Superficial siderosis of the central nervous system with epilepsy originating from traumatic cervical injury: illustrative case

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BACKGROUND Superficial siderosis of the central nervous system (SSCNS) is a rare condition that results from hemosiderin deposition in the brain, brainstem, cerebellum, and spinal cord as a result of chronic, repeated, and recurrent subarachnoid hemorrhage. SSCNS that originates in the spinal cord is rarely reported, and epilepsy as a manifestation of such a case has not been reported before.

OBSERVATIONS The authors reported a rare case of SSCNS with epilepsy originating from traumatic cervical injury and presented a literature review of all reported SSCNS cases that originated in the spine. The patient was a 29-year-old man with a 16-year history of progressive headache accompanied by seizures, ataxia, and sensorineural hearing loss. He had experienced a traumatic cervical injury at age 7. Magnetic resonance imaging revealed a characteristic hypointense rim around the pons and cervical spinal cord on susceptibility-weighted imaging scans. Cerebrospinal fluid examination during a headache episode confirmed subarachnoid hemorrhage and increased intracranial pressure. Surgical exploration revealed a C6 dural defect with bone spurs inserted into the dura mater. After the patient underwent dura mater repair and shunt implantation, his symptoms disappeared completely except for hearing loss.

LESSONS This rare case indicated that symptomatic epilepsy followed by SSCNS can be eliminated by complete repair of the cervical dura mater.

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KEYWORDS superficial siderosis; epilepsy; trauma; spine

Superficial siderosis of the central nervous system (SSCNS) is an uncommon and unrecognized disorder characterized by hemosiderin deposition on the surface of the brain, brainstem, cerebellum, and spinal cord as the result of chronic or intermittent bleeding into the subarachnoid space, which causes irreversible damage to the CNS and results in a series of neurological manifestations. The typical triad of SSCNS symptoms includes progressive cerebellar ataxia, central motor disability, and sensorineural hearing loss. Other symptoms, such as chronic increased intracranial pressure (ICP) and hydrocephalus, develop in approximately one-third of patients because of obstruction of the ventricular foramina and/or malabsorption of cerebrospinal fluid (CSF). Epilepsy is a rare manifestation of SSCNS as discussed in the literature, although patients with SSCNS may have a prior history of head trauma or surgical procedures. Most reported cases of SSCNS originate from traumatic brain injury and intracranial hemorrhage or surgery; relatively few cases originate from spinal injury, and in this latter group, epilepsy has not been reported. Medical or surgical treatments for SSCNS are often ineffective, and most reported cases progress slowly and inexorably. Although treatment with deferoxamine and cochlear implants have been tried, successful therapy still depends on determining the etiology of chronic bleeding and precise treatment. We report an interesting case of SSCNS originating from a traumatic cervical injury. The patient experienced intractable epilepsy and increased ICP in addition to the typical triad. His condition was treated successfully with repair of the dura mater.

Illustrative Case

A 29-year-old male patient presented with a 16-year history of progressive headache accompanied by intractable seizures and sensorineural hearing loss. The headache consisted of sudden-onset frontal or occipital radiating pain that was often accompanied by photophobia, diplopia, nausea, and vomiting. The pain was aggravated when he lay flat, and it gradually increased in severity and frequency. In the previous year, every headache was accompanied by generalized
tonic-clonic seizures, which fully subsided several minutes later. Although various antiepileptic drugs were used, the seizures were not controlled effectively. The patient also developed progressive deafness and ataxia within the previous 5 years, and the hearing loss was obvious on the left side. When he was referred to our hospital, epilepsy with headache onset was occurring approximately twice a month; furthermore, the patient was unable to walk independently, and the hearing loss in the left ear was almost complete.

When he was 7 years old, the patient had experienced a fall that resulted in a short period of disturbance in consciousness and neck pain. Neurological examination upon admission to the hospital revealed a deterioration of memory, decreased visual acuity without papilledema, nystagmus, hearing loss, positive Rinne test result, and ataxic gait. Other cranial nerve and sensory examinations produced normal results. On brain magnetic resonance imaging (MRI), axial T2-weighted images (Fig. 1A) and susceptibility-weighted imaging scans (Fig. 1B) showed a characteristic rim of hypointensity along the dorsolateral pons, which indicated the deposition of hemosiderin. CSF examination at headache onset indicated a high ICP (29 cm H₂O) and blood (>1,000 red blood cells per mm³), suggesting subarachnoid hemorrhage. Except for a slight increase in protein concentration, no specific positive results were shown in biochemical or immunoelectrophoresis examinations of CSF. No abnormalities were present on routine electroencephalography except for a wide slow wave throughout the brain. To determine the reason for the subarachnoid hemorrhage, digital subtraction angiography of the brain and spinal cord was performed; however, neither aneurysms nor arteriovenous malformations were found. Based on the prior history of falling, further MRI of the cervical spine was performed, which revealed a hypointense lesion in the subdural and epidural space of C6–7 on sagittal T2-weighted images (Fig. 1C). This finding indicated the possibility of a previous hemorrhage. Cervical computed tomography (CT) examination revealed hyperostoeogeny of the left C7 lamina (Fig. 1D) with a bone spur protruding into the spinal canal (Fig. 1E), which indicated a previous fracture. Because the osteophyte was believed to be the cause of recurrent subarachnoid hemorrhage, surgical exploration was indicated.

During the operation, we found a wide deposition of hemosiderin around the spinal cord, and a protruding osteophyte (Fig. 2) pierced the dura and the subdural space of the spinal cord, leading to an apparent dural defect. The osteophyte was removed, and the subcutaneous fascia was sutured over the dural defect in a watertight fashion. The patient recovered well after the operation; however, he experienced...
| Case No. | Author & Year | Age (yrs), Gender | Etiology | Major Symptoms | Time (yrs) | MRI Findings | CSF Findings | Location of Dural Defect | Closure Method | Outcome (FU) | Postop MRI Findings |
|----------|---------------|-------------------|----------|----------------|------------|--------------|--------------|-------------------------|----------------|--------------|---------------------|
| 1        | Kumar et al., 2005 | 42, M | Head injury at 10 yo | Gait ataxia, deafness, anosmia, incontinence | 8 | Epidural fluid collection from C4 to T9 | Xanthochromia, ICP: NM, RBCs: 1,133 | T2–3 | Muscle graft | No change (6 mos) | Reduction of fluid collection |
| 2        | 51, F | Head injury at 16 yo | Gait ataxia, deafness, incontinence | 4 | Epidural fluid collection from T1 to T3 | NM | T2 | — | — | Reduction of fluid collection |
| 3        | 52, M | Lt brachial plexus & spinal injury at 10 yo | Gait ataxia, deafness, tinnitus, incontinence | 7 | Epidural fluid collection from C3 to L5 | NM | T11 | — | — | — |
| 4        | Kumar et al., 2006 | 42, M | Lt brachial plexus & spinal injury at 20 yo | Gait ataxia, deafness | — | Epidural fluid collection from T1 to T5, C7–T1 pseudomeningocele | Xanthochromia, RBCs: 0, ICP: NM | — | — | No change (2 yrs) |
| 5        | Holle et al., 2008 | 59, M | Thoracic disc herniation | Gait ataxia, limb incoordination, slurred speech, deafness, anosmia | 3 | Epidural fluid collection from C5 to T6, disc herniation | Xanthochromia, RBCs: NM, ICP: 50 cm H₂O | T5–6 | Glue-coated collagen sponge | Improvement of headache, deterioration of cerebellar syndrome |
| 6        | Shih et al., 2009 | 70, M | — | Gait ataxia, deafness, tinnitus, cognitive decline | 2 | Epidural fluid collection from T2 to T8 | Xanthochromia, RBCs: 11, ICP: NM | T4–5 | Dural patch, dural sealant | No change (15 mos) |
| 7        | Kumar et al., 2009 | 64, M | C4–7 laminectomy | Gait ataxia, deafness | 10 | Epidural fluid collection from C3 to T11 | Xanthochromia, RBCs: 464, ICP: 4 cm H₂O | T7–8 | Free fat graft, sealant | Improvement of gait (6 mos) | Resolution of fluid collection |
| 8        | Ikeda et al., 2010 | 71, F | — | Gait ataxia, deafness | 7 | Epidural fluid collection from C7 to T12 | Xanthochromia, RBCs: >30,000, ICP: NM | T2–3 | — | No change (1 yr) |
| 9        | Kumar et al., 2010 | 54, M | Motor vehicle accident | Gait ataxia, deafness, slurred speech | 5 | Epidural fluid collection from C2 to T7 | Xanthochromia, RBCs: 1,243, ICP: 175 cm H₂O | T3 | Suture | Improvement of neck pain (4 mos) | Resolution of fluid collection |
| 10       | Cheng et al., 2011 | 53, M | Arachnoid cyst | Gait ataxia, deafness, dizziness | 2 | Epidural fluid collection from C7 to T4 | Xanthochromia, RBCs: 661, ICP: 115 cm H₂O | — | Glue | Improvement of gait (6 mos) | Resolution of fluid collection |
| 11       | Boncoraglio et al., 2012 | 69, M | Surgery for L4–5 disc herniation | Cerebellar ataxia | 4 | Epidural fluid collection from C2 to T9, T6–7 cord herniation | Xanthochromia, RBCs: NM, ICP: NM | T6–7 | Patch, fibrin glue | No change (6 mos) | Resolution of fluid collection |
| Case No. | Author & Year | Age (yrs), Gender | Etiology | Major Symptoms | Time (yrs) | MRI Findings | CSF Findings | Location of Dural Defect | Closure Method | Outcome (FU) | Postop MRI Findings |
|----------|---------------|-------------------|----------|----------------|------------|--------------|--------------|-------------------------|----------------|--------------|---------------------|
| 12       | Egawa et al., 2013 | 67, M | — | Headache, gait ataxia, deafness, dysarthria | 30 | Epidural fluid collection from C2 to T8 | Xanthochromia, RBCs: 1,000, ICP: 10 cm H₂O | T2–3 | Free muscle graft, fibrin glue patch | Improvement of headache, deterioration of neurological symptoms | Resolution of fluid collection |
| 13       | — | 54, M | — | Gait ataxia, tinnitus, slurred speech, diplopia | 4 | Epidural fluid collection from C7 to T8 | Xanthochromia, RBCs: 1,000, ICP: 13 cm H₂O | T1–2 | Suture, muscle graft | No change (18 mos) | Resolution of fluid collection |
| 14       | Yokosuka et al., 2014 | 53, M | Cervical laminectomy & removal of cervical schwannoma | Schizophrenia | 26 | Pseudomeningocele | Xanthochromia, RBCs: 768–1,034, ICP: normal | — | Autologous fat | No change (12 mos) | Resolution of pseudomeningocele |
| 15       | Schievink et al., 2016 | 33, M | — | Headache, nausea, emesis, tinnitus, low-back pain | 2 | Extensive ventral thoracolumbar extradural CSF collection & hematoma within lumbar ventral CSF collection | — | T9–10 | — | Improvement of all symptoms (12 mos) | Resolution of fluid collection |
| 16       | O’Hare et al., 2016 | 62, F | — | Headache, blurred vision, aural fullness, neck pain | 2 | Intrathecal hemorrhage & extensive spinal extradural CSF collection | — | — | Patch | Improvement of all symptoms (8 mos) | Resolution of fluid collection |
| 17       | — | 61, M | Extensive dural ectasia | Urinary retention, deafness, tinnitus | — | Extensive dural ectasia | — | T5–11 | — | — | Resolution of pseudomeningocele |
| 18       | Ryu SM et al., 2016 | 55, M | — | Gait ataxia, excretion disorder, tinnitus | 2 | — | Xanthochromia, RBCs: 15,260, ICP: NM | C1–2 | — | — | — |
| 19       | Madkouri & Grelat, 2017 | 58, M | Dural arteriovenous fistula | Cerebellar ataxia, pyramidal signs, dysarthria, deafness, cognitive impairment | — | — | — | C3–4, C5–6, C6–7 | Suture | Improvement of all symptoms (1 mo) | Resolution of fluid collection |
| 20       | Sakoda et al., 2017 | 64, M | Head injury | Headache, dizziness, deafness | 2 | Dural defect at T2–3 level on anterior side of spinal canal | Xanthochromia, RBCs: 4,144, ICP: 1 cm H₂O | T3 | Autologous fascia of neck muscle | Improvement of all symptoms (7 mos) | Resolution of fluid collection |
| Case No. | Author & Year | Age (yrs), Gender | Etiology | Major Symptoms | Time (yrs) | MRI Findings | CSF Findings | Location of Dural Defect | Closure Method | Outcome (FU) | Postop MRI Findings |
|----------|---------------|------------------|----------|----------------|------------|--------------|--------------|------------------------|----------------|--------------|---------------------|
| 21       | Takai et al., 2017<sup>20</sup> | 58, M | —       | Gait ataxia, dysarthria, deafness | 5          | Dural defect at several spinal levels from C4 to T7 | Xanthochromia, RBCs: 2,800–3,300, ICP: 4 cm H₂O | T1 | — | — |
| 22       | Hiraka et al., 2018<sup>20</sup> | 58, M | —       | Gait ataxia, deafness | — | Epidural fluid collection from C3 to T10 | Colorless, RBCs: NM, ICP: 130 cm H₂O | T1 | — | — |
| 23       | Bower et al., 2018<sup>31</sup> | 67, F | Marfan syndrome | Gait ataxia, deafness, urinary incontinence | 10 | Thoracic & lumbar spine dural ectasia | — | — | — |
| 24       | Hosokawa et al., 2018<sup>12</sup> | 62, M | —       | Gait ataxia, deafness, spasticity | 8 | Epidural fluid collection from T1 to T4 | Xanthochromia, RBCs: NM, ICP:NM | — | — | — |
| 25       | 60, M | —       | Gait ataxia, deafness | 3 | Epidural fluid collection from C1 to T4 | Xanthochromia, RBCs: NM, ICP:NM | — | — | — |
| 26       | 49, M | —       | Gait ataxia, deafness | 12 | Epidural fluid collection from T1 to T4 | Xanthochromia, RBCs: NM, ICP:NM | — | — | — |
| 27       | 68, F | —       | Gait ataxia, deafness | 2 | Epidural fluid collection from T1 to T4 | Colorless, RBCs: NM, ICP:NM | — | — | — |
| 28       | 74, F | —       | Gait ataxia, deafness | 13 | Epidural fluid collection from T1 to T4 | Colorless, RBCs: NM, ICP:NM | T7–8 | Suture | Improvement of headache, stability of other symptoms (17 mos) | Resolution of fluid collection |
| 29       | Arishima et al., 2018<sup>13</sup> | 50, M | Surgery for subdural hematoma | Gait ataxia, motor disturbance of bilat upper limbs | 10 | Epidural fluid collection from C2 to T12 | Xanthochromia, RBCs:NM, ICP:20 cm H₂O | C7 | Suture | Improvement of all symptoms (17 mos) | Resolution of fluid collection |
| 30       | 59, M | Surgery for subdural hematoma | Motor disturbance of rt upper & lower limbs | 0.25 | Epidural fluid collection from C2 to T2 | Xanthochromia, RBCs:NM, ICP:0 cm H₂O | T1–2, T3–4 | Synthetic dura material | Improvement of all symptoms (6 mos) | Resolution of fluid collection |
| 31       | Camlar et al., 2018<sup>34</sup> | 58, F | Thoracic spinal surgeries | Gait ataxia, deafness, dizziness | 0.75 | Dural defect at T1–2 level | Xanthochromia, RBCs:NM, ICP:11 cm H₂O | T8–9 | Suture | Improvement of all symptoms (24 mos) | Resolution of fluid collection |

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| Case No. | Author & Year | Age (yrs), Gender | Etiology | Major Symptoms | Time (yrs) | MRI Findings | CSF Findings | Location of Dural Defect | Closure Method | Outcome (FU) | Postop MRI Findings |
|---------|---------------|-------------------|----------|----------------|------------|--------------|--------------|----------------------|----------------|----------------|---------------------|
| 32      | Brembilla et al., 2018 | 48, M | — | Gait ataxia, deafness | 3 | Osteophyte at T8–9 level | Xanthochromia, RBCs: 6,000, ICP: 6 cm H₂O | — | — | Stability of deafness (24 mos) |
| 33      | Nasri et al., 2018 | 48, M | Head injury at 2 yo | Motor disturbance of bilateral upper limbs, urinary dysfunction | 10 | Dural pseudomeningoceles of Lt C3 to C7 nerve roots | — | T1–2 | Suture | No change | Resolution of fluid collection |
| 34      | Machino et al., 2019 | 71, M | — | Gait ataxia, giddy feeling, dizziness | 5 | Epidural fluid collection from C7 to T5 | — | T1–2 | Fat & fascia lata | No change (12 mos) | Clivus reconstruction |
| 35      | Vellutini et al., 2019 | 35, M | Head injury at 15 yo | Deafness | — | Arachnoidocele | Xanthochromia, RBCs: NM, ICP: NM | T3 | Suture, fibrin sealant | Death (8 mos) | — |
| 36      | Nathoo et al., 2020 | 74, M | Head injury | Gait ataxia, deafness, dizziness, cognitive impairment, urinary retention | 2 | Epidural fluid collection from T2 to T5 | — | T1–2, T5–6 | Suture | Improvement of headache, stability of other symptoms (17 mos) | Resolution of fluid collection |
| 37      | Katoh et al., 2020 | 74, F | — | Gait ataxia, deafness | 7 | Epidural fluid collection from C7 to T10 | — | C6–7 | Fibrin glue | Improvement of headache (36 mos) | Resolution of fluid collection |
| 38      | Wia˛cek et al., 2020 | 63, M | Rt brachial plexus & spinal injury at 35 yo | Gait ataxia, deafness, slurred speech, headache | 7 | Epidural fluid collection from C3 to T12 | Xanthochromia, RBCs: NM, ICP: 5 cm H₂O | T9–10 | Dura substitute, fibrin glue, autologous fat graft, absorbable gelatin sponge | Improvement of all symptoms (16 mos) | Resolution of fluid collection |
| 39      | Cornips et al., 2020 | 56, M | Transdural thoracic disc herniation | Headache, cognitive dysfunction | 0.1 | — | — | T7–8 | Dura substitute, fibrin glue, autologous fat graft, absorbable gelatin sponge | Improvement of all symptoms (4 mos) | Resolution of fluid collection |
| 40      | 33, M | Transdural thoracic disc herniation | Headache, dizziness | Epidural fluid collection from C2 to T12 | 2 | — | — | C7 | Suture | Improvement of all symptoms | Resolution of fluid collection |
severe headache and vomiting after the drainage tube was pulled out. Lumbar puncture was performed, and test results indicated an extremely high ICP (>33 cm H₂O) on the 15th day after the operation. However, there were no red blood cells in the CSF, and the protein concentration was normal. Although the ventricle was not obviously enlarged, increased ICP was diagnosed, and a lumbar-peritoneal shunt was placed to drain CSF and decrease ICP. The patient recovered uneventfully, and his headache disappeared immediately. His progress was followed up regularly. At the 18-month follow-up visit, the patient was free from headache and seizures, and his ataxia had improved greatly; however, his deafness had not improved.

**Discussion**

**Observations**

SSCNS is a rare neurodegenerative disease that results from toxic accumulation of hemosiderin on the surface of the brain and spinal cord. Although the number of reported cases is increasing, the natural history and clinical evolution of SSCNS are poorly understood. Further identification and resolution of the bleeding source do not elicit prompt clinical recovery or radiological reversal of SSCNS in most cases, leading to a major challenge in further diagnosis and treatment. Most clinical signs and symptoms of superficial siderosis are believed to be related to the anatomical distribution of hemosiderin deposits within the neural system.10,11 Hemosiderin is apt to deposit in tissues that are exposed to abundant CSF, such as the vermis, superficial sulci and gyri, basal frontal lobe, temporal lobe, brainstem, and spinal cord as well as cranial nerves I, II, and VII, which leads to the typical triad of progressive gait ataxia, central motor disability, and sensorineural hearing loss. Other manifestations have been reported, such as diplopia, hyposmia, amnesia, headache, and seizures.12,13 Because most of the damage to the CNS is irreversible, it is vital to determine the etiology and intervene as early as possible. Although extensive diagnostic examinations are used to determine the causative pathologies of bleeding conditions, the etiology of more than 30% of subarachnoid hemorrhage cases remains unknown.13

Recently, attention has been drawn to the association between SSCNS and dural defects in the spinal canal. We searched all related English-language literature in PubMed, GeenMedical, and other databases and identified 41 cases of SSCNS8,12,14–42 associated with spinal dural defects (Table 1). The cases included 33 male and 8 female patients with an average age of 60.6 years (ranging from 33 to 74 years). The common definite causes were as follows: trauma (11/41), previous surgery (5/41), intervertebral disc herniation (4/41), dural ectasia (2/41), Marfan syndrome (1/41), and dural arteriovenous fistula (1/41). There were 17 cases in which the etiology was not reported. The duration from symptom onset to surgery averaged 6.81 years (ranging from 0.1 to 30 years). The most prevalent clinical manifestations were gait ataxia (31/41) and sensorineural hearing loss (28/41), followed by headache (7/41), tinnitus (6/41), dizziness (5/41), urinary incontinence (6/41), cognitive decline (4/41), limb incoordination (3/41), slurred speech (3/41), dysarthria (3/41), anosmia (2/41), neck pain (2/41), diplopia (1/41), nausea (1/41), emesis (1/41), and blurred vision (1/41). MRI indicated spinal dural defects located in the cervical spine in 5 patients and in the thoracic vertebrae in 23 patients. Most of the CSF examinations showed xanthochromia, increased red blood cells, and intracranial hypotension. Considering that SSCNS was caused by spatially defined lesions with dural defects, 34 patients were treated with reparative surgery. The repair techniques included direct suturing (8 patients), muscle grafts (4 patients), fat grafts (6 patients), fibrin glue (9 patients), patches (4 patients), gelatin sponges (3 patients), and artificial dura mater (1 patient). Postoperative MRI in most cases
showed a reduction or disappearance of epidural effusion. Among the patients with reported results, the prognosis was improved in 10 patients, partially improved in 9 patients, unchanged in 9 patients, and worsened in 3 patients. The improvement rate of headache symptoms was the highest (100%, 7/7), followed by gait instability symptoms (19.4%, 6/31); sensorineural hearing loss was not likely to improve (0%, 0/28).

In our study, the patient with SSCNS was confirmed to have intermittent subarachnoid hemorrhage caused by a cervical osteophyte that resulted in a dural defect. The repeated activity of the osteophyte led to a small amount of bleeding, which entered the subarachnoid space through the dura defect, causing the deposition of hemosiderin on the surface of the spinal cord and brain and the generation of clinical symptoms. As a result of removal of the bone spurs and repair of the dura mater, subarachnoid hemorrhage was avoided, and the symptoms improved dramatically.

Our patient’s epileptic manifestation may be related to the increase in ICP. It has been reported that the causal relationship between intracranial hypertension and epilepsy events is evident clinically and that increased cranial pressure can induce seizures. Our patient experienced severe headache before epilepsy events, accompanied by increased ICP, which further confirmed the relationship. Our patient also had elevated ICP before dural closure and even higher pressure after dural closure. We speculate that malabsorption of CSF due to dysfunction of the pachyion granulations caused by recurrent subarachnoid hemorrhage may result in chronic intracranial hypertension. Before dural closure, the dural fistula could drain some of the CSF, which is why the patient’s headache was partially relieved when he changed his position. After the dura defect was closed, the extra CSF could not be absorbed and resulted in higher ICP, which was ultimately resolved by shunt surgery.

Lessons
Our patient represents an extremely rare case of SSCNS with epilepsy originating from traumatic cervical injury. Although this situation is rare, an active search for the cause of subarachnoid hemorrhage, followed by accurate treatment, will ensure a good prognosis for such patients.

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Conception and design: Duan. Acquisition of data: Yuan, Shen. Analysis and interpretation of data: Yuan, Wang. Drafting the article: Xu. Critically revising the article: Duan. Approved the final version of the manuscript on behalf of all authors: Duan.

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