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CASE REPORT

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Duropathy as a rare motor neuron disease mimic: from bibrachial amyotrophy to infratentorial superficial siderosis

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Abstract

Background Bibrachial amyotrophy associated with an extradural CSF collection and infratentorial superficial siderosis (SS) are rare conditions that may occasionally mimic ALS. Both disorders are assumed to be due to dural tears.

Case presentation A 53-year-old man presented with a 7-year history of slowly progressive asymmetric bibrachial amyotrophy. Initially, a diagnosis of atypical motor neuron disease (MND) was made. At re-evaluation 11 years later, upper limb wasting and weakness had further progressed and were accompanied by sensorineural hearing loss. MRI of the brain and spine demonstrated extensive supra- and infratentorial SS (including the surface of the whole spinal cord) as well as a ventral longitudinal intraspinal fluid collection (VLISFC) extending along almost the entire thoracic spine. Osteodegenerative changes were observed at C5-C7 level, with osteophytes protruding posteriorly. The bony spurs at C6-C7 level were hypothesized to have lesioned the dura, causing a CSF leak and thus a VLISFC. Review of the MRI acquired at first evaluation showed that the VLISFC was already present at that time (actually beginning at C7 level), whereas the SS was not. 19 years after the onset of upper limb weakness, the patient additionally developed parkinsonism. Response to levodopa, brain scintigraphy with ^{123}I-ioflupane and brain MRI with nigrosome 1 evaluation were consistent with idiopathic Parkinson's disease (PD). On the latest follow-up 21 years after symptom onset, the VLISFC was unchanged, as were upper arm weakness and wasting.

Conclusions Based on the long-term follow-up, we could establish that, while the evidence of the VLISFC was concomitant with the clinical presentation of upper limb amyotrophy and weakness, the radiological signs of SS appeared later. This suggests that SS was not per se the cause of the ALS-like clinical picture, but rather a long-term sequela of a dural leak. The latter was instead the causative lesion, giving rise to a VLISFC which compressed the cervical motor roots. Dural tears can actually cause several symptoms, and further studies are needed to elucidate the pathophysiological correlates of "duropathies". Finally, as iron metabolism has been implicated in PD, the co-occurrence of PD with SS deserves further investigation.

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Keywords Superficial siderosis, Bibrachial amyotrophy, Duropathy, Dural leak, Ventral longitudinal intraspinal fluid collection, ALS mimics

Introduction

Bibrachial amyotrophy associated with an extradural cerebrospinal fluid (CSF) collection and infratentorial superficial siderosis (SS) are rare conditions that can occasionally mimic amyotrophic lateral sclerosis (ALS).

Classic infratentorial SS results from the deposition of hemosiderin in the subpial layers of the brain and spinal cord and is characterized by sensorineural hearing loss, cerebellar ataxia, and myelopathy-related symptoms [1, 2]. Around 5–10% of cases present lower motor neuron (LMN) features with muscle wasting [1], and to our knowledge, since the first description of the disease in 1908 [3], six cases have been associated with a motor neuron disease (MND) phenotype [4–9].

Most infratentorial SS cases are caused by a dural defect that leads to a persistent low-volume bleeding into the subarachnoid space (SAS) from fragile vessels at its borders [2, 10, 11]. Additionally, extra-arachnoid fluid collections caused by CSF extravasation have been frequently reported [12, 13], and – since they are commonly located ventrally and extend longitudinally through multiple spinal levels – the term ventral longitudinal intraspinal fluid collection (VLISFC) was coined [14].

Yet, the prototypical condition related to CSF leaking is spontaneous intracranial hypotension (SIH) and in its context three types of leaks have been identified [15]. SS has been mostly observed in patients with type 1a dural defects – i.e. a small ventrally located dural slit due to osteodegenerative spinal disease –, and occasionally with type 2 defects (meningeal diverticula) [16, 17]. Apart from SS, ventral spinal CSF leaks have been associated with other two long-term sequelae: bibrachial amyotrophy and spinal cord herniation [18, 19].

We hereby report a case of infratentorial SS associated with VLISFC that developed more than a decade after the onset of isolated bibrachial amyotrophy and was initially misdiagnosed as atypical, slowly progressive MND. Moreover, 19 years after the initial symptoms, the patient developed parkinsonism, making this the second described case of parkinsonism associated with infratentorial SS.

Case presentation

A 53-year-old man initially presented in December 2008 with a 7-year history of asymmetrical (left greater than right) upper arm weakness and amyotrophy. He was diagnosed with a slowly progressive flail arm variant of ALS and referred for follow-up. The patient was admitted to our unit almost 11 years later (July 2019) complaining of progressive muscle weakness. His past medical history was notable for tympanoplasty in the right ear due to spontaneous rupture of the tympanic membrane. There was no history of neurosurgical interventions, trauma, macrovascular bleeding events or orthostatic headache. Upon admission, cognitive functions were intact and mental status appeared normal with no speech or language deficit. Cranial nerves were spared except for bilateral sensorineural hearing loss and conductive hearing loss on the right side. Manual muscle strength testing demonstrated weakness on the Medical Research Council scale (right/left) in deltoid (4/2), biceps brachii (4/3), and supraspinatus (5/3). There was a marked reduction in muscle bulk of shoulder girdles, more pronounced on the left. Lower-extremity, respiratory and bulbar musculature was spared. Deep tendon reflexes of the upper extremities (biceps, triceps, and brachioradialis) were normal on the right and weak on the left. Plantar responses were flexor. There was no limb or gait ataxia.

Needle electromyography (EMG) showed chronic neurogenic changes in the upper limbs, more marked on the left and with a proximal>distal gradient, and, to a lesser extent, in the lower limbs, without signs of active denervation. More specifically, the deltoid muscles (innervated by spinal segments C5 and C6) had polyphasic motor unit potentials with increased amplitude and duration recruited with a single-unit pattern on the left and a poor transitional/single-unit pattern on the right; the biceps brachii muscles (C5-C6) (and the right brachioradialis muscle, C5-C6) had polyphasic potentials with increased amplitude and duration recruited with a poor transitional/single-unit pattern on the left and a poor transitional pattern on the right; the triceps muscles (C6-C7-C8) (and the right flexor carpi radialis, C6-C7) had polyphasic potentials with normal or increased amplitude and increased duration recruited with a rich transitional pattern; the first dorsal interosseus muscles (C8-T1) had polyphasic potentials with normal amplitude and duration recruited with a rich transitional pattern. The examined muscles in the lower limbs (right vastus medialis (L2-L3-L4), right and left tibialis anterior (L4-L5), left medial gastrocnemius (S1-S2)) had potentials with normal amplitude and duration recruited with a normal pattern.

CSF analysis revealed xanthochromia and a raised red blood cell (RBC) count (1300/μL; normal value, 0/μL). Brain magnetic resonance imaging (MRI) disclosed diffuse hemosiderin deposition: specifically, marked hypointensity on T2-weighted imaging (T2WI) along the contours of the posterior cranial fossa structures (brainstem, cerebellar folia and vermis) (Fig. 1-A) as well as

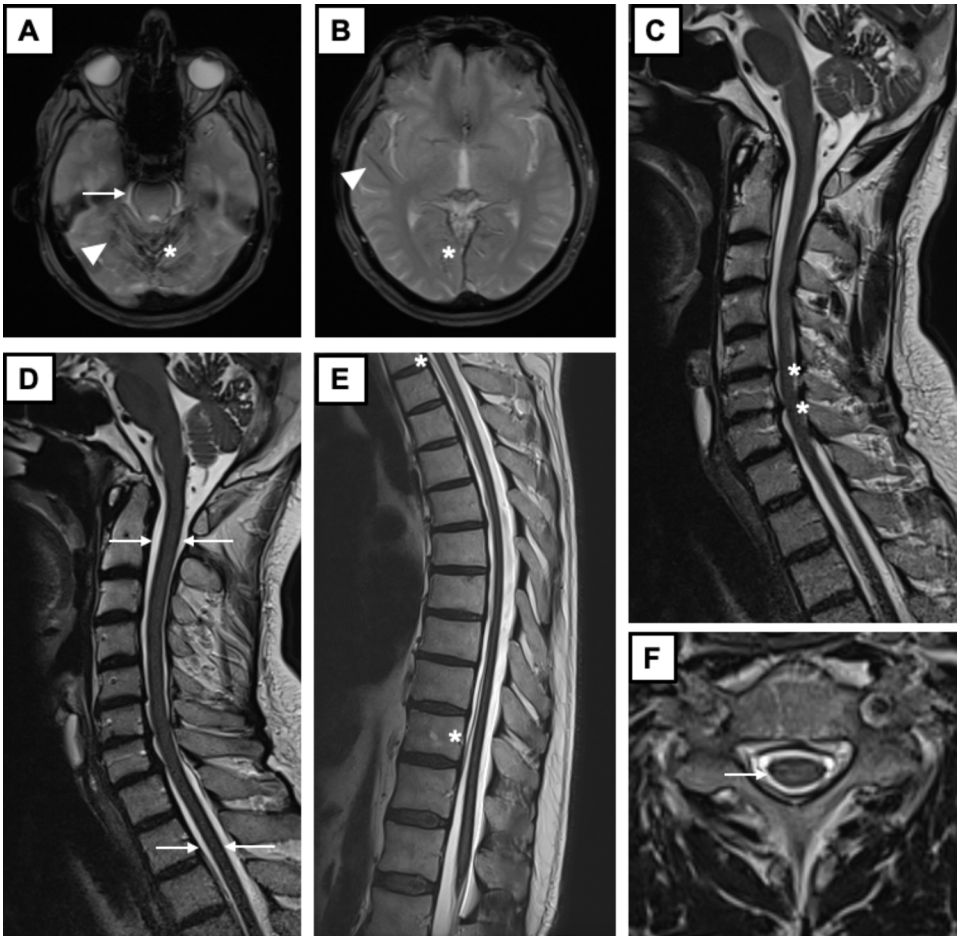

Fig. 1 MRI images of July 2019. **A**: axial GRE image showing infratentorial hemosiderin deposition (pons (white arrow), cerebellar vermis (white asterisk), cortex of cerebellar hemispheres (white arrowhead)). **B**: axial GRE image showing supratentorial hemosiderin deposition (right temporal and insular cortex (white arrowhead), bilateral medial parieto-occipital cortex (white asterisk)). **C**: T2-TSE sagittal image of the cervical cord showing osteophytes and disc protrusions at C5-6-7 levels (white asterisks). **D** and **E**: T2-TSE sagittal images of the cervical and dorsal cord, respectively, showing hypointense rim of hemosiderin deposition along the entire length of the cord (white arrows) and a VLISFC extending from T2-T3 level to T11 level (white asterisks). **F**: axial T2-SPACE image at C6 level showing the hypointense rim of hemosiderin deposition on the cord surface (white arrow) but no signal alterations of the anterior horns. *GRE*, *gradient-recalled echo. MRI*, *magnetic resonance imaging. TSE*, *turbo spin echo. VLISFC*, *ventral longitudinal intraspinal fluid collection*

sulcal hemosiderin deposits on frontal cortices, Sylvian surfaces of the temporal lobes, and in sulci of parasagittal frontal, parietal, and occipital cortices (Fig. 1-B). MRI of the spinal cord showed osteodegenerative changes at C5-C7 levels (bulging of the corresponding intervertebral discs and osteophytes protruding posteriorly into the SAS) (Fig. 1-C) and a hypointense rim of hemosiderin deposition along its entire length with a VLISFC extending from T2-T3 level to T11 level (Fig. 1-D-E). There were no radiological signs of intracranial hypotension. Anterior horns of the cervical cord appeared intact on axial view (Fig. 1-F). Regarding the lower spine, disc protrusions were present from L2 to S1, with impingement on the dural sac and on the lower parts of intervertebral foramina, a finding which was most evident at L3-L4 level, where a slight spinal stenosis was observed. We reviewed the MRI acquired 11 years before, which included a gradient-recalled echo (GRE) sequence but did not show signs of hemosiderin deposition. Interestingly, osteophytes at C5-C7 level and the VLISFC were already evident at that time; the latter appeared more voluminous compared to our current observation, beginning at C7 level (Fig. 2). We thus hypothesized that the bone spur at C6-C7 level had lesioned the dura. Although 3D T2 DRIVE sequences were employed, no dural leak was found. We did not proceed to perform digital subtraction myelography and/or CT myelography [2] given the disease duration, the relative stability of neurologic deficits and the progressive reduction of the VLISFC. The patient was diagnosed with bibrachial amyotrophy caused by a VLISFC from an unidentified type 1a CSF leak and SS as late complication of a long-standing dural tear.

Notably, more than one year later (September 2020) the patient developed rest tremor of the right hand. He was

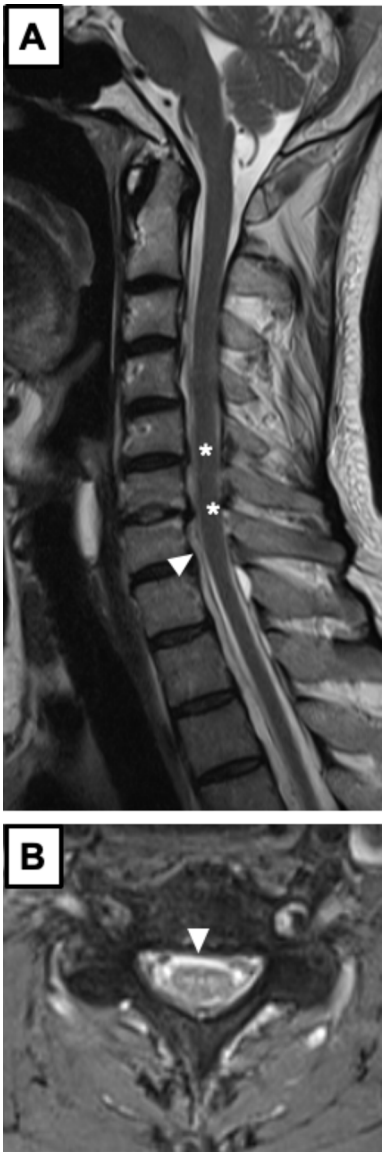

Fig. 2 MRI images of December 2008. **A**: sagittal T2-TSE image showing osteophytes at C5-6-7 levels (white asterisks) and the VLISFC beginning at C7 level (white arrowhead), but no rim of hemosiderin deposition along the surface of the cord. **B**: axial T2-me2d image at C6 level showing the VLISFC (white arrowhead) but no hemosiderin deposition on the cord surface. *MRI, magnetic resonance imaging. TSE, turbo spin echo. VLISFC, ventral longitudinal intraspinal fluid collection*

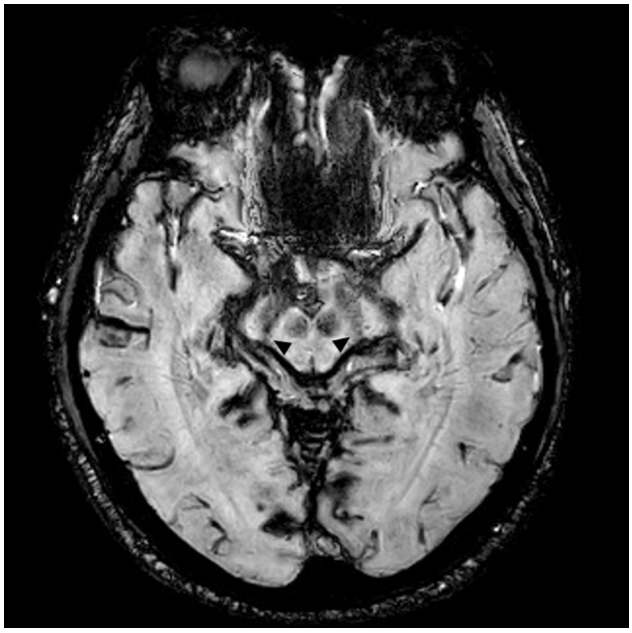

Fig. 3 MRI image of Agust 2021. SWI image showing bilateral absence of the hyperintense signal of nigrosome 1 (black arrowheads). *MRI magnetic resonance imaging. SWI, susceptibility-weighted imaging*

therefore again admitted to our unit in May 2021. Neurological examination confirmed the tremor and disclosed bradykinesia and mild rigidity of the right upper limb, in addition to the already known neuromuscular deficits. A levodopa challenge test was positive. Brain scintigraphy with ^{123}I-ioflupane (DaTSCAN®) revealed depletion of the dopaminergic presynaptic nerve terminals in the putamina, while 3-Tesla (3T) brain MRI showed loss of nigrosome-1 signal bilaterally, pointing to dopaminergic neuron loss in the pars compacta of the substantia nigra (SN) (August 2021) (Fig. 3). MRI of the brain and spine confirmed previous findings. Dopamine substitution therapy was started (melevodopa/carbidopa 100/25 mg tid along with a rotigotine transdermal patch of 4 mg/24 hours), with a positive effect on tremor and rigidity.

On the latest neuroradiological follow-up in October 2022, the VLISFC was unchanged (Fig. 4). Analysis of CSF obtained by a further lumbar puncture in December 2022 revealed ongoing chronic microscopic subarachnoid bleeding, with elevated RBC count (1000/μL; normal value, 0/μL), presence of siderophages on microscopic examination, and increased ferritin level (101.6 ng/mL; normal value, <12 ng/mL [20]). The patient remained clinically stable in terms of upper arm weakness and wasting, as confirmed at the latest neurological evaluation in February 2024.

Discussion

Dural abnormality has been observed as a common denominator in infratentorial SS (dural tear recognized as its cause in over 80% of patients [2]), SIH, multisegmental amyotrophy and spinal cord herniation, leading to the introduction of the term "duropathies" [14]. The pathophysiology and the relationship between overlapping duropathies remain uncertain. The type of dural defect can partially account for symptoms, but it remains unclear why some patients develop only one manifestation of the possible spectrum of duropathies while others present with overlapping conditions.

The first report establishing the link between dural leak, intraspinal fluid collection and anterior horn cell

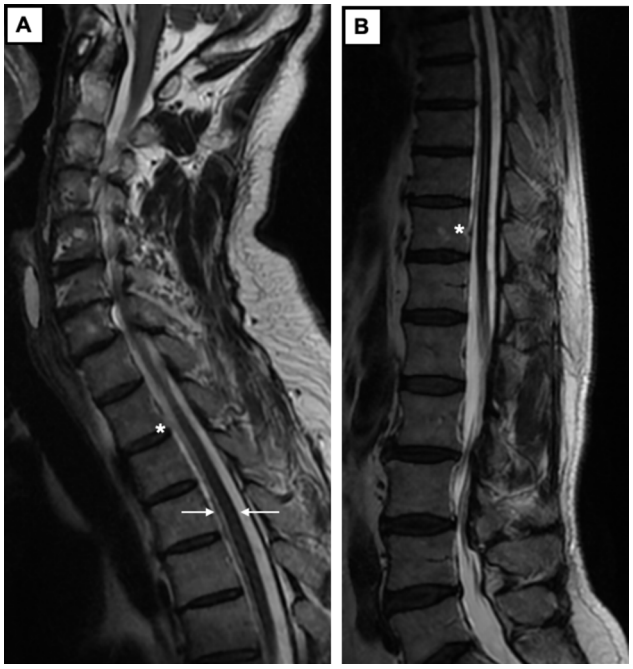

Fig. 4 MRI images of October 2022. Sagittal T2-TSE images of the cervical (**A**) and lumbar spine (**B**) showing persistence of the hypointense rim of hemosiderin along the surface of the cord (white arrows) as well as persistence of the VLISFC, beginning approximately at T2-T3 level (white asterisk in image **A**) and ending at T11 level (white asterisk in image **B**). *MRI, magnetic resonance imaging. TSE, turbo spin echo*

damage dates back to 2011 [21]. In a recent review of bibrachial amyotrophy as a rare manifestation of intraspinal fluid collection, 44 patients have been described; of note, only 39% of cases were associated with the presence of SS [22]. In another study, SS has been documented in 9% of the patients with a CSF leak [11]. Additionally, it was shown that in patients with a chronic dural leak and SIH, the risk of developing SS was about 50% and that of bibrachial amyotrophy about 20% at 15 years of follow-up [18]. Although the connection between SIH and SS – with the former typically preceding the latter – has been well established, some patients with SS have no history of SIH-related symptoms.

In the past, LMN involvement in infratentorial SS has been thought to originate from arachnoiditis or radiculopathy, with some authors suggesting the deposition of hemosiderin at the motor nerve root exit zone [9] or directly at the level of anterior horn cells. As for the pathophysiology of multisegmental amyotrophy in association with a VLISFC (whether accompanied by SS or not), two mechanisms have been suggested: compression from the VLISFC resulting in anterior horn cell damage and/or stretching of the motor nerve roots due to posterior displacement of the spinal cord by the VLISFC [23–25]. The pathological analysis of a case of bibrachial amyotrophy with SS and dural tear suggested the pivotal role of VLISFC in the development of anterior horn damage and excluded hemosiderin deposition as its main cause [26]. Of note, in all but one of the reported cases of infratentorial SS presenting with an ALS-like phenotype, intraspinal fluid collections have been described, upholding this hypothesis [4–9]. Moreover, we are not aware of cases of infratentorial SS preceding the development of brachial amyotrophy. On the contrary, the infratentorial SS could be responsible for the patient's bilateral sensorineural hearing loss, while the conductive component on the right side was secondary to previous spontaneous rupture of the right tympanic membrane, which had been treated with tympanoplasty. This aspect of our case is – independently from the main issue of bibrachial amyotrophy – a reminder that the occurrence of not otherwise explained bilateral sensorineural hearing loss should prompt diagnostic consideration of SS.

In our case, clinical and electrodiagnostic findings suggested the localization of pathology to cervical motor roots or to anterior horn cells at C5-C6 levels and to a lesser extent at lower cervical levels [27]. The patient's history was not indicative of arachnoiditis as he did not complain of back, neck or radicular pain. Wasting and weakness preceded by many years the radiological appearance of hemosiderin deposition, suggesting – in agreement with previous reports – that infratentorial SS per se is not likely to be responsible for the ALS-like phenotype [24]. On the other hand, it appears equally unlikely that the C5-C6 spinal cord segments were affected by the fluid compression starting at C7 vertebral level. On the contrary, the C5-C6 spinal nerve roots might have been affected along their way to the corresponding intervertebral foramina; indeed, the C5 motor nerve roots have been reported to be particularly vulnerable to cervical cord tethering due to VLISFCs [22, 25]. The relative stability of the patient's condition throughout the 21-year clinical course might be partially explained by the lack of increase of the fluid collection over the years. Collaterally, the modest neurogenic changes found on needle EMG of the lower limbs may be attributed to the degenerative changes of the lower spine observed on MRI.

Most cases of VLISFC occur between cervical and thoracic levels, a finding that in part can be explained by the high prevalence of osteodegenerative abnormalities at this level; indeed, dural defects have been reported to be caused by osteophytes, a disc-osteophyte complex, or calcification [13, 23]. In fact, even though a dural leak was not identified in our case, MRI revealed osteophytes at C5-C6-C7 levels that might have been the cause of the dural tear, as further suggested by the fluid collection starting at C7 level. It remains unclear why, despite the longitudinally extensive intraspinal fluid collection, only the cervical segment was neurologically affected. A possible explanation for that could be greater mobility of the

cervical spine. Dynamic spinal imaging might be needed to assess the relationship between neck motion, possible VLISFC displacement, and spinal cord compression, similarly to what is performed for diagnosing Hirayama disease, where, however, the neurological deficits (wasting and weakness) and the EMG changes are more distal (C8-T1 segments) and the primary pathology is posterior rather than anterior (forward shift of the posterior wall of the dural sac) [28]. Regarding the lacking identification of the precise site of the dural tear and consequent CSF leak, our choice of a conservative approach was motivated by the stability of the clinical picture and the will to avoid invasive procedures. Nevertheless, we recognize that a slightly more aggressive conduct would have been justified, especially considering that symptoms of SIH may also appear later than bibrachial amyotrophy in cases of VLISFC, as reported by other authors [27]. However, in the latter case, the time interval between the appearance of weakness and the development of orthostatic headache was 26 months at most, while our patient did not report headache even at the latest follow-up occurring 22 years after onset of bibrachial amyotrophy.

After more than one year of clinical stability following the diagnosis, at the age of 65, our patient developed features suggestive of Parkinson's disease (PD). The latter is a frequent neurological disorder, with an estimated prevalence of 0.4% in the general population aged 60 to 69 [29]. Both the good motor response to dopaminergic medication and neuroimaging support the idiopathic origin of the disorder in this case. Additionally, the late occurrence of extrapyramidal involvement without significant worsening of other SS-related features suggests its independent origin rather than its secondary nature as a manifestation of SS progression. Besides, SS pathology is characterized by marginal siderosis; hence, direct iron deposition in deep structures such as the SN and striatum appears anatomically unlikely, even more without involvement of cerebral peduncles. Conversely, given the proximity of the SN to the CSF of the interpeduncular cistern and the fact that brain parenchyma can be affected up to a depth of 3 mm in SS [30], a causal relationship between SS and parkinsonism in our case cannot be excluded either, especially in lack of pathological data. Accumulation of iron has been implicated in the pathophysiology of many neurodegenerative diseases, including PD, in which an increased iron content has been specifically observed in the areas of dopaminergic neuron loss within the SN [31]. We found one report of parkinsonism occurring together with neuroradiological findings of SS, in which the authors ascribed all neurological manifestations to the widespread central nervous system (CNS) lesions due to SS rather than to idiopathic nigrostriatal degeneration [32]. Further studies are needed to elucidate the link between iron metabolism abnormalities, SS, and PD.

Conclusions

We have described an unusual clinical case characterized by long-standing asymmetrical upper-limb amyotrophy and weakness with neuroimaging evidence of infratentorial SS and a VLISFC. The longitudinally extended evaluation of the case over time enabled us to establish that, while the finding of the VLISFC was concomitant with the clinical presentation of upper limb weakness and wasting, the radiological signs of infratentorial SS appeared later. This suggests that the ALS-like clinical picture was not secondary to infratentorial SS. The actual causative lesion had instead to be an unidentified dural leak, which had given rise to a VLISFC. The latter, in turn, compressed the cervical motor roots. The infratentorial SS was rather a long-term sequela of the dural tear, being produced by chronic bleeding into the SAS. However, infratentorial SS was probably responsible for the patient's long-standing bilateral sensorineural hearing loss. Conversely, the possible link between the later development of parkinsonism/PD and the earlier clinical manifestations is uncertain. As infratentorial SS and bibrachial amyotrophy are only some of the diverse clinico-pathological manifestations of dural tears, further studies are needed to elucidate the pathophysiological correlates of "duropathies". Finally, as iron metabolism might be relevant for the etiopathogenesis of PD, the co-occurrence of PD with SS deserves further investigation.

Abbreviations

ALS	Amyotrophic lateral sclerosis
CNS	Central nervous system
CSF	Cerebrospinal fluid
DRIVE	Driven equilibrium fast spin echo
EMG	Electromyography
GRE	Gradient recalled echo
LMN	Lower motor neuron
MND	Motor neuron disease
MRI	Magnetic resonance imaging
PD	Parkinson's disease
RBC	Red blood cell
SAS	Subarachnoid space
SIH	Spontaneous intracranial hypotension
SN	Substantia nigra
SS	Superficial siderosis
T2WI	T2-weighted imaging
VLISFC	Ventral longitudinal intraspinal fluid collection

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Author contributions
The study was conceived by FV. VI performed a literature search and wrote the manuscript. FV critically reviewed the pathophisiological hypotheses presented in the discussion. FV, CC, AS, GC and AE provided the neuroradiological images. CC and AE discussed the diagnostic hypotheses from the neuroradiological point of view. EC and EC contributed neurochemical data. FV, VS and NT critically reviewed the manuscript. All authors approved the final version of the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
The study was approved by the Ethics Committee of IRCCS Istituto Auxologico Italiano (code 2021_05_18_04). The patient whose case is described in this manuscript provided written informed consent for participation.

Consent for publication
The patient whose case is described in this manuscript provided written informed consent for publication.

Competing interests
The authors declare no competing interests.

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