Case report

Posterior spinal artery syndrome showing marked swelling of the spinal cord: A clinico-pathological study

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Objective: To describe a rare autopsy case of posterior spinal artery syndrome with marked swelling of the spinal cord, an unusually subacute onset and short clinical course.

Methods: Case report.

Findings: An 84-year-old Japanese woman presented with bilateral muscle weakness of the lower legs and sensory disturbance 1 week after head contusion. Neurological findings worsened gradually. She developed phrenic nerve paralysis and died of respiratory failure 6 weeks after the onset of neurological symptoms. On pathological examination, the spinal cord was markedly swollen in the cervical and upper thoracic segments. Microscopically, there was loss of myelin sheath in the bilateral posterior columns and neuronal loss of the posterior horns in all of the spinal segments. However, findings were unremarkable in the bilateral anterior columns and bilateral anterior horns in most of the spinal segments. Posterior spinal arteries had no stenosis, occlusion, or thrombosis. We considered that pathogenesis was infarction associated with head injury.

Conclusion: To our knowledge, this is the first report of a case of posterior spinal artery syndrome with a markedly swollen spinal cord and poor prognosis.

Keywords: Myelopathy, Posterior spinal artery syndrome, Paraparesis

Introduction

To date, there have been few reports describing posterior spinal artery syndrome. The definition of posterior spinal artery syndrome is infarction in the posterior spinal artery area, i.e. bilateral posterior columns, bilateral posterior horns, and posterior segments of bilateral lateral columns. Generally, the incidence of spinal cord infarction is lower than that of cerebral infarction because spinal arteries have many more anastomoses and much less atherosclerosis than cerebral arteries. Most individuals have 4–10 anterior radicular arteries, as well as 10–23 posterior radicular arteries.¹ The posterior spinal arteries are smaller in diameter and more evenly distributed than the anterior ones. Furthermore, patients with infarction of the spinal artery region range from young to old, whereas cerebral

artery infarction occurs much more often in the elderly. Therefore, infarction of the spinal cord, particularly in the distribution of the posterior spinal arteries, is extremely rare.

Generally, the onset of posterior spinal artery syndrome is variable, i.e. acute, subacute, or chronic. Whatever the events, posterior spinal artery syndrome usually has a favorable prognosis. However, this patient had a short clinical course and poor prognosis. To date, there have been few reports describing posterior spinal artery syndrome with pathological descriptions. Furthermore, etiology of posterior spinal artery syndrome has not been clarified. Here, we report a rare autopsy case of posterior spinal artery syndrome from clinical and pathological standpoints.

Case report

After a fall, an 84-year-old Japanese woman consulted a neurosurgeon in our hospital for a bruise on the back of

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the head and neck. Initially, she had had no neurological disturbances, but she gradually developed neck pain, muscle weakness, and sensory disturbance of the lower extremities. She consulted a family doctor and, because a cervical spinal cord injury was suspected, was admitted to the department of neurology in our

Figure 1 Spinal MRI showing marked swelling (indicated by yellow arrows) of the spinal cord in the cervical and upper thoracic segments. (A) T1-weighted sagittal image indicating marked swelling of the cervical and upper thoracic segments of the spinal cord with signal elongation. (B, C) T2-weighted sagittal image revealing that there is widespread marked swelling of the cervical and upper thoracic segments of the spinal cord with signal elongation. (The cervical and upper thoracic spinal region (B), the lower cervical and thoracic spinal region (C).) (D) Post-contrast-enhanced T1-weighted image demonstrating heterogeneous enhancement in the upper thoracic segments of the spinal cord. (E) A T2-weighted axial image indicating the cervical spinal cord is markedly swollen, and occupies the cerebrospinal fluid space.

hospital 5 days after the head contusion. Vital signs revealed no pathological findings. On neurological examination, her consciousness was clear. Cranial nerve examination was normal. However, she had severe muscle weakness in the upper and lower extremities. Deep tendon reflexes of the upper and lower extremities were extremely weak, and she also presented with rectourinary disturbance. Deep sensory disturbance was identified in the bilateral upper and lower extremities, but no superficial sensory deficit was seen. Spinal magnetic resonance imaging (MRI) showed marked swelling of the spinal cord on T1-weighted images with signal elongation and T2-weighted images with signal elongation in the cervical and upper thoracic segments (Fig. 1). Cerebrospinal fluid obtained by lumbar puncture contained no cells, elevated total protein (813 mg/100 ml), and 66 mg/100 ml glucose, while pressure was within the normal range. No atypical cells were found in the cerebrospinal fluid.

She was clinically diagnosed as having cervical cord injury based on neurological examination and spinal MRI findings. She was treated with high-dose steroid (methylprednisolone 1000 mg/day) therapy. However, neurological symptoms progressed, and she developed hypoventilation coma associated with paralysis of the respiratory muscles about 6 weeks after the onset of neurological symptoms. Death was attributed to respiratory failure. Autopsy of the brain and systemic organs was performed.

Pathological examination and findings

Macroscopic findings

The brain weighed 1200 g. Macroscopically, there was no evidence for cerebral or cerebellar herniation or subarachnoid hemorrhage. At the cut surface, the brain showed no hemorrhage or hematoma (Fig. 2A). No tumor was identified in the brainstem, cerebellum, or cerebrum. The cervical and upper thoracic segments of the spinal cord were markedly swollen and softened (Fig. 2B). Especially in the upper thoracic segments of the spinal cord, we could not discriminate the internal spinal structure, such as the boundary between the gray and white matter, which was indistinguishable. However, the other segments, including the lower thoracic, lumbar, and sacral segments of the spinal cord revealed no deformities (Fig. 2B). There was no hematoma around the spinal cord. No mass lesions were found in the central nervous system or other organs.

Microscopic findings

Microscopically, there was loss of myelin sheath in the bilateral posterior columns in all the spinal segments

Figure 2 Pathological findings. (A) Macroscopic findings of the brain. The frontal cut surface of the brain shows neither hemorrhage nor edema. (B) Macroscopic finding of the spinal cord. The cervical and upper thoracic segments of the spinal cord show marked swelling, whereas the other segments show unremarkable changes. The swollen area of the spinal cord is indicated by black arrows. (C) Cervical cord is markedly swollen with demyelination of the bilateral posterior region (Klüver-Barrera staining). (D) Lumbar cord has no swelling with demyelination of the bilateral posterior region (Klüver-Barrera staining). (E, F) Microscopic findings of the spinal cord (hematoxylin and eosin staining). (E) Neuronal cells are preserved in the anterior horn of the cervical cord. (F) Neuronal cells are decreased and pyknotic in the posterior horn of the cervical cord. Many reactive glial cells are identified. Bars: (A, B) 10 mm, (C, D) 5 mm, (E, F) 50 μ m.

(Figs. 2C–F). Axons were relatively preserved in the same areas. There was also neuronal loss in the bilateral posterior horns in all the spinal segments (Fig. 2F). In

the swollen area, i.e. the cervical and upper thoracic segments, there was mild loss of myelin sheath with minor lymphocyte infiltration in the posterior areas in the

bilateral lateral columns (Figs. 2C–D). Axons were relatively preserved in the same areas. Furthermore, there was widespread granulation tissue with capillary proliferation, marked gliosis, mild lymphocytic infiltration, and mild macrophage aggregation in the lower cervical and upper thoracic segments of the spinal cord, probably due to the contusion. Glial cells had mild nuclear atypia, but no pleomorphism. There were no significant changes in the bilateral anterior columns or bilateral anterior horns in any of the spinal segments (Fig. 2E) except for the narrow segments of the lower cervical and upper thoracic cord. Neither thrombi nor fibrocartilaginous emboli were found in the anterior spinal artery or posterior spinal arteries. There were no arteriovenous malformations, or mass lesions in either the spinal cord or the spine. No mass lesions were found in the thoracic cavity, i.e. aortic aneurysm, hematoma, or neoplasm. Furthermore, there was no sign of any malignancy in the central nervous system or other organs.

Discussion

Posterior spinal artery syndrome is rare. Williamson² first reported an autopsy case of posterior spinal artery syndrome in 1895. To our knowledge, only about 30 cases have been reported.^{1–24}

There are various causes of posterior spinal artery syndrome. For example, until the 1950s, meningovascular syphilis was a major cause of posterior spinal artery syndrome.^{3–5} Since then, infarction of the posterior spinal artery territories has also been a major cause of posterior spinal artery syndrome.^{1,6–13,25} Infarction of posterior spinal arteries may be caused by thromboembolization,^{14,15} arteriosclerosis,^{15,16,26} dissecting aortic aneurysm,^{17,18} or minor indirect trauma to the posterior spinal artery.^{15,19} Furthermore, iatrogenic phenol intrathecal injection therapy has been reported as a predisposing factor for posterior spinal artery syndrome.²⁰ In many cases, no occlusions were detected in the spinal blood vessels.

Recently, some cases have been diagnosed clinically by MRI or angiography. For example, Gutowski *et al.*⁸ reported an acute onset posterior spinal artery syndrome case with fair prognosis. That patient experienced an infarction in the territory of the unilateral posterior spinal artery. It was hypothesized that the discrete infarction was probably due to partial left vertebral artery dissection secondary to sneezing. They clinically diagnosed the case as posterior spinal artery syndrome by MRI. His neurological abnormalities recovered gradually. Fukuda and Kitani⁹ described an acute onset case of posterior spinal artery syndrome with a

fair prognosis. Their case experienced infarction in the upper cervical cord region during antelexion. They clinically diagnosed the case as posterior spinal artery syndrome by MRI and angiography. The neurological disturbance of the case recovered gradually. Kaneki *et al.*¹⁰ reported a subacute onset posterior spinal artery syndrome case with fair prognosis. They clinically diagnosed the case as posterior spinal artery syndrome by MRI. They suggested that posterior spinal artery syndrome shows variety in its clinical course because of its complicated anastomosis network. No pathological examinations were performed in the above three cases.

To date, case reports of posterior spinal artery syndrome with pathological examination are very rare.^{1–3,5,13,15,19–23} For instance, Takahashi *et al.*²³ reported an acute onset case with a long clinical course of more than 1 year. On pathological examination, the case had infarction in the region of the posterior spinal arteries in the bilateral upper cervical spinal cord. Hegedüs and Fekete¹ reported an acute onset posterior spinal artery syndrome case. The patient's neurological condition improved slowly, but she suddenly died of pulmonary thromboembolism. On pathological examination, there was widespread infarction in the region of the posterior spinal arteries in the bilateral lower lumbar and sacral spinal cord. Stone and Roback¹⁹ reported a case of myelomalacia, symptoms of which began while the patient was hanging by her arms from the limb of a tree. They hypothesized that her myelomalacia may have been precipitated by ischemia due to injury to the cervical spinal arteries or the arteries origin. We considered that the pathogenesis of our case was infarction associated with head injury.

Our case had subacute onset (about 10 days), a short clinical course (about 50 days), and poor prognosis, while posterior spinal artery syndrome generally has fair prognosis, i.e. spontaneous recovery. To date, there have been few reports of posterior spinal artery syndrome with a short clinical course and poor prognosis. Furthermore, there have been few reports of posterior spinal artery syndrome cases with markedly swollen spinal cord as in our case. Our case had no neurological symptoms before she experienced a severe head and neck contusion and developed neck pain and several neurological symptoms within five days after the contusion. Pathologically, there was loss of myelin sheath in the bilateral posterior columns and neuronal loss of the posterior horns in all the spinal segments, while there were unremarkable changes in the bilateral anterior columns and bilateral anterior horns in most

of the spinal segments. Martin *et al.*²⁷ described that a case with typical acute traumatic central cord syndrome after minor hyperextension injury to the neck. There were disk protrusions, but no vertebral fracture or displacement. Consequently, we hypothesized that pathogenesis of the spinal cord lesion in our case was infarction of the posterior spinal arteries caused by acute trauma of the spinal cord due to the head and neck contusion.

From clinical and radiological standpoints, we listed transverse myelitis, multiple sclerosis, intravascular lymphoma, diffuse gliomatosis, arteriovenous malformation, and vitamin B₁₂ deficiency as differential diagnoses. Some reasons that this case is unusual for transverse myelitis are as follows. Firstly, cerebrospinal fluid obtained by lumbar puncture did not reveal an increase in cells. Secondly, although this woman had been treated with high-dose steroid therapy, neurological symptoms progressed. Furthermore, our case presented with deep sensory disturbance in the bilateral upper and lower extremities, but no superficial sensory disturbance, which is unusual for transverse myelitis. Generally, in multiple sclerosis there are not only spinal lesions, but also random intracranial lesions. In this case, however, lesions were mainly distributed in the bilateral posterior columns and bilateral posterior horns in most segments of the spinal cord, and no intracranial lesions were found. Furthermore, histologically, perivascular lymphocyte cuffing, which is compatible with pathological findings of multiple sclerosis, was not present. This patient had no past history of autoimmune diseases. Furthermore, there was no invasion of lymphoma cells. There was widespread granulation tissue with capillary proliferation and multiple glial cells in the upper thoracic segments, but glial cells were non-neoplastic, probably due to the contusion. We could not find any arteriovenous malformation, thrombosis, or obstruction of spinal vessels. As for vitamin B₁₂ deficiency, this woman was well nourished and had no history of gastrectomy.

Conclusion

In conclusion, our assessment was posterior spinal artery syndrome caused by acute trauma of the spinal cord, characterized by a markedly swollen spinal cord and poor prognosis.

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