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【 CASE REPORT 】

Sudden-onset C8 Radiculopathy due to a Plexiform Schwannoma of the Cervical Nerve Root

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Abstract:
Plexiform schwannoma is a rare variant of schwannomas and usually presents with chronic symptoms. We herein report a case of cervical nerve plexiform schwannoma that presented with unusually sudden severe left radiculopathy of the eighth cervical nerve after physical exercise. Coronal short-tau inversion recovery (STIR) magnetic resonance imaging (MRI) revealed a multinodular tumor along the eighth cervical nerve. The tumor was partially resected. A pathological analysis revealed that the tumor was a schwannoma, and we diagnosed the case as a plexiform schwannoma. The unusual sudden-onset presentation in this case was considered to be caused by the unusual localization of the tumor involving the nerve root and mechanical stress due to physical exercise.

Key words: plexiform schwannoma, multinodular, peripheral nerve tumor, STIR, sudden-onset

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Introduction

Schwannomas are benign tumors originating from Schwann cells and are the most common nerve sheath tumors. Conventional schwannomas often develop in acoustic, spinal, or peripheral nerves, where they form an encapsulated, globular, single mass within a single fascicle. Spinal or peripheral nerve schwannomas can cause chronic sensory symptoms triggered by compression or percussion, but they seldom produce persistent pain or severe weakness (1, 2). Since total resection can often be achieved, the outcome is usually favorable, even in cases with a large deep-seated lesion affecting major peripheral nerves. Plexiform schwannomas are a rare variant of schwannomas that account for 4.3% of all schwannomas. They typically develop in the dermis or subcutaneous tissue but occasionally occur in deep-seated nerves. They are characterized by a plexiform or multinodular growth pattern involving multiple fascicles. Although plexiform schwannomas also show a chronic course that reflects their histopathologically benign nature, in contrast to conventional schwannomas, plexiform schwannomas in major nerves often cause motor deficits with poor functional prognosis due to the difficulty of their total resection (3, 4).

We herein report a case of a plexiform schwannoma of the eighth cervical (C8) nerve presenting with unusually sudden and severe sensorimotor palsy. This case and its pathomechanism are of interest because both the clinical course and localization of the tumor are atypical for plexiform schwannoma.

Case Report

A man in his 60s presented with the sudden onset of left hand palsy. He had been feeling a dull pain in the left upper back since playing golf two days earlier. While pressing his back against the corner of a pillar, he suddenly felt a painful sensation in his left arm. Dysesthesia on the ulnar side of the forearm and hand remained afterwards. The next morning, he noted severe weakness of the left hand. He visited an orthopedic clinic and underwent magnetic resonance imaging (MRI) of the cervical spine including T1- and T2-weighted sagittal and T2-weighted axial imaging (Figure A), but no causative lesion was diagnosed. The back pain resolved within several weeks; however, his hand weakness

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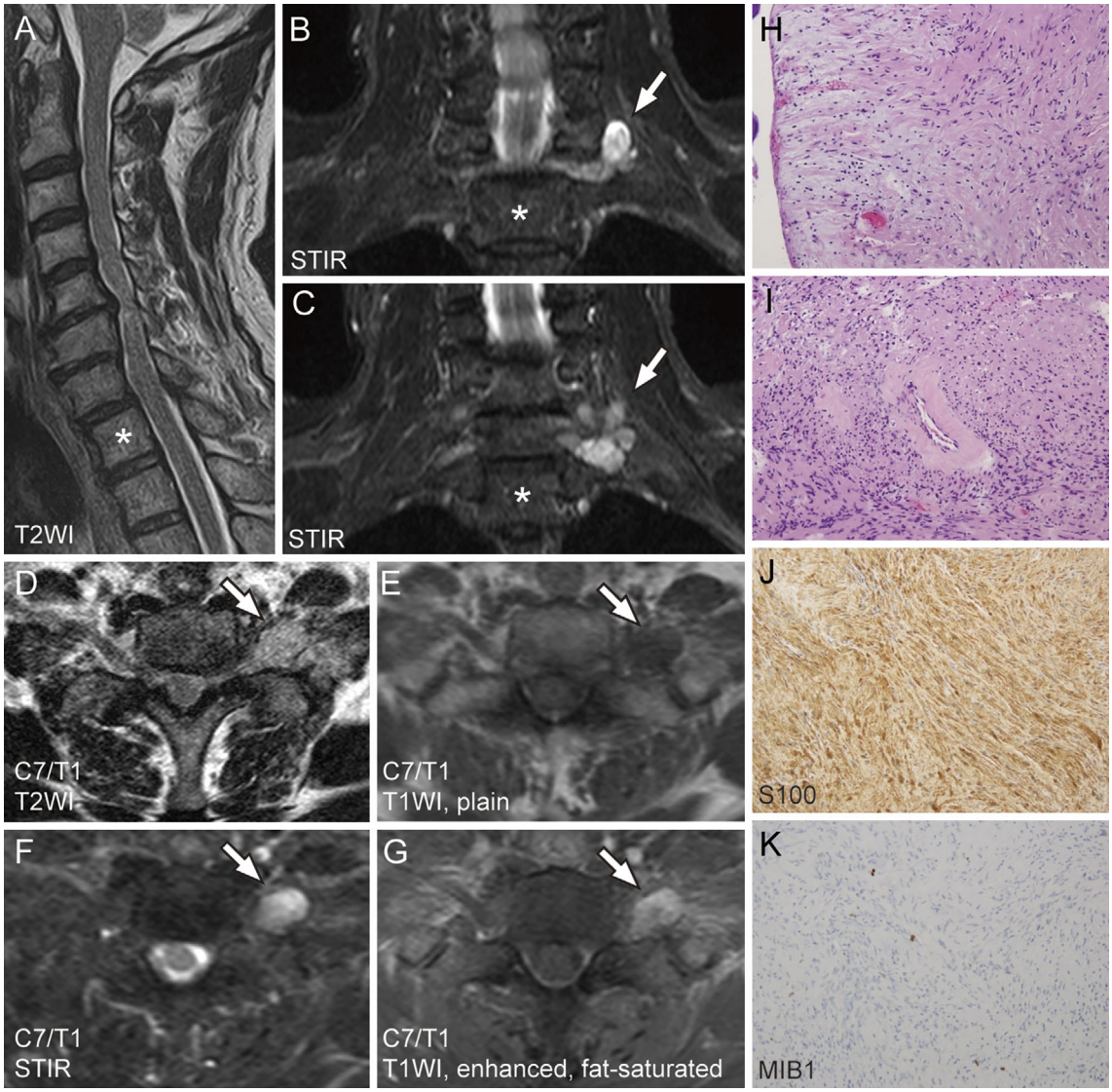

Figure. **(A) Magnetic resonance imaging (MRI) of the cervical spine performed two days after the symptom onset. Sagittal T2-weighted imaging (T2WI) shows spinal cord compression at the C5/6 level, but the lesion causing the left C8 and T1 level palsy remains undiagnosed. The asterisk indicates the T1 vertebral body in A to C. (B, C) Coronal short-tau inversion recovery (STIR) imaging reveals multinodular lesions (arrows) along the enlarged left C8 nerve root. (D) Axial T2WI. The arrow indicates thickening of the left C8 nerve root. (E) Axial T1-weighted imaging (T1WI). The arrow indicates thickening of the left C8 nerve root. (F) STIR imaging clearly delineates the cervical nerve lesion. (G) Axial fat-saturated T1WI with gadolinium enhancement. The cervical nerve lesion shows heterogeneous enhancement. (H) The lesion protruding upward from the cervical nerve (B, arrow) is resected and histologically evaluated. An admixture of a more cellular area comprising spindle-shaped cells (Antoni A pattern, right side) and a hypocellular, looser area (Antoni B pattern, left side) is observed [Hematoxylin and Eosin (H&E) staining, ×200]. The Antoni A pattern is dominant. (I) Hyalinized vessels and palisading of the nuclei are observed (H&E staining, ×200). (J) Diffuse immunoactivity against S100 protein is observed (×200). (K) MIB1 is scarcely immunopositive, and the labeling index is 1-2% (×200).**

and dysesthesia remained unimproved, and he developed wasting of the left forearm and hand. He visited our hospital five weeks after the onset. At that time, he mentioned that he had experienced subtle, intermittent dysesthesia on the ulnar side of the left hand for the past two years. This facet of the history was initially disregarded until elicited repeatedly.

Neurological examination revealed severe weakness and moderate wasting of the left flexor carpi ulnaris, flexor digitorum profundus and all intrinsic hand muscles, graded 1/5 on the Medical Research Council (MRC) scale. The weakness was less severe in the left flexor carpi radialis and extensor digitorum (ED) (2/5 on the MRC scale) and mild in the left extensor carpi radialis (4/5 on the MRC scale). The

deltoid, biceps brachii, brachioradialis and triceps brachii muscles were normal bilaterally. The dysesthesia was localized on the ulnar side of the left hand and forearm. Touch sensation was decreased in the area. Tendon reflexes were diminished in the upper limbs and exaggerated in the lower limbs bilaterally. Plantar responses were flexor bilaterally. These neurological findings suggested left C8 and first thoracic (T1) nerve lesions along with myelopathy.

A nerve conduction study of the left arm revealed the absence of median and ulnar compound muscle action potentials recorded at the abductor pollicis brevis and abductor digiti minimi, respectively. Sensory nerve action potentials (SNAPs) of the left ulnar nerve were markedly reduced in amplitude, while those of the median nerve recorded at the second digit were normal. Electromyography showed active denervation exclusively in the paralyzed muscles. These electrophysiological findings suggested postganglionic lesions affecting the left C8 and T1. The cell count in the cerebrospinal fluid (CSF) was slightly elevated (9/μL, mononuclear cell predominance), and protein level was mildly elevated (88.3 mg/dL). CSF glucose level was normal (CSF/plasma ratio: 0.58), and cytology also yielded negative results. Cervical MRI using a short-tau inversion recovery (STIR) sequence revealed multinodular hyperintense lesions along the left C8 nerve that were heterogeneously enhanced by gadolinium (Figure B-G). The previous T2-weighted transverse images, on further review, indicated the same lesion, and the size of the lesions had not changed over the past five weeks. The MRI findings combined with neurological and electrophysiological findings suggested a C8 nerve schwannoma expanding to the lower trunk. The lesion protruding upward was partially resected, preserving the cervical nerve to minimize further motor and sensory deficits, and C4-7 laminoplasty was performed for coexisting spinal canal stenosis.

Histological examination revealed an admixture of a dominant, more cellular area composed of spindle-shaped cells (Antoni A pattern) and a hypocellular, looser area (Antoni B pattern) (Figure H), with vascular hyalinization (Figure I). Immunohistochemistry demonstrated diffuse immunoreactivity against S100 protein and a low MIB1 labeling index (1-2%) (Figure J and K). These findings were consistent with schwannoma. His motor and sensory deficits remained unimproved postoperatively.

Discussion

We reported a rare case of plexiform schwannoma arising in the left cervical nerve roots and the brachial plexus and presenting with sudden-onset sensorimotor hand palsy.

Although most schwannomas can be resected without any neurological sequelae, post-resection deterioration in the neurological function occurs in some cases, and it is sometimes only possible to perform subtotal resection in order to preserve the parent nerve function. Such cases of tumors involving multiple fascicles of the parent nerve are a subtype of schwannomas called plexiform schwannomas (3, 4). Preoperative MRI findings are important for distinguishing between conventional and plexiform schwannomas. In conventional schwannomas, the lesions are well-defined, have discrete margins and are globular in shape, with the parent nerve having a normal appearance. In contrast, plexiform schwannomas often feature individual enlarged fascicles coursing through and beyond the primary lesion and have a more tubular or fusiform appearance. Alternatively, plexiform schwannomas can appear as multiple nodules of varying size that are strung along a single nerve or fascicle. While conventional schwannomas show marked enhancement after gadolinium administration, plexiform schwannomas show somewhat less predictable enhancement. In some cases, the globular portion of the lesion is enhanced, but the more plexiform portion is not (4). Histologically, conventional schwannomas consist of a random admixture of two classical architectural patterns termed Antoni A (dense tissue with spindle-shaped cells) and Antoni B (loose tissue with stellate cells). Plexiform schwannomas show a histological spectrum across conventional, cellular and mixed schwannoma and are typically dominated by dense Antoni A tissue (1). The involvement of multiple fascicles is an important histological characteristic of plexiform schwannomas, but the plexiform nature of the lesions cannot always be histologically confirmed, as only a limited biopsy specimen is available in many cases. Therefore, preoperative MRI findings and intraoperative macroscopic findings of the plexiform or multinodular nature provide critical information for pathologists (4). Schwannomatosis and neurofibromatosis type 2 are each diagnosed in 5% of cases with plexiform schwannomas (3).

In the present case, the multinodular growth pattern with an enlarged parent nerve, which was clearly delineated by coronal STIR imaging, suggested the possibility of a plexiform schwannoma. Histological examination revealed typical findings of a schwannoma with an Antoni A dominant pattern. Although we could not confirm multifascicular involvement in the limited biopsy sample, the characteristic gross MRI findings, including the multinodular growth pattern and the long lesion expanding from the C8 nerve root to the lower trunk, strongly suggested a plexiform schwannoma. Schwannomatosis and neurofibromatosis type 2 were ruled out based on the absence of a family history, multiple tumors or skin abnormality.

In a large case series, most plexiform schwannomas were found to be located superficially, and only 3% affected major nerves (3). Only five cases of plexiform schwannomas affecting the spinal nerve roots or plexus have been reported, including two cases involving the cervical roots (4, 5), one involving the cervical and thoracic roots (6), one involving the cervical roots to the brachial plexus (7) and one involving the brachial plexus (3). Furthermore, upper limb weakness has been reported in three cases (4, 5, 7), and respiratory failure due to compression of the lung and airway has been reported in one case (6). Each

of these cases exhibited a chronic course. A possible mechanism underlying the unusual sudden-onset occurring in our case is proposed below.

The clinical presentations of schwannomas are usually chronic, and sudden neurological deficits are rare. Spontaneous intratumoral hemorrhage reportedly caused facial nerve palsy in a vestibular schwannoma (8), quadriparesis in a craniovertebral junction schwannoma (9) and median nerve palsy in a carpal tunnel schwannoma (10). In our case, intratumoral hemorrhage was not detected by MRI. Instead, physical exercise and mechanical compression to the patient's back seemed to have caused the acute neurological deterioration. Such mechanical stress may have damaged the spinal nerve root, as the involved nerve root was thickened and fixed to the intervertebral foramen, making it vulnerable to mechanical stress.

In contrast to conventional schwannomas, complete resection of the tumor without causing a neurological deficit is difficult in cases of plexiform schwannomas. A small fascicle associated with conventional schwannomas can be sacrificed without consequences, but in plexiform schwannomas, the tumor has complex growth associated with multiple fascicles, and their complete resection implies the sacrifice of the involved fascicles (4). Only a biopsy or subtotal resection preserving the parent nerves can be performed in most cases. Few reports on the treatment outcomes of plexiform schwannomas affecting the nerve roots have been published (5, 7). In one case, all preoperative symptoms were completely resolved by extensive resection of the intradural and extradural parts of the tumor (5). In another case, only a biopsy was performed because of the wide spread of the tumor and because intraoperative functional mapping was not possible (7). In our case, only a biopsy was performed because of the difficulty of resecting the tumor without sacrificing the parent nerves and because, even if the tumor could be resected, the functional prognosis would be poor, as axonal damage had already occurred.

We encountered a case of plexiform schwannoma arising in the cervical nerve. Plexiform schwannoma in spinal nerves may cause sensorimotor deficit, and mechanical stress can elicit acute deterioration of the radiculopathy. Spinal plexiform schwannoma is a rare but important etiology of acute-onset radiculopathy.

The authors state that they have no Conflict of Interest (COI).

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