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Arteriovenous malformation of the spinal cord

Three-dimensional reconstruction of the intramedullary abnormal vessels

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Summary. We report a case of spinal arteriovenous malformation (AVM) in which detailed morphological examination of the intramedullary vascular lesion was carried out employing serial section studies. The relationship of the malformation to parenchymal lesions was evaluated. An abnormal vessel at the spinomedullary transition, whose lamina elastica was partially interrupted, was suggestive of a shunt vessel. The location of the intramedullary abnormal vessels and foci of softening of the spinal cord almost overlapped in distribution. Three-dimensional reconstruction of the intramedullary abnormal vessels demonstrated marked elongation and tortuosity of the original vascular architecture and they ended in the extramedullary abnormal vessels with thickened wall. Thus, our study showed that the intramedullary abnormal vessels were the result of secondary changes subsequent to pressure and volume overload inflicted upon the spinal venous system through AVM. Compression by tortuous elongation of the intramedullary vessels is the apparent cause of the parenchymal softening. A traumatic factor in the aetiology of the AVM was also discussed, since the patient had had two preceding episodes of traffic accidents with cranial and lumbar injury. Trauma seemed to be significant factor as a trigger of symptoms.

Key words: Arteriovenous malformations – Spinal cord – Three-dimensional reconstruction – Vascular tortuosity – Trauma

Introduction

Spinal vascular malformations have received attention in the literature since Hebold's first report in 1885 (Hebold 1885; Gaupp 1888; Lindemann 1912; Nonne 1913). Foix and Alajouanine (1926) described detailed pathology of the disease which is often referred to after their names, and vascular abnormalities per se have been considered to be responsible for the spinal cord degeneration seen (Greenfield and Turner 1939; Stolze 1950; Scholz and Manuelidis 1951; Brion et al. 1952; Bodechtel and Erbslöh 1957; Jellinger et al. 1968; Jellinger 1978).

The technical advance in the field of neuroradiology, especially selective arteriography of the spinal cord, and the development of surgical treatments of vascular malformations of the spinal cord have established that Foix-Alajouanine's disease is an arteriovenous malformation (AVM) (Brion et al. 1952; Djindjian 1963; DiChiro et al. 1971; Doppmann 1971; Yasargil 1971; DiChiro et al. 1973; Kendall et al. 1977; Logue 1979; Oldfield et al. 1983; Doppmann et al. 1985; Riche et al. 1985; Heros et al. 1986; Théron et al. 1986).

We examined the vascular and parenchymal lesions of a spinal AVM in an autopsy case with serial section study along with three-dimensional reconstruction of the intramedullary abnormal vessels and found that elongation and tortuosity of the original parenchymal vasculature were the essential alteration of intramedullary vascular abnormality accompanying the spinal AVM.

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Case report

A 75 year old man was admitted to the Department of Neurology, University of Tokyo Hospital, in July 1980, because of progressive gait disturbance and dysuria. He had been involved in two traffic accidents at the ages of 60 (in 1965) and 72 (in 1977) and sustained a cranial and a lumbar injury, respectively. He noticed sensory disturbance in the left toes, gradually spreading to the whole left lower extremity in August 1978, ten months after the last accident. The sensory disturbance

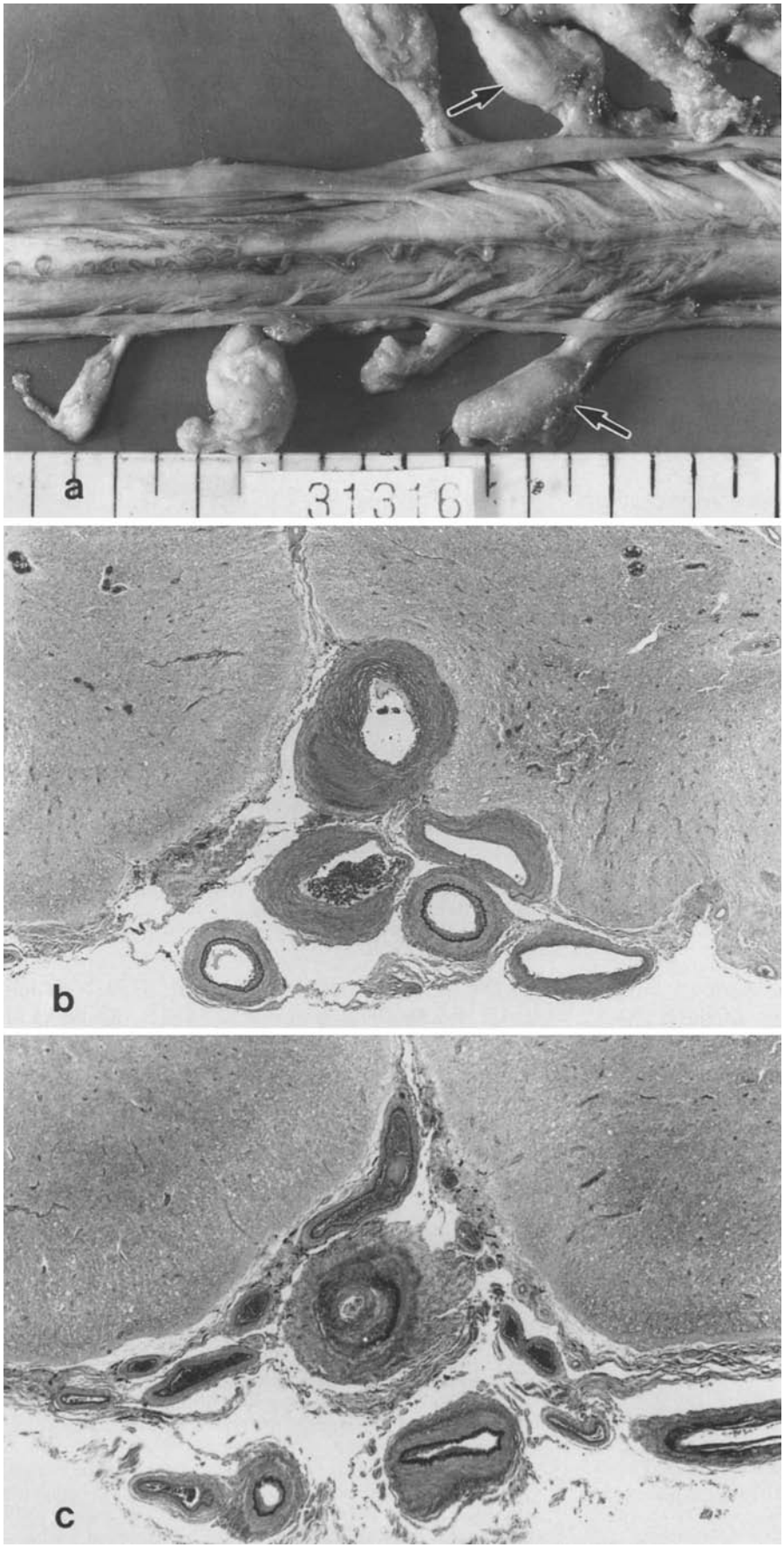

Fig. 1.a Tortuous vessels of the ventral surface of the lower cervical and the upper thoracic spinal cord. *Arrows* indicate the dorsal root ganglia of C8.
b Extramedullary abnormal vessels (4 cross sections) at C7 showing dilataion and irregular thickening of their walls. Two cross sections of artery are present. Elastica van Gieson stain, ×30.
c An abnormal vessel with thickened intima and partially interrupted its lamina elastica interna at the spinomedullary transition. Elastica van Gieson stain, ×30

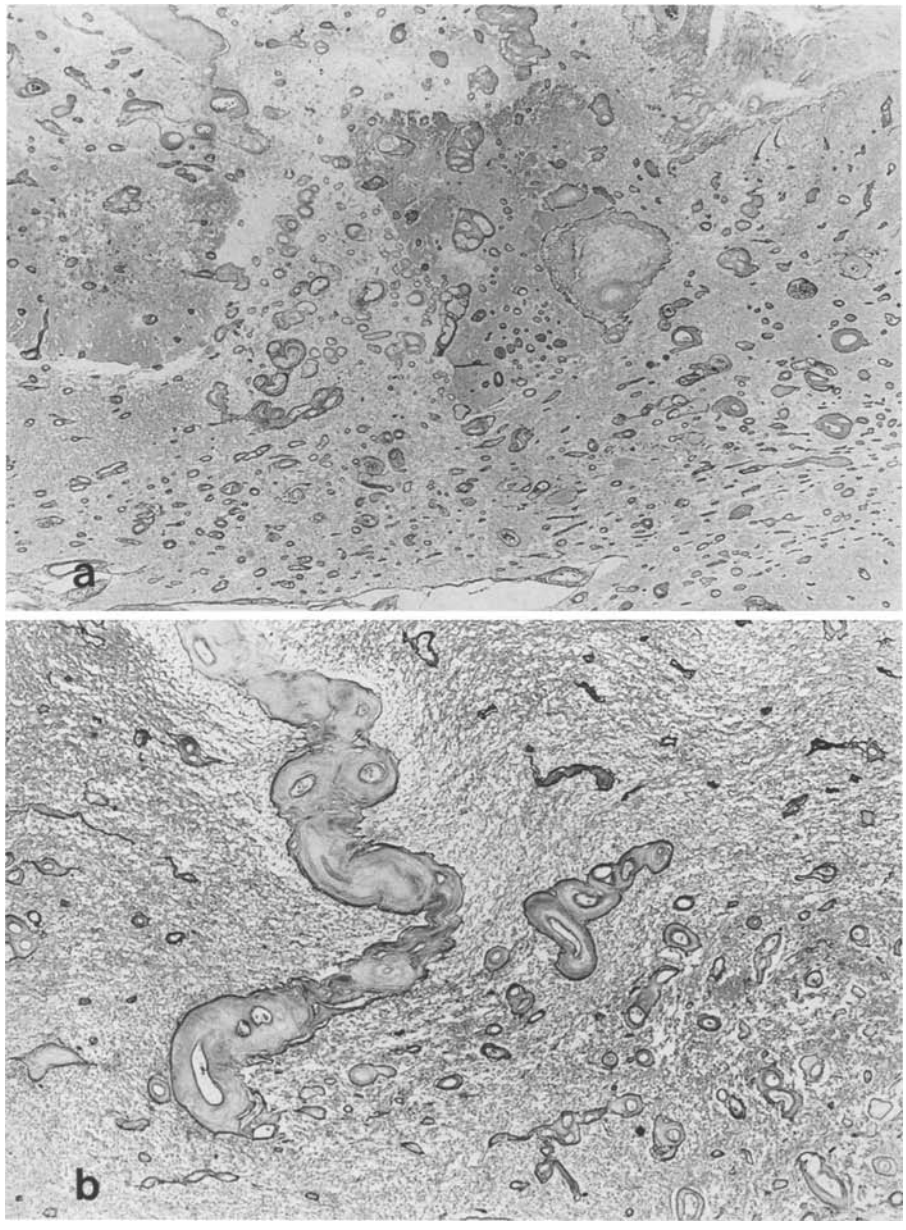

Fig. 2. a Numerous small abnormal vessels of the posterolateral portion of C8 and coagulation necrosis of the parenchyma in upper left part. Reticulin stain, ×40.
b Tortuous vessels with hyalinized wall of the left posterior funiculus of C7. Reticulin stain, ×75

extended to the right lower extremity in several weeks, and was followed by muscle weakness of both lower extremities and subsequently by recto-urinary disturbance. In addition, sensorimotor disturbances of the left upper extremity became apparent in November. The symptoms were gradually progressive and the patient began to notice frank gait disturbance by the end of the year. Neurological examination at the time of admission revealed sensory disturbance of all modalities below C4-level, muscle atrophy and weakness of the left upper extremity, decreased deep tendon reflexes of both the lower and the left upper extremities, bilateral extensor plantar responses and an atonic urinary bladder. X-ray films of the lumbar spine showed compression fracture of the L2 spine. Cerebrospinal fluid was normal. Myelography revealed an abnormally tortuous contour of the vessels on the ventral surface of the spinal cord from C4 to C6 and selective vertebral arteriography showed an abnormal vessel on the ventral surface of the upper cervical cord, presumably fed by the vertebral artery. His symptoms progressed into tetraparesis with dysphagia. He died of aspiration pneumonia at the age of 80. The total clinical course was eight years and one month.

Autopsy examination showed slight muscular atrophy of both the lower and left upper extremities without deformity, and severe aspiration pneumonia of the right lung. The brain (1290 g) showed slight ischaemic change in the cerebral and cerebellar cortical neurons.

Macroscopically, the major neuropathological changes were confined to the spinal cord. Dilatated and slightly tortuous vessels from C1 to Th12, most prominent over the ventral surface of the cervical cord were found and the radicular veins were also slightly dilatated (Fig. 1a). The cut surface of the cervical cord showed brown discoloration, predominantly in

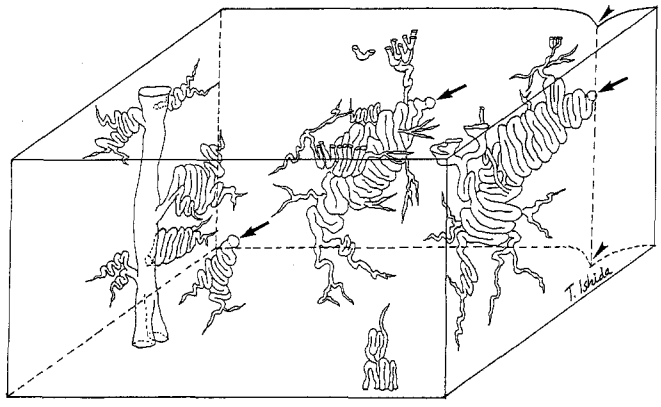

Fig. 3. Three-dimensional reconstruction of the intramedullary abnormal vessels of C7 discloses markedly tortuous elongation of original vasculature showing anomalous configuration and these vessels end in the extramedullary vessels (*arrows*). The posterior lateral sulcus is schematically indicated (*arrowheads*)

the left posterior portion and in the margin of the cord. The posterior spinal arteries were unremarkable in gross appearance.

Microscopically, several extramedullary vessels over the ventral and dorsal surfaces appeared abnormal, with irregular dilatation and thickening of the wall (Fig. 1b). There was an abnormal vessel with thickened intima and partially interrupted lamina elastica interna on the ventral surface of the spinomedullary transition approximately 0.5 cm below the medulla oblongata (Fig. 1c). These vessels were presumably histologically representative of the AVM itself. Numerous small and tortuous intramedullary abnormal vessels, predominantly in the left posterior horn and posterior funiculus were noted (Fig. 2a). These intramedullary vessels had no lamina elastica and the lumina were stenotic with thick hyalinized wall (Fig. 2b). In some, the lumen was narrowed to a pinpoint and the wall was focally laden with a small amount of haemosiderin, suggestive of old haemorrhage. Fibrin thrombus was found in a vessel of the deep white matter of the anterior funiculus of C8. The softening of the cord, which chiefly affected the left posterior horn and the white matter with atrophy of the anterolateral funiculus was noticed from C1 to Th12. The anterior horn was relatively preserved. Coagulation necrosis which Scholz (1949) described as "Plasmatische Infiltrationsnekrose" was observed in the left posterior funiculus of the C8 (Fig. 2a). The area of softening was more widespread than intramedullary vascular lesion.

A series of 5 μm-thick serial sections covering the seventh cervical segment were prepared from a paraffin-block. They were stained with elastica van Gieson stain, reticulin stain and haematoxylin-eosin stain alternately. Manual three-dimensional reconstruction of intramedullary abnormal vessels measuring larger than 20 μm in diameter (radial vein level) was carried out. The reconstruction disclosed that markedly tortuous elongation of the nascent vessels was present and gave rise to the anomalous appearance as seen in Fig. 2a, b (Fig. 3). There were no extraneous budding or neoformation of vessels. These abnormal vessels ended in the extramedullary thick walled vessels.

Discussion

Through the reconstruction of the small intramedullary abnormal vessels, it was confirmed that the abnormality was subsequent to elongation and tortuousity of precapillary vessels in a limited space in the spinal cord. The abnormal intramedullary vasculature is often considered as characteristic of spinal AVM along with softening with coagulation necrosis of the spinal parenchyma, occurring at the site of the most predominant intramedullary vascular lesion. The tortuous elongation of the vessels, however, appears to be merely secondary to chronic volume and pressure overload brought about in the venous system. Aminoff et al. (1974) described that marked capillary proliferation suggestive of long-standing hypoxia was found in the affected segments of the cord. But our study showed only elongation and tortuosity of the original vascular architecture. Hyalinized walls of the intramedullary abnormal vessels suggest excessive infiltration of plasma proteins similar to the hyalinized arterioles of the kidney, spleen and other organs in essential systemic hypertension (Biava et al. 1964).

Djindjian et al. (1978) reported that the steal phenomenon was the predominant pathogenesis of the signs and symptoms of spinal AVM. In addition to ischaemia due to steal, compression by mass may be significant in the development of neurological symptoms. It is considered that tortuous elongation of the intramedullary abnormal vessels indicated above would have played a role as a compression factor in our case.

The property of coagulation necrosis ("Plasmatische Infiltrationsnekrose", Scholz 1949), is similar to nonhaemorrhagic venous infarction due to thrombotic occlusion of the leptomeningeal venous vasculature, as described by Kim et al. (1984). No such finding was identified in the present case, however, in which only a minute thrombus in a vessel of the anterior funiculus of C8 was found.

AV fistulae have been considered essential to the pathogenesis of spinal AVM (Doppman 1971; Merland et al. 1980; Oldfield et al. 1983; Doppman et al. 1985; Heros et al. 1986). Benhaiem et al. (1983) described AV fistulae in the retromedullary AVMs within the dura mater by serial section of the surgical material. Benhaiem-Sigaux et al. (1985) reported retromedullary AV fistulae associated with Klippel-Trenaunay-Weber syndrome. In our case, the presence of a connection between the vertebral artery and the abnormal vessels was suggested angiographically but could not be confirmed histologically in the autopsy specimen. A vessel of the spinomedullary transition about 0.5 cm below the medulla oblongata (Fig. 1c), in which the lamina elastica partially interrupted, may correspond to and represent the AV shunt

suspected on angiography. The connection between this abnormal vessel and the vertebral artery, however, was not confirmed histologically, because we could not remove the dura and surrounding tissue containing the branches of the vertebral artery.

Merland et al. (1980) described that the shunt in their cases was very small and its precise location was difficult to determine even with selective arteriography and angiotomography. AV fistulae demonstrated by Benhaiem et al. (1983) also had a very small communication between an artery and a vein in the transdural location. Extensive histological examination of the spinal cord, with consideration of angiographic findings, including the dura mater is required in order to locate the connection between the feeder and the drainer in an autopsy case of spinal AVM.

Sarget (1925); Ritter (1927); Haberland (1950); Scholz and Manuelidis (1951); Kothe (1953); Schliack and Fölsch (1958) and Lange-Cosack and Peisker (1965) reported a case or cases in which the symptoms of AVM appeared after a trauma. In the eight cases thus reported, with an age distribution which ranges from 18 to 71 years, male preponderance (all males), and the site of AVM as located in the lower thoracic and lumbosacral region do not substantially differ from those without history of trauma. The most common age of the onset of spinal AVM with and without trauma is the fifth and sixth decades and male to female ratio is 4 to 1 (Aminoff and Logue 1974). The majority of AVM are sited in the thoracolumbosacral region (Yasargil 1971; Aminoff and Logue 1974). The time interval between the trauma and development of symptoms varied and ranged over a few months to several years. Our case had sustained two traffic accidents involving head and lumbar region, respectively. The former accident was twelve years prior and the latter was ten months prior to the onset of the symptoms. Benhaiem et al. (1983) proposed that an AV fistula of the dura of the spinal cord was an aquired lesion, however, the relationship to trauma was not discussed. However, Béraud (1972) considered that traumatic factors do not play a significant role in the pathogenesis of the disorder and the concept that AVM is congenital in aetiology appears to be generally accepted (Yasargil 1971; Béraud 1972; Jellinger 1978). Although the spinal AVM in our case could be traumatic in origin, we should mention that trauma may have played a part in the manifestation of symptoms.

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