Case Report

Isolated spinal cord compression syndrome revealing delayed extensive superficial siderosis of the central nervous system secondary to cervical root avulsion

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Context: Cervical root avulsion secondary to traumatic plexus injury is a rare etiology of superficial siderosis (SS) of the central nervous system (CNS). We describe the case of an isolated progressive compressive myelopathy revealing this complication and discuss the pathogenesis of such a presentation, its clinical and imaging peculiarities with a literature review.

Findings: We report on the case of a 48-year-old man with history of left brachial plexus injury at the age of 2 years. Since the age of 38 years, he had presented with a progressive paraplegia, bladder and erectile dysfunction, neuropathic pain and sensory level. The diagnosis was made by spinal cord and brain magnetic resonance follow-up imaging revealing hypointensity T2-weighted gradient echo linear dark rim around the entire neuraxis and cervical dural pseudomeningoceles. These MRI findings were suggestive of extensive hemosiderin deposition consolidating the diagnosis of SS of CNS.

Conclusion/Clinical relevance: Our case report illustrates diagnosis difficulties in unusual or paucisymptomatic presentations of SS. A history of brachial plexus trauma with nerve root avulsion should prompt gradient-echo T2-weighted imaging to bring out such a complication. Superficial siderosis of the CNS should be included in the panel of differential diagnosis of the parathospastic syndromes and compressive myelopathy.

Keywords: Superficial Siderosis, Spastic paraplegia, Spinal cord compression, Myelopathy, Pseudomeningocele

Introduction

First described by Hamill *et al.* in 1908 as a form of hemochromatosis,^{1,2} superficial hemosiderosis or superficial siderosis (SS) of the central nervous system (CNS) is a rare neurological condition with an estimated incidence of 0.15%.^{2,3} The hallmark of SS is chronic deposition of hemosiderin along the pial and subpial structures of the brain and spinal cord, as a result of recurrent or persistent bleeding in the subarachnoid space. This subarachnoid hemorrhage is demonstrated by characteristic linear low-intensity signals along the surface of the neuraxis on susceptibility T2-weighted imaging (WI) or T2* WI magnetic resonance imaging (MRI), and leads to damage to the neural tissues.³ Clinically, SS becomes symptomatic in only 15% of

patients, with a delay between presumed onset of bleeding and first symptoms varying from four months to 37 years.^{1,4} Patients with SS usually present with slowly progressive and irreversible cerebellar ataxiasensorineural hearing loss in 81% of symptomatic patients.^{1,5,6} A myelopathic presentation is also recognized in 53% of patients, but is exceptionally isolated.¹ We hereby described an unusual case of late-onset SS developed 36 years after cervical root avulsion secondary to traumatic plexus injury and revealed by isolated progressive spinal cord compression syndrome. We discussed pathogenesis of such a presentation, its clinical and imaging peculiarities with a literature review.

Case report

A 48-year-old man, born to non-consanguineous parents, with no peculiar family history, had at the age of two a

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Figure 1 Magnetic resonance imaging of the neuraxis. T2-weighted gradient echo showing linear hypointensity due to extensive hemosiderin coating around the entire neuraxis including cerebral falx and sylvian fissures (D), the folia of the posterior fossa and brainstem, together with cerebellar atrophy prevailing on the superior vermis (C) and along the cord surface (F and G). The presence of lesions was ambiguous on T2-weighted MR imaging (A, B and E). Cervical spine MR imaging revealed cervical dural pseudomeningocele (H).

severe trauma affecting head, spinal cord and left upper limb during a major car crash. His left upper limb suffered from lack of strength and amyotrophy, which persisted with no other sequelae. At the age of 38, he began to present progressive weakness of first his left, and then his right lower limbs, associated with erectile and urinary dysfunction requiring self-bladder catheterization. He became totally bedridden 6 years later, with decreased sensitivity and neuropathic pain in lower limbs, which was more marked on the left side. At the age of 46 years, neurological examination showed pyramidal syndrome in lower limbs with brisk deep tendon reflexes, Babinski sign and spastic paraplegia. The left upper limb was atrophic with proximal motor deficit and abolished deep tendon reflexes. He had a sensitive level localized at the 10th dorsal vertebrae involving tactile, thermal and nociceptive sensitivity, in addition to hypopallesthesia in lower limbs. The hematological and biochemical laboratory tests were normal. The immunological tests (antinuclear antibodies, anti-native DNA antibodies, anti-extractable nuclear antigens (ENA) antibodies, anti-neutrophil cytoplasm antibodies (ANCA), anti-phospholipid antibodies, Rheumatoid Factor), tumor markers (Prostate-Specific Antigen (PSA), Carcinoembryonic antigen

(CEA), α -fetoprotein (AFP), CA19-9 and CA-125), antineuronal antibodies (anti-Cv2, anti-PNMA2, anti-Ri, anti-Yo, anti-Hu, anti-Recoverin, anti-SOX1, anti-Titin) and infectious serologies (HIV, hepatitis B and C, syphilis and borreliosis) were all negative. Lumbar puncture was not performed for absence of consent. Electromyogram (EMG) showed multiple radiculopathy of the left upper limb from C3 to C7. Brain and spinal magnetic resonance imaging (MRI) revealed T2-weighted gradient echo linear dark rim of hypointensity around the entire neuraxis including cerebral falx and sylvian fissures (Fig. 1D), the folia of the posterior fossa and brainstem, associated with cerebellar atrophy prevailing on the superior vermis (Fig. 1C). In addition, the susceptibility-weighted MR images of the spinal cord showed similar findings along the cord surface (Figs. 1F and G). The presence of lesions was ambiguous on T2-weighted MR imaging (Figs. 1A, B and E). Cervical spine MR imaging also revealed post-traumatic dural pseudomeningoceles of the left C3 to C7 nerve roots. Magnetic Resonance Angiography (MRA) was normal. These MRI findings were suggestive of an extensive hemosiderin deposition on the surface of both hemispheres of the brain (supra and infra-tentorial regions) as well as the spinal cord in accordance with the

Table 1 Literature review of patients with brachial plexus/root avulsion and delayed superficial siderosis.

Authors	Age (years)/ Sex	Interval from injury to symptoms (years)	Root avulsion level	Presenting signs and symptoms	Interventions
Lewey <i>et al.</i> ; 1942 ⁸	NS/NS	NS	NS	posterior fossa exploration; hemosiderin-stained cerebellum	NS
Bracchi <i>et al.</i> , 1993 ⁹	NS/Male	17	C6-C7	Hearing loss, ataxia, anosmia, paraparesis, incontinence, facial nerve palsy	None
Bonito <i>et al.</i> , 1994 ⁹	47/Male 51/Male	17 10	C-8 C-8	anosmia, hypotonia, ataxia progressive ataxia, genitourinary dysfunction	None repair of pseudomeningocele, coagulation of scar
Tapscott <i>et al.</i> , 1996 ⁹	46/Male	16	C5-C6	tinnitus, sensorineural hearing loss, imbalance	exploration of C-5 pseudomeningocele; large vein in the wall cauterized
Maggioni <i>et al.</i> , 1997 ⁹	51/Male	21	NS	hearing loss, ataxia	None
Durieux <i>et al.</i> , 1999 ⁹	NS/Male	20	NS	Hearing loss, ataxia, myelopathy	None
Konitsiotis <i>et al.</i> , 2002 ⁹	45/Male	19	Cervical roots	hearing loss, ataxia; paraparesis, and headaches	None
Kole <i>et al.</i> , 2004 [9]	51/Male	22	C8-T1	hearing loss, ataxia, dysarthria	Epidural vein coagulation
Cohen-Gadol <i>et al.</i> , 2004 ⁹	42/Male	13	C7 et T1	hearing loss, ataxia, paraparesis, memory disturbance	coagulation of abnormal vessels in pseudomeningocele
	52/Male 69/Male	37 36	C7-T1 T1	hearing loss, ataxia hearing loss, ataxia, decreased sense of taste and smell	resection and ligation of pseudomeningocele None
Anquilina <i>et al.</i> , 2005 ⁹	34/Male	9	C7-T1	Hearing loss, ataxia, anosmia, paraparesis, urinary incontinence, anisocoria	Coagulation of abnormal and friable vessels
Miliaras <i>et al.</i> , 2006 ⁹	45/Male	17	C7	Hearing loss, ataxia, paraparesis, memory disturbance, headache	Excision of meningeal diverticulum
Aoyama <i>et al.</i> , 2009 ¹⁰	36/Male	30	C1-C2	hearing loss and ataxia	Removal of Malpositioned spinal instrument
Papadimas <i>et al.</i> , 2009 ⁹	49/Male	21	NS	Ataxia, spastic paraparesis	None
Nunes <i>et al.</i> , 2011 ¹¹	38/Male	NS	NS	Ataxia, hearing loss, tinnitus and memory loss	NS
Tari-Capone <i>et al.</i> ; 2013 ⁹	20/Male	6	C7-T1	Headache, meningism	Coagulation of abnormal vessels in pseudomeningocele
	54/Male	5	NS	Ataxia, hearing loss, cognitive impairment and micturition disturbance	Resection of pseudomeningocele None
Saft <i>et al.</i> ; 2014 ¹²	48/Male	26	C6-T1	Noon pseudoradicular lumbosacral Pain	Surgery of a pseudomeningocele
Decramer <i>et al.</i> ; 2016 ¹³	60/Male	19	NS	Ataxia, hearing loss, hyposmia, tinnitus	Posterior cervical laminectomy, tissue graft

Continued

Table 1 Continued

Authors	Age (years)/ Sex	Interval from injury to symptoms (years)	Root avulsion level	Presenting signs and symptoms	Interventions
Sinha <i>et al.</i> ; 2017 ¹⁴	NS/NS	NS	C7-T1	Ataxia, hearing loss, dysarthria, tinnitus	C6 to T1 laminectomies Coagulation of intradural bleeding vessels
	NS/NS	18	C5-C6	Left hemiparesis, ataxia, dysarthria, urinary urgency, diplopia, cognitive decline	Reparation of dural defect Application of artificial dura and tissue glue followed by layered closure
	NS/NS	23	C1-C3	ataxia, dysarthria, dysphagia, diplopia, anosmia, hearing loss,	C1-C3 laminectomy Microscopic intradural exploration a Separation and cauterization of a bleeding point from the dissection site Dural reparation
Our patient	48/Male	36	C3-C7	Isolated spinal cord compression syndrome	None

NS, not specified.

diagnosis of SS of CNS. He received vitamin B, amitriptyline and pregabalin for neuropathic pain and baclofen for spasticity. Corticosteroids were considered, yet never attempted for recurrent urinary infections. Surgical intervention for pseudomeningoceles was also discussed but not performed for absence of consent from the patient.

Discussion

We reported on an exceptional case of isolated progressive spinal cord compression syndrome revealing CNS superficial hemosiderosis secondary to cervical root avulsion by traumatic plexus injury with formation of cervical pseudomeningoceles. In fact, brachial plexus injury and nerve root avulsion are commonly associated.⁷ However, this association has been recognized as an extremely rare etiology of deposition of hemosiderin on leptomeninges and subpial layers of brain and spinal cord. Table 1 summarizes all case records of cervical nerve root avulsion or brachial plexus injury complicated with CNS-SS in the literature.⁸⁻¹⁴ The injury of nerve root sleeves leads to occult extradural CSF leak through small defects in the meninges with spinal extra-arachnoid fluid collection formation.¹⁵⁻¹⁷ These fluid-filled cavities, located within the spinal canal and outside the thecal sac, have been referred to as pseudomeningoceles.^{4,17} Subarachnoid bleeding due to traumatic pseudomeningocele of the brachial plexus is a very unusual cause of superficial siderosis.^{17,18} The associated dural tears may lead to scar formation with fragile vessels that could bleed intermittently and slowly causing SS.⁴ Presence of an osmotic gradient between the cystic collection and the subarachnoid space with a valve-like mechanism and pulsatile CSF dynamics lead to the expansion of the extradural spinal cyst. Consequently, this can cause cord compression and cord signal changes.¹⁶ Hence, pseudomeningoceles may present with symptoms related to compressive myelopathy from the collection or SS.^{17,20} Although pyramidal and sensory signs and bladder dysfunction are common in SS, sensory symptoms or a sensory level, as found in this case, are rarely present and may relate to brainstem or spinal cord involvement in SS.^{4,15,21,22} The long interval between the brachial plexus trauma and the onset of symptomatic SS in our patient is consistent with previously reported long presymptomatic phase (Table 1). Our patient complained of progressive spastic paraplegia for 8 years. To our knowledge, the isolated myelopathic presentation suggestive of spinal cord compression syndrome in such a context has never been reported (Table 1).⁴ In our patient, despite

extensive investigations including neuroimaging studies in multiple medical institutions, no etiological cause was established until recently. MRI is the diagnostic modality of choice.⁴ Gradient-echo T2-weighted images have higher sensitivity for hemosiderin deposition imaging typically showing a rim of hypointensity with a predilection for superior vermis, quadrigeminal plate and basal cerebral surface but may also involve the sylvian fissure, interhemispheric fissure, and cortical sulci as found in our case.^{3,4} Nevertheless, particularly in the early stages, the findings may be subtle, the T2 hypointensity following the contours of the brain and spinal cord may be easily missed. In our patient, several brain and spinal cord MRI have been performed without gradient-echo T2-weighted images hampering the detection of SS related abnormalities.^{3,4,15} Cerebellar atrophy is often present in SS as found in our case.³ Because SS is associated with a progressive and disabling course, thorough imaging evaluation of the entire neuraxis is indicated to locate accurately any potential bleeding source, such as pseudomeningocele as in our patient.^{4,15} Currently, there is no evidence that any treatment, including removal of an identified source of bleeding could impede disease progression.^{2,23} Iron chelation therapy by desferrioxamine or copper chelation therapy by trienterine did not show significant success in SS.^{2,4} However, it has been recently demonstrated that deferiprone, a lipid-soluble iron chelator that is able to penetrate the blood-brain barrier, could be effective at improving deposition of hemosiderin and clinical symptoms of SS.¹⁶ Besides, steroid therapy, which was considered for our patient, could constitute a new track for the treatment of siderosis of CNS.²³ Treatment of SS also involves identification and surgical correction of the potential bleeding source.^{4,16} Excision of the pseudomeningocele, coagulation of friable vessels in the scarred wall of the pseudomeningocele repair and closure of dural defects caused by root avulsion injury could be a reasonable therapeutic strategy to prevent further intrathecal bleeding and additional neuronal injury.^{4,15} However, because the hemosiderin deposits secondary to preoperative bleeds may continue to injure neuronal tissues, prevention of further hemorrhage may not make a significant difference in the overall clinical outcomes.⁴ In our case, surgical intervention was not approved by the patient who received only symptomatic treatment.

Conclusion

Our case report illustrates clinical and imaging difficulties hampering an early diagnosis of SS, especially with unusual or paucisymptomatic presentations. Even after

a prolonged delay, a history of brachial plexus trauma with nerve root avulsion should prompt thorough imaging investigation of the entire neuraxis imperatively including gradient-echo T2-weighted images to bring out such a complication. SS of the CNS should be included in the panel of differential diagnosis of the parathospastic syndromes and compressive myelopathy.

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