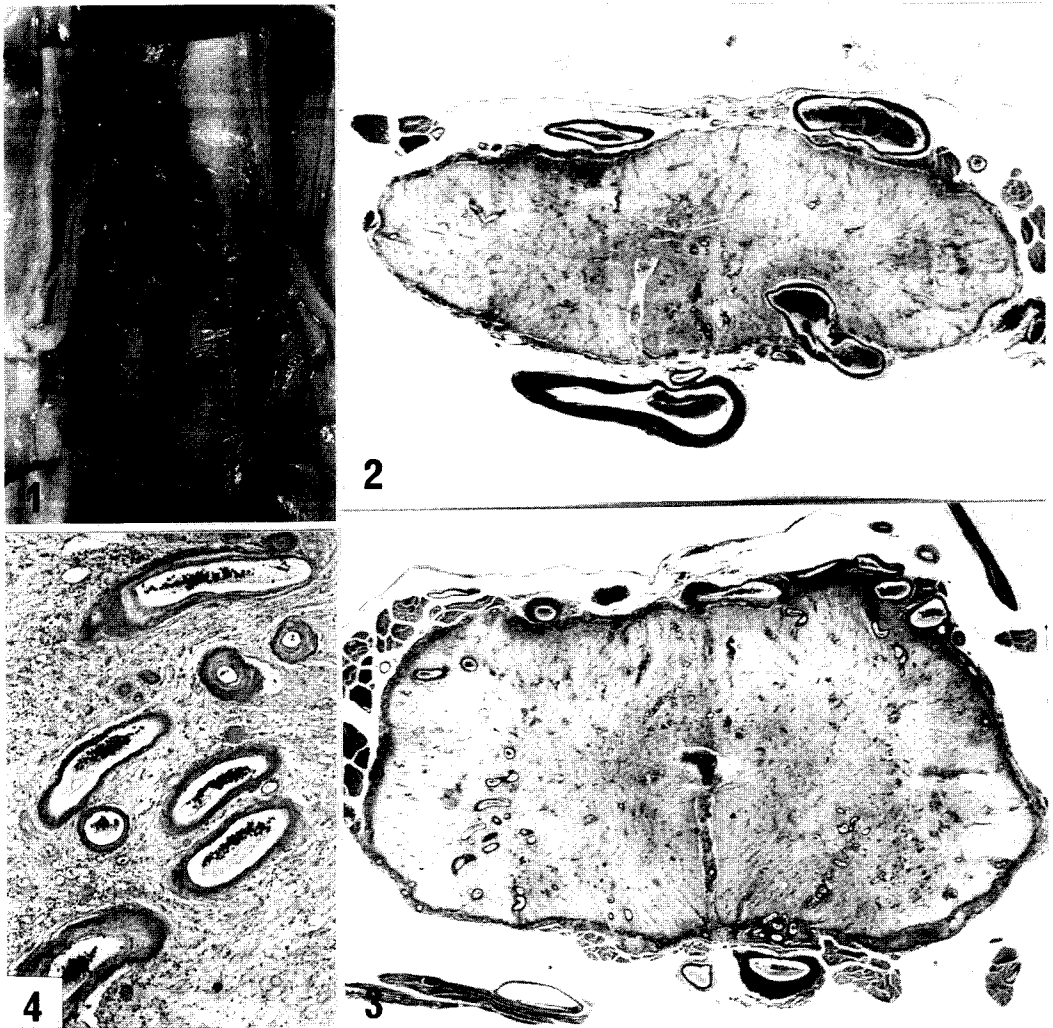


Fig. 1 Abnormally dilated and tortuous blood vessels on the posterior surface of the thoracic spinal cord.

Figs. 2 (Th12) and 3 (L4) The spinal cord is completely necrotic with dilated blood vessels outside and a large number of blood vessels of small calibers inside. Azan staining, x10

Fig. 4 The intramedullary blood vessels show thickened walls and luminal dilatation. L4, elastica van Gieson, x60.

occupying lesion. Nocturnal delirium appeared after one week of hospitalization. Muscular weakness of the legs progressed and he was unable to urinate, requiring urine catheterization. Bedsores developed. The CSF test conducted in the 7th hospital month showed a decrease in the protein content to 88 mg/dl and a normal cell count, 4/mm³. At that time, muscle rigidity was recognized in the neck and upper limbs, and dysarthria became apparent. Muscular atrophy of the legs progressed, and the patient complained of pain in the legs all day long. Paraplegia of both legs, delirium and muscle rigidity gradually progressed. During 8th month in hospital, disorientation and memory disturbance increased in severity, accompanied by exacerbation of muscle atrophy of the legs. Prednisolone was administered at the highest dose of 30 mg/day for 2 months, but it was ineffective. These symptoms gradually exacerbated until the age of 74 (4 years after onset) when he became unable to eat, requiring pernasal nutrition. He died of pneumonia at the age of 75.

The brain weighed 1,265 g. Macroscopically, the cerebral hemispheres showed no marked changes in appearance or on the cut surface. The substantia nigra in the midbrain was depigmented. In the spinal cord, the vein on the posterior surface was engorged and tortuous starting at Th5. These changes were milder at Th8-10, but were marked at Th10-L2 (Fig. 1). On the anterior surface, an engorged abnormal blood vessel distinguishable from, and parallel to, the anterior spinal artery, was present in Th4-7. A markedly swollen blood vessel originated in the right anterior funiculus of spinal cord and gradually moved toward the center during the descending course. On the cut surface, the spinal cord preserved its original form and became necrotic from the lower segment of the thoracic region to sacral region (Fig. 5).

Light microscopic findings: The cerebral cortex showed no marked changes, except for a small number of neurofibrillary tangles in the parahippocampal gyrus and the amygdalae with-

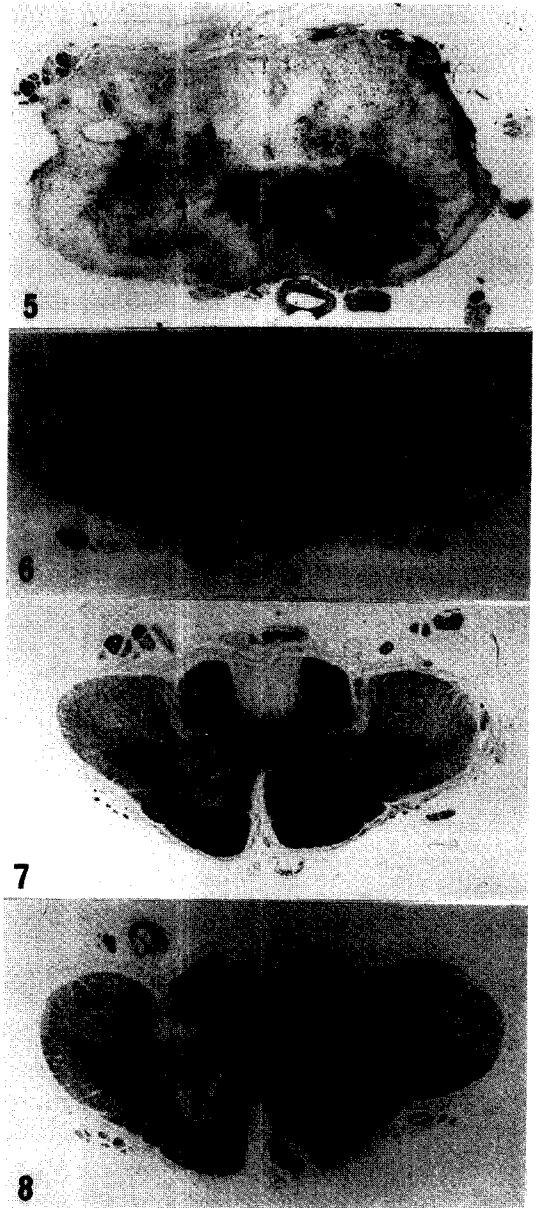


Fig. 5 Complete transverse necrosis. L4, Klüver-Barrera (KB) staining, x8

Fig. 6 Small amounts of myelin sheath remain in part of the lateral and anterior funiculi. Th10, KB staining, x10

Fig. 7 The lateral corticospinal tract (pyramidal tract) is vacuolated, and the fasciculus gracilis shows secondary changes. Th6, KB staining, x10

Fig. 8 The lateral corticospinal tract has become degenerated, but less than at Th6. Th4, KB staining, x10

out senile plaques. A small number of cortical type of Lewy bodies were found in the parahippocampal gyrus, the insula and the cingulate. In the substantia nigra, there was a moderate decrease in the nerve cells, and Lewy bodies of brain-stem type were present in the cytoplasm of the remaining neurons. Brain-stem type Lewy bodies were also observed in the substantia innominata, nucleus of the oculomotor nerve, locus caeruleus, raphe and dorsal nucleus of vagal nerve.

Vascular changes in the spinal cord: Two changes, luminal dilatation and wall thickening, were noted. Marked extramedullary vascular changes consisting of luminal dilatation and wall thickening were observed in the segment from the lower thoracic to sacral cord (Figs. 2 and 3), but luminal dilatation alone was present in Th 4 and 6. Intramedullary vascular changes also included both types of changes in the severely affected segment (Fig. 4), but luminal dilatation was mild in Th 8 and 10 without wall thickening in the anterior funiculus of Th 8. Neither of these changes was present in Th 6. Most of the thickened blood vessels had no internal elastic lamina and appeared to be a vein, venule or capillary. The vascular walls were eosinophilic in hematoxylin-eosin staining, light pink or reddish purple in elastica van Gieson staining, blue in Azan staining and pink-purple in silver impregnation (PAP). The vascular walls were negative in anti-beta protein staining. Most of the thickened walls hyalinized, but some showed fibrinoid degeneration. Most of the lumina were narrowed or occluded, but some were dilated. There was little glial or phagocytic reaction in these abnormal blood vessels. Vascular thickening and hyalinization were also noted in the blood vessels in the roots of the lower thoracic to sacral cord.

Parenchymal changes in the spinal cord: The white and gray matters of the spinal cord below the middle thoracic level had lost the structures, being present within the trabecula of the connective tissue, with only part of the original structures of the anterior funiculus and the external

margin of the spinal cord remaining. There was precipitation of fibrin and colloid in the lumbar spinal cord. The gray matter no longer contained the stroma, but preserved anterior horn cells. While some of these cells appeared normal, many showed central chromatolysis, hypertrophy of the processes or spheroids. In Th4-6 showing a slight lesion, the lateral pyramidal tract showed spongy demyelination (Figs. 7 and 8). Each myelin sheath was enlarged, without or with only a trace of wall, being stained light and clear. Most axons were enlarged. Numerous phagocytes aggregated in the vascular wall and were distributed inside the lateral funiculus (Fig. 9). Perivascular exudation was absent. The more distant from the nidus, the less marked were the changes in the lateral funiculus (Figs. 5-8): Vacuolation of the lateral funiculus was even milder in Th2, but was more marked on the left side. There was perivascular phagocyte infiltration. In C8 the lateral funiculus was vacuolated, without perivascular phagocyte infiltration. From the cervical cord to upper thoracic cord, the fasciculus gracilis showed secondary degeneration including disappearance of both the myelin sheath and axon, hyalinization of the vascular wall and phagocyte infiltration. The lateral spinothalamic tract showed spongy degeneration and axonal

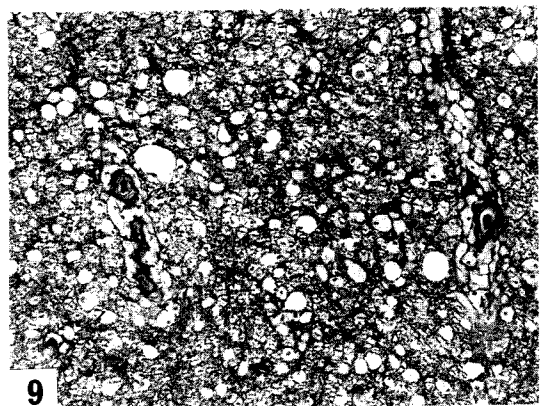


Fig. 9 The parenchyma is vacuolar, and the wall of the myelin sheath has become thinner. Azan staining, x150.

swelling with a few phagocytic infiltration. The posterior spinocerebellar tract looked pale, and showed only a mild demyelination.

Discussion

In comparison with the clinical picture of F-A syndrome (1-4), coexistence of organic mental syndrome and parkinsonism led to an erroneous diagnosis. The neurologic symptoms improved slightly at first, but progressed slowly. The age at initial onset was 70. These factors are rather atypical of F-A syndrome. Symon *et al.* (5) divided spinal AVM into dural AVM's and spinal cord angioma in describing clinical findings and surgical efficacy. Rosenblum *et al.* (6) reviewed 81 cases of spinal AVM and divided them into dural arteriovenous fistulas and intradural AVM to describe the respective characteristics. Dural AVM's reported by Symon *et al.* is thought to be identical to dural AV fistulas reported by Rosenblum *et al.* Spinal AVM is characterized by onset at middle age, symptoms including progressive paraplegia, the absence of subarachnoid hemorrhage, aggravation of symptoms when the patient moves and the localization of the nidus, which is the lower thoracic to lumbar spinal cord. Many authors state that these characteristics are identical to those of F-A syndrome. The pathological findings of our case were consistent with those of reported cases of F-A syndrome (1, 12-18). However, since we did not suspect spinal AVM at autopsy, we did not examine the extrabural blood vessels in detail.

The changes in the lateral pyramidal tract of the middle thoracic spinal cord seemed to be noteworthy. As mentioned in light microscopic findings, the wall of the myelin sheath had become thin, with axonal swelling. In addition, phagocytes were present inside the lateral funiculus and perivascular area. These changes are identical to those reported by Jellinger and Sturm (19) as early changes in radiation necrosis. We also found the same changes in the lateral

funiculus of the upper thoracic spinal cord which are fairly away from the nidus in our patient with radiation necrosis. Necrosis in radiation necrosis of the spinal cord is attributed to injuries to vascular endothelial cells due to radiation, whereas in spinal AVM, to a disturbed venous return due to congestion. Although radiation necrosis and spinal AVM have different etiological causes, both of them show congestion, and the changes in the lateral funiculus are probably the result of early congestive changes. As Criscuolo *et al.* (20) argued, venous congestion is likely to be present in the early stage of spinal AVM, that this disease must be taken into consideration as a differential diagnosis for myelopathy in individuals of middle or older age, especially in males.

Acknowledgments. The authors wish to thank Dr. H. Fujita (Jikei Hospital) for reviewing our manuscript.

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Received June 26, 1991; accepted August 16, 1991.