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ARTICLE

Specific electromyography characteristics can distinguish longitudinally extensive transverse myelitis from congestive myelopathy due to spinal dural arteriovenous fistula: a retrospective study

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Abstract

Aims/Background Although electromyography has been extensively used in the diagnosis of neurological diseases, there is no comprehensive understanding of the electromyography manifestations of spinal dural arteriovenous fistula. Given the widespread use of electromyography in the diagnosis of neurological conditions, it is worthwhile to holistically analyse the electromyography findings of spinal dural arteriovenous fistula to differentiate it from neurological diseases that share similar clinical manifestations. The aim of this study is to evaluate whether electromyography can distinguish spinal dural arteriovenous fistula from longitudinally extensive transverse myelitis.

Methods We holistically reviewed files of all patients who were diagnosed with spinal dural arteriovenous fistula or longitudinally extensive transverse myelitis at The First Medical Centre of PLA General Hospital from 1 January 2010 to 31 December 2020. We compared the symptomology, epidemiology, and imaging results of patients with spinal dural arteriovenous fistula and longitudinally extensive transverse myelitis, placing emphasis on their electromyography manifestations. Student's t test was used to analyse normally distributed data, while Chi-square test was used to compare classification statistics.

Results Lesions of spinal dural arteriovenous fistula shown on images tend to appear at lower lumbar and sacral segments, whereas lesions of the cervical and upper thoracic segments are more characteristic of longitudinally extensive transverse myelitis. Spinal dural arteriovenous fistula patients and longitudinally extensive transverse myelitis patients overlap in terms of clinical manifestations. After comparison, the two groups of patients had different demographics (age, sex), onset mode, predisposing factors before onset, and electromyographic features. The electromyographic features of patients with spinal dural arteriovenous fistula were associated with neurogenic damage ($p < 0.001$).

Conclusions In patients with spinal dural arteriovenous fistula, electromyography can help clinicians to identify early disease, avoid patient treatment delay, and eliminate unnecessary treatment.

Key words: Electromyography; Longitudinally extensive transverse myelitis; Neuromyelitis optica spectrum disorder; Spinal dural arteriovenous fistula

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Introduction

Spinal dural arteriovenous fistula (SDAVF) is a common disease associated with vascular malformation in the spinal cord, with an occurrence rate of 50–80% of all spinal vasculopathies (Jellema et al, 2006). The disease was first described by Foix and Alajouanine in 1926 after they had examined the clinical files and autopsy results of two patients with 'progressive myelopathy' (Jellema et al, 2006). Although almost 100 years have passed since the initial description of SDAVF, it is still clinically challenging to definitively diagnose SDAVF. Despite the rapid technological progression of clinical imaging modalities, over 60% of patients with SDAVF continue to be misdiagnosed with various forms of longitudinally extensive transverse myelitis (LETM) due to their similar characteristics on imaging. It has been shown that establishing a correct diagnosis of SDAVF via spinal angiography takes

an average of 281 days if it is preceded by a misdiagnosis (Hunt et al, 2018), and a delay in diagnosis can significantly increase the risk of paralysis (Iovtchev et al, 2015; Brinjikji et al, 2016). Therefore, it is essential to identify which methods are effective in the prompt diagnosis of SDAVF to avoid unnecessary delay in treatment due to misdiagnosis.

Longitudinally extensive transverse myelitis is defined as myelitis that extends over at least three vertebral segments as shown in spinal cord magnetic resonance imaging (MRI) (Collongues et al, 2014). While LETM is related to aquaporin-4 (AQP4)-antibody (Ab) neuromyelitis optica spectrum disorders (NMOSD), it also includes myelin oligodendrocyte glycoprotein (MOG)-Ab associated disease and other unclassified myelitides (Maillart et al, 2020). Furthermore, LETM constitutes the most confounding differential for SDAVF due to their similar imaging characteristics and symptoms involving both upper and lower motor neuron lesions (Jellema et al, 2006; Kister et al, 2016). Both SDAVF and LETM can manifest as long-segment spinal cord lesions, and the clinical manifestations may include lower limb weakness, lower limb sensory abnormalities, and urinary and defaecation disorders. Therefore, the similarity of clinical manifestations and the rarity of both diseases often make it difficult for clinicians to differentiate between the two diseases.

Electromyography (EMG) has been used to monitor neural functions and their improvement during and after surgery in SDAVF patients (Gopalakrishna et al, 2020); however, whether EMG is a potentially effective test in differentiating SDAVF from LETM remains unaddressed. While the EMG characteristics of SDAVF patients have been described in the literature (Linden and Berlit, 1995), the number of patients was small and the data were incomplete, so the EMG characteristics of SDAVF patients could not be fully described. Therefore, the main objective of the current study is to holistically compare and contrast the EMG manifestations of SDAVF and LETM to examine if EMG can be clinically useful in differentiating SDAVF from LETM.

Methods

Subjects

We holistically reviewed the medical records of all patients who were diagnosed with SDAVF via either distal subtraction angiography (DSA) or surgery from 1 January 2010 to 31 December 2020. We extracted and analysed their gender, age, symptom progression (predisposition, sphincter dysfunction, lower limb weakness, superficial sensory impairment, deep sensory impairment), imaging results, and EMG manifestations. We applied the same analysis to patients who were diagnosed with LETM. A total of 38 SDAVF patients and 54 LETM patients were included in our study.

To participate in the study, SDAVF patients must have met the following criteria:

(1) Fistula identified by spinal DSA or surgery.
(2) Available preoperative EMG test results.
(3) Age ≥16 years at onset of disease.

The exclusion criteria for SDAVF patients were as follows:

(1) Any history of endovascular or surgical intervention.
(2) Other spinal cord lesions (spinal stenosis, syringomyelia, among others).

On the other hand, to participate in the study, LETM patients must have met the following criteria:

(1) A single spinal cord lesion spanning no less than 3 vertebral bodies.
(2) Available EMG test results before steroid or immunomodulation therapy.
(3) Age ≥16 years at onset of disease.
(4) AQP4-Ab and MOG-Ab in serum, as tested using a cell-based assay.

The exclusion criteria for LETM patients were as follows:

(1) Myelopathy that was caused by vascular, granulomatosis, paraneoplastic, metabolic, or infectious diseases (Maillart et al, 2020).
(2) Other spinal cord lesions (spinal stenosis, syringomyelia, among others).

Imaging evaluation

MRI: Magnetic resonance imaging results were read by one radiologist and validated by two senior attending neurologists. The extent of T2 hyperintense lesions was characterised

by the number of vertebral bodies involved. For example, a lesion between C3 and C5 was characterised as 3 vertebral bodies.

EMG: Electromyography results were read and validated by two senior attending neurologists.

Statistical analysis

Continuous variables with a normal distribution were analysed by Student's t test. Chi-square test was used to compare classification statistics. Data are presented as n (%). Variables with a normal distribution are expressed as mean ± SD, and in the case of non-normality, the medians are presented. Categorical data are expressed as counts or percentages. The abovementioned data were analysed by SPSS version 24.0 software (IBM Corp, Armonk, NY, USA), and $p < 0.05$ was considered statistically significant. This study was approved by the Institutional Ethics Committee of Chinese PLA General Hospital (approval no. S2023-587-01). Written informed consent was obtained from each participant.

Results

Baseline characteristics

A total of 38 SDAVF patients and 54 LETM patients met our inclusion criteria, and the baseline characteristics are summarised in Tables 1 and 2. The imaging features of typical SDAVF patients are depicted in Figure 1. The imaging features of an NMOSD patient are depicted in Figure 2. The distribution of lesions in the spinal cord of LETM and SDAVF patients is graphically depicted in Figure 3.

Among all SDAVF patients in our study, 29 were male and 9 were female. The mean age of onset was 58.4 ± 8.8. In terms of the location of fistulas, 4 (10.5%) were located above C7, 21 (55.3%) at T1–T12, 8 (21.0%) at L1–L5, and 5 (13.2%) below T12. The disease progression ranged from 0.7–120 months, with a median of 12 months. In most cases (33, 86.8%), SDAVF progressed insidiously, while the remaining cases showed a subacute onset. During disease progression, 52.6% of patients showed various degrees of symptom relapse and remission, while the remaining cases followed a slowly progressive pattern. The detailed information can be found in Table 1.

Among all LETM patients in our study, 8 were male and 46 were female. The mean age of onset was 42.4 ± 14.6. The disease appeared acutely in 44.4% of cases and subacutely in the

Table 1. Baseline demographics and characteristics of patients with spinal dural arteriovenous fistula (*n*=38)

Variable	Median	*n*	%	Range
Sex (female/male)		9/29	23.7%/76.3%	
Age of onset	60			35–75
Duration of symptoms (months)	12			0.7–120
Level of fistula				
above C7		4	10.5%	
T1–T12		21	55.3%	
L1–L5		8	21.0%	
below T12		5	13.2%	
Onset situation (insidious/subacute)		33/5	86.8%/13.2%	
Symptom fluctuation (yes/no)		20/18	52.6%/47.4%	

Table 2. Baseline demographics and characteristics of patients with longitudinally extensive transverse myelitis (*n*=54)

Variable	*n*	Median	Range	%
Sex (female/male)	46/8			85.2%/14.8%
Age of onset		44	16–72	
Related antibody				
AQP-4(+)	41			75.9%
Mog(+)	2			3.7%
negative	11			20.4%
Onset situation (acute/subacute)	24/30			44.4%/55.6%

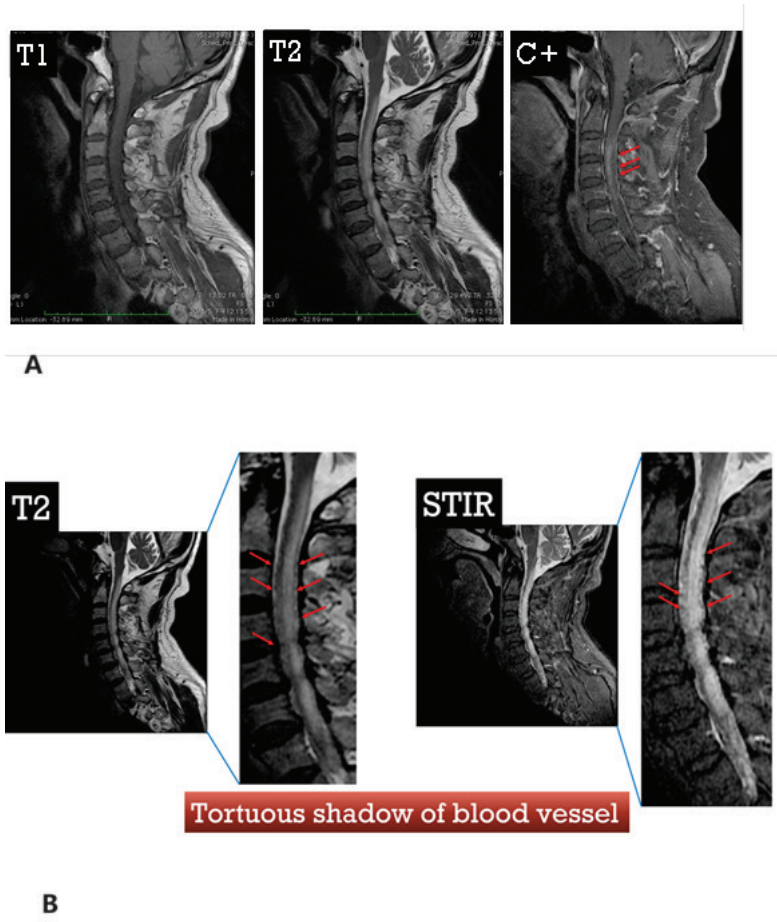

Figure 1. Images of a spinal dural arteriovenous fistula patient. (A) Imaging findings of a spinal dural arteriovenous fistula patient showing long T1 and long T2 signals at the C1-T1 pyramidal level, revealing obvious enhancement (red arrows). (B) Tortuous blood vessels are seen on the dorsal side of the spinal cord at the C2-C5 level (red arrows). In the short TI inversion recovery (STIR) T2 sequence, the fat was suppressed, and the lesion was more pronounced.

remaining cases. Additionally, 41 (75.9%) patients showed AQP4 antibody positivity, while 2 (3.7%) showed Mog antibody positivity. The detailed information can be found in Table 2.

After comparing the SDAVF and LETM groups, we found that LETM tended to appear at a younger age than SDAVF (LETM mean age: 42.4 ± 14.6; SDAVF mean age: 58.4 ± 8.8; $p < 0.001$). Additionally, SDAVF predominantly effected males (76.3%), whereas LETM favourably targeted females (85.2%). Both SDAVF and LETM patients were likely

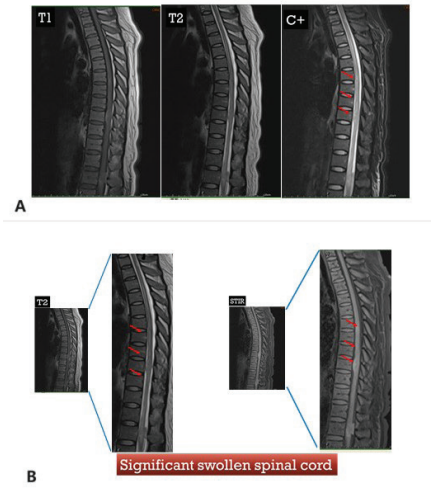

Figure 2. Images of a neuromyelitis optica spectrum disorders patient. (A) Imaging findings of a neuromyelitis optica spectrum disorders patient showing long T1 and long T2 signals of the long segment of the spinal cord, revealing obvious enhancement (red arrows). (B) The lesions are not obvious on T1, but in the short TI inversion recovery (STIR) T1 sequence, the fat was suppressed, and the lesion was more pronounced (red arrows). The entire spinal cord shows significant swelling.

to develop sphincter dysfunction, deep sensation impairment, and superficial sensation impairment (p=0.175, 0.26, 0.473, respectively). There was a statistically significant difference in the presence of influencing factors (p=0.006) and the development of lower limb weakness (p=0.005) between the LETM group and the SDAVF group. After comparing the MRI manifestations of LETM and SDAVF, we found that both conditions presented as longitudinally extensive spinal cord lesions, with no significant differences in lesion length (SDAVF: 7.2 ± 2.3 vertebral bodies; LETM: 7.4 ± 4.2 vertebral bodies; p=0.785). The detailed information can be found in Table 3.

A detailed comparison of EMG between the two groups is shown in Table 4. Regarding the EMG test, we analysed motor and sensory neuron conduction, needle EMG results, and somatosensory evoked potentials. In the LETM group, all 54 patients had needle EMG results, along with motor and sensory conduction results. Only 49 patients had somatosensory evoked potential results. Additionally, 12 patients showed neurogenic damage on the needle EMG tests, and 38 patients showed abnormalities in somatosensory evoked potentials. We found abnormalities in peripheral nerve conduction in 6 patients. On the other hand, in the SDAVF group, all 38 patients had needle EMG results in which 23 patients showed neurogenic damage. Additionally, 36 patients had motor and sensory nerve conduction results, and 26 patients had somatosensory evoked potential results in which 23 patients showed abnormalities. Finally, 7 patients also showed peripheral nerve conduction abnormalities.

Our study revealed that both LETM and SDAVF patients were likely to show EMG abnormalities. Only 9 patients in the LETM group and 4 patients in the SDAVF group presented with normal EMG results. After comparing the two conditions, we found that LETM patients were more susceptible to abnormalities in somatosensory evoked potentials (38/49, 77.6%), whereas SDAVF patients were more susceptible to both abnormalities in somatosensory evoked potentials (23/26, 88.5%) and abnormalities in the needle EMG test (28/38, 73.7%).

We then compared the extent of neurogenic damage, abnormalities in somatosensory evoked potentials, neurogenic damage combined with abnormalities in somatosensory evoked potentials, and peripheral nerve damage. There was a statistically significant difference in the extent of neurogenic damage between the two groups ($p < 0.001$). Both LETM and SDAVF patients were prone to develop abnormalities in somatosensory evoked potentials, although there was no statistically significant difference (p=0.249). However, statistically significant differences were noted in neurogenic damage combined with abnormalities in somatosensory evoked potentials ($p < 0.001$). Both LETM and SDAVF tended not to develop

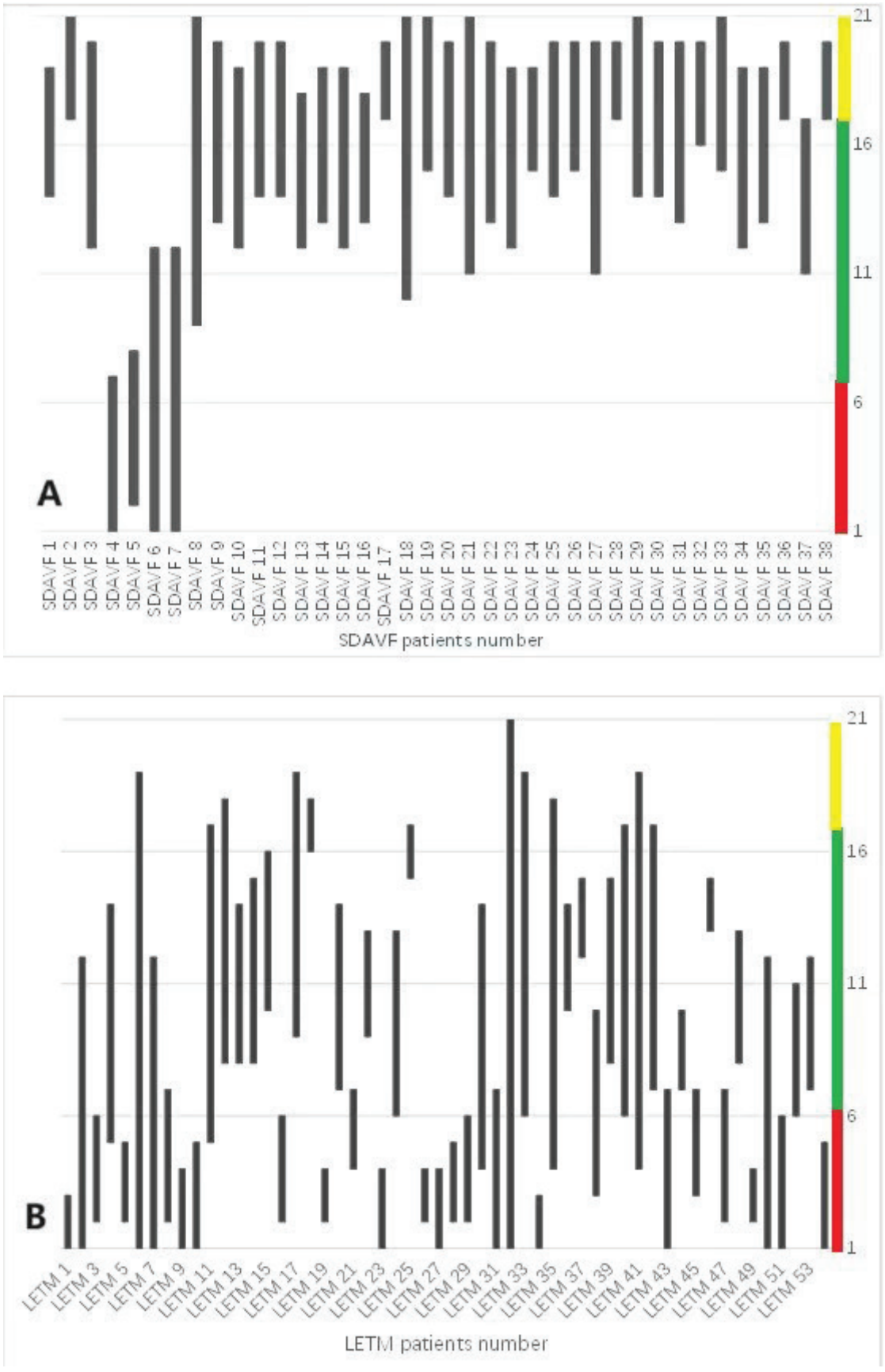

Figure 3. Bar graphs demonstrating lesion distribution along the spinal axis for subjects with confirmed longitudinally extensive transverse myelitis (B) and spinal dural arteriovenous fistula (A). On the vertical axis, red indicates the cervical region, green indicates the thoracic region, and yellow indicates the lumbar region. SDAVF, spinal dural arteriovenous fistula; LETM, longitudinally extensive transverse myelitis.

Table 3. Comparison of clinical magnetic resonance imaging for subjects with spinal dural arteriovenous fistula and longitudinally extensive transverse myelitis

Category	Factor	SDAVF	LETM	*p*-value	*t*/Chi-square
Total subjects		38	54		
Demographics	Age	58.4 ± 8.8	42.4 ± 14.6	<0.001	-6.032
	Sex (female)	23.7% (9)	85.2% (46)	<0.001	35.088
MRI location	Vertebral body span	7.2 ± 2.3	7.4 ± 4.2	0.785	0.274
Clinical symptom	Predisposition	5.3% (2)	27.8% (15)	0.006	7.506
	Sphincter dysfunction	76.3% (29)	63% (34)	0.175	1.842
	Lower limb weakness	100% (38)	81.5% (44)	0.005	7.895
	Superficial sensory impairment	81.6% (31)	87% (47)	0.473	0.515
	Deep sensory impairment	73.7% (28)	83.3% (45)	0.26	1.267

MRI, magnetic resonance imaging; SDAVF, spinal dural arteriovenous fistula; LETM, longitudinally extensive transverse myelitis.

Table 4. Comparison of electromyography features with spinal dural arteriovenous fistula or longitudinally extensive transverse myelitis

Category	Factor	SDAVF	LETM	*p*-value	Chi-square
Total subjects		38	54		
EMG	Peripheral nerve damage	19.4% (7/36)	11.1% (6/54)	0.271	1.214
	Somatosensory evoked responses	88.5% (23/26)	77.6% (38/49)	0.249	1.332
	Neurogenic damage	73.7% (28/38)	22.2% (12/54)	<0.001	24.037
	Neurogenic damage with abnormal somatosensory evoked responses	65.4% (17/26)	14.3% (7/49)	<0.001	20.383
	Only abnormal somatosensory evoked responses	21.4% (6/28)	62.0% (31/50)	<0.001	11.849
	Completely normal	10.5% (4/38)	16.7% (9/54)	0.405	0.693

Note: In the SDAVF group, 2 patients were missing data of peripheral nerve conduction, and 12 patients were missing data of somatosensory evoked potentials. In the LETM group, 5 patients were missing data of somatosensory evoked potentials. EMG, electromyography.

peripheral nerve damage (p=0.271). Finally, completely normal EMG test results were rare in both conditions, with no statistically significant differences in both groups (p=0.405).

Finally, we analysed and summarised the typical EMG manifestations of SDAVF. Our results indicated that SDAVF presented with normal nerve conduction velocity but abnormal needle EMG and somatosensory evoked potential test results. The abnormalities of needle EMG can distinguish SDAVF patients from LETM patients. Tables 5–8 show the characteristic EMGs of an SDAVF patient and an LETM patient.

Discussion

Our study summarised the characteristics of SDAVF and LETM, comparing the demographics of patients, symptomology, MRI manifestations, and EMG test results. In this observational

Table 5. Needle electromyography studies of a spinal dural arteriovenous fistula patient

	Spontaneous	Activity		Motor	Unit	Potential (1)	Motor	Unit potential (2)
	IA	Fib	PSW	Dur	PPP	Amp	WF	Peak voltage
Rt TAM	N	N	N	16.8	33%	3088	SP	4.1
Lt TAM	N	N	N	17.6	56%	2387	SP	5.0
Rt QDF	N	N	N	11.1	17%	495	-	-
Lt QDF	N	N	N	12.0	-	549	SP	1.8
Rt GCM	N	2+	2+	19.0	-	1575	-	-
Lt GCM	N	2+	2+	16.3	20%	4009	SP	5.0
AS	N	N	N	11.4	31%	472	MP	1.1

The patient's needle electromyography showed neurogenic damage to both lower limbs. IA, insertional activity; Fib, fibrillation; PSW, positive sharp wave; Dur, duration; PPP, polyphasic pattern; Amp, amplitude; WF, wave form; Rt, right; Lt, left; TAM, tibialis anterior muscle; QDF, quadriceps femoris; GCM, gastrocnemius muscle; N, normal; AS, anal sphincter; SP, simplex pattern; MP, mixed pattern.

Table 6. Somatosensory evoked studies of a spinal dural arteriovenous fistula patient

Upper limb									
Nerve	**Stimulation site**	**Latency**		**(ms)**		**Amplitude**		**(μV)**	
Name		N9		N20		N9		N20	
Lt Median	Wrist	9.9		19.6		4.4		3.6	
Rt Median	Wrist	9.9		19.6		4.6		3.3	
Lower limb									
Nerve	Stimulation site	Latency		(ms)		Amplitude		(μV)	
Name		P1	N1	P2	N2	P1	N1	P2	N2
Lt Tibial	Ankle	34.2	44.0	57.7	69.5	1.4	4.4	6.2	9.5
Rt Tibial	Ankle	34.2	43.0	55.0	68.7	2.3	11.0	12.7	10.7

The patient's electromyography showed no obvious abnormalities. Rt, right; Lt, left; P, positive; N, negative.

Table 7. Needle electromyography studies of a longitudinally extensive transverse myelitis patient

	Spontaneous		activity	Motor	Unit	Potential (1)	Motor	Unit potential (2)
	IA	Fib	PSW	Dur (ms)	PPP	Amp (μV)	WF	Peak voltage (mV)
Rt FDI	N	N	N	9.6	9%	403	-	3.3
Rt ED	N	N	N	9.8	6%	428	-	1.3
Rt BBM	N	N	N	10.6	5%	561	MP	2.1
Lt TAM	N	N	N	11.8	20%	566	-	2.7
Lt QDF	N	N	N	10.2	-	408	SP	1.3
Rt QDF	N	N	N	10.4	20%	558	SP	1.6

The patient's needle electromyography showed no obvious abnormalities. IA, insertional activity; Fib, fibrillation; PSW, positive sharp wave; Dur, duration; PPP, polyphasic pattern; Amp, amplitude; WF, wave form; Rt, right; Lt, left; FDI, first dorsal interosseous; ED, extensor digitorum; BBM, biceps brachii muscle, TAM, tibialis anterior muscle; QDF, quadriceps femoris; N, normal; SP, simplex pattern; MP, mixed pattern.

Table 8. Somatosensory evoked studies of a longitudinally extensive transverse myelitis patient

Upper limb									
Nerve	Stimulation site	Latency		(ms)		Amplitude		(μV)	
Name		N9		N20		N9		N20	
Lt Median	Wrist	9.9		19.8		1.6		5.6	
Rt Median	Wrist	9.2		18.8		2.8		8.3	
Lower limb									
Nerve	Stimulation site	Latency		(ms)		Amplitude		(μV)	
Name		P1	N1	P2	N2	P1	N1	P2	N2
Lt Tibial	Ankle	44.0	52.2	69.2	78.7	2.6	2.7	0.9	0.8
Rt Tibial	Ankle	44.5	54.2	70.7	80.5	1.8	2.8	2.3	1.6

The patient's electromyography showed that the P40 latency of the bilateral tibial nerves was prolonged. Rt, right; Lt, left.

study, we found that SDAVF and LETM predominantly affect patients of different age and sex. The demographic distribution is consistent with the existing literature (Jellema et al, 2006; Wingerchuk et al, 2015). While our study shows no significant difference in the length of spinal cord lesions between LETM and SDAVF, the location of lesions tended to be different. SDAVF lesions manifested on MRI are more likely to be found on the lower thoracic and lumbar segments, whereas lesions of LETM tended to appear on the upper thoracic and cervical segments. Such a difference in the location of lesions has been documented elsewhere (Jellema et al, 2006; Wingerchuk et al, 2015; Kister et al, 2016). In SDAVF, the location of fistulas primarily occurs at thoracic and lumbar segments, which often corresponds with the MRI findings.

Regarding the symptomology, there was a statistically significant difference in the rate of myasthenia of lower extremities and the existence of inducing factors. We found that all SDAVF patients showed myasthenia of lower extremities, while 81.5% of all LETM patients also showed the same symptom. Therefore, although statistical significance is evident, we believe that myasthenia of lower extremities is not an effective differentiating factor due to its high occurrence rate in both groups. On the other hand, 27.8% of LETM patients demonstrated influencing factors before disease onset, such as infection and diarrhoea, whereas only 5% of SDAVF patients developed the disease after heavy lifting. Thus, while the existence of influencing factors can be used as a differentiating factor, it is not a major one due to its relative low occurrence rate in both conditions. Finally, our study compared the EMG results of LETM and SDAVF patients. SDAVF patients were more prone to neurogenic damage. Both SDAVF and LETM presented as abnormalities in somatosensory evoked potentials, while peripheral nerve damage was rare in both conditions. On the other hand, SDAVF typically presented as neurogenic damage combined with posterior funiculus injury, whereas only posterior funiculus injury was usually seen in LETM. Additionally, evidence of lower motor neuron lesions was unique to SDAVF, while posterior funiculus injury was often seen in both SDAVF and LETM. Therefore, given the unique EMG characteristics of SDAVF, we believe that needle EMG is a useful diagnostic test in differentiating LETM from SDAVF.

With the development and popularisation of angiography, our understanding of the pathophysiology of SDAVF is increasing. Rather than vascular steal phenomenon, spinal compression, or spinal cord bleeding, venous hypertension is believed to be the primary pathophysiology of SDAVF (Aminoff et al, 1974). While the development of contrast-enhanced magnetic resonance angiography has contributed to our understanding of SDAVF, the golden diagnostic tool for SDAVF remains DSA (Jellema et al, 2006). In our study, we found that the primary symptoms of SDAVF are sensory deficits, motor defects, gait disturbances, and urinary and stool retention. We also found that SDAVF predominantly affects middle-aged males with the fistula occurring primarily at thoracic-lumbar segments. These findings are consistent with the existing literature (Jellema et al, 2006; Iovtchev et al., 2015; Hunt et al, 2018).

Due to the versatility and unpredictability of the initial symptoms of SDAVF, misdiagnosis often occurs. SDAVF presenting as long segmental myelopathy is often misdiagnosed as LETM, leading to unnecessary treatment before surgical treatment of SDAVF. LETM is a central nervous system demyelinating disease (Lucchinetti et al, 2014; Wingerchuk et al, 2015; Lopez et al, 2021). In its acute phase, LETM usually presents as spinal cord oedema, along with continuous or discontinuous hyperintense lesions in the spinal cord. In our study, we found that the main symptoms of LETM include sensory deficits, urinary and faecal retention, and muscle weakness, which is consistent with the existing literature (Wingerchuk et al, 2015; Maillart et al, 2020). It should be noted that most of our LETM patients had a diagnosis of NMOSD. Histologically, NMOSD often manifests as macrophage infiltration on oligodendrocytes and astrocytes, axonal death, astrocyte damage, and vascular granulocyte infiltration. In our study, three NMOSD patients showed reduced conduction velocity of peripheral nerves. Such an observation raises an intriguing question as to whether NMOSD, which is often believed to affect CNS exclusively, can potentially lead to peripheral neuropathy. The study of lower motor neuron damage in NMOSD patients is limited. In a case report, a patient who was subsequently diagnosed with NMOSD had been misdiagnosed with lumbosacral nerve root disorder (Kim et al, 2016); however, due to the existence of the GD1b antibody in this patient, it is unclear whether the damage of lumbosacral nerves is a manifestation of NMOSD or a symptom of other clinical conditions. Regarding the EMG results, 12 patients in the NMOSD group showed needle EMG test results indicative of neurogenic damage. However, a detailed review of the medical history of these 12 patients revealed that 4 of them had a history of connective tissue diseases, while 2 of them had a history of shingles. Additionally, it has been shown that autoimmune diseases that can cause peripheral neuropathy, such as systematic lupus erythematosus, Sjogren's syndrome, and myasthenia gravis, are common comorbidities of NMOSD (Kamm and Zettl, 2012; Lucchinetti et al., 2014; Lopez et al,

2021). Therefore, our study shows that, while EMG is a viable test for the differential diagnosis of NMOSD, interpreting results in the context of the patient's past medical history and symptoms is especially important in determining the clinical significance of the EMG test results in diagnosing NMOSD.

Spinal dural arteriovenous fistulas are abnormal shunts between the root medullary artery and the medullary vein (Krings and Geibprasert, 2009), classically located below the pedicle of the vertebral arch and within the dura mater. The most common location for shunting includes the dorsal dura root sleeve between the two adjacent nerve roots that lie between the interlingual foramen and the dura (Da Ros et al, 2021). Shunting leads to an increase in venous pressure in the medullary vein, leading to 'arterialization' of the medullary vein, which ultimately leads to spinal cord oedema, spinal artery hypoperfusion, and progressive hypoxia. Because venous hypertension is a major pathophysiologic feature of SDAVF, patients frequently experience worsening symptoms during prolonged standing, activity, or Valsalva exercise (Vuong et al, 2016), all of which increase venous system stress. Furthermore, because the dura mater branches and the radiculomedullary arteries supply the nerve roots, the development of lower motor neuron lesions in our SDAVF patients can be explained.

Of all SDAVF patients in our study, 7 showed peripheral nerve damage in the EMG test; however, such peripheral neuropathy can be explained by a history of chronic diabetes mellitus in three patients. One patient had unilateral damage of the median nerve possibly due to compression of abnormally developed bone. However, we could not find alternative explanations for the peripheral nerve damage of the other 3 patients; therefore, we attribute it to the nerve root damage that potentially caused by SDAVF. Ten SDAVF patients in our study showed no signs of lower motor neuron lesions in the EMG test. Of these 10 patients, 7 patients presented with fistulas at mid- and low-thoracic levels, two above the C3 level, and one at lumber level. The tendency for fistulas to occur at mid- and lower-thoracic levels can be explained by the vascular organisation at these segments. The descending aorta supplies the majority of arteries at the level of T3–L4, whereas T1 and T2 levels and cervical segments are supplied by the intercoastal artery and the carotid and vertebral arteries, respectively (Vuong et al, 2016). Damage to the nerve roots adjacent to the fistula can explain the development of nerve root pain in our patients. Due to the progressive nature of SDAVF, the longer the disease duration, the more likely the appearance of neurogenic damage.

Additionally, we found evidence of lower motor neuron lesion in a patient who had the shortest disease progression (20 days); therefore, a possible explanation for normal needle EMG results in SDAVF patients is that EMG on paravertebral muscle is not performed. Finally, it has been reported that neurogenic damage caused by SDAVF was misdiagnosed as ALS (Santos et al, 2021). Thus, even though a characteristic EMG pattern of SDAVF was evident, misdiagnosis still happens due to the physician's lack of awareness on the potential value of EMG in diagnosing SDAVF. This further emphasises the need for our holistic review.

There are several limitations in our current study. Firstly, our study is a retrospective observational study that establishes an association between EMG abnormalities and SDAVF diagnosis. EMG by itself can neither replace DSA nor indicate fistula location. A combination of symptomology, imaging, and EMG is needed to rule out other differentials such as LETM. Secondly, while our study holistically compares the EMG manifestations of LETM and SDAVF, as LETM constitutes the main differential for SDAVF, we do not do so for other differentials of SDAVF. Future studies will need to include EMG analysis of more diseases before determining the strength of EMG in diagnosing SDAVF. Thirdly, we have not evaluated the potential changes in EMG between pre- and post-surgery SDAVF patients. Such an approach is valuable as EMG can be used to monitor nerve conductions during SDAVF surgery and to measure neurological improvement post-surgery (Gopalakrishna et al, 2020). There are also studies that monitor the electromyographic changes of spinal cord arteriovenous and intramedullary spinal cord cavernous malformations during surgery (Li et al, 2019). Finally, it has been reported that some characteristics of MRI have predictive value for the prognosis of SDAVF patients (Pu et al, 2017). In the future, a larger sample of patients will need to be followed to improve the comparison of EMG before and after

rehabilitation, and to observe whether the changes in EMG can indicate the prognosis of SDAVF patients.

Conclusions

Spinal dural arteriovenous fistula is a rare disease. We systematically reviewed 38 SDAVF cases and 54 LETM cases and revealed their differences in disease demographics, symptomology, disease progression, and imaging results. Our study shows that SDAVF predominantly affects male, whereas LETM favourably targets females, but both SDAVF and LETM patients are likely to develop sphincter muscle dysfunction, deep sensation loss, and superficial sensation loss. After comparing the MRI manifestations of LETM and SDAVF, we found that both conditions present as longitudinally extensive spinal cord lesions with no significant differences in lesion length, but there were differences in the areas where the lesions were concentrated. The lesions of patients with SDAVF were concentrated in the thoracolumbar segment, while those of the patients with LETM were concentrated in the cervicothoracic segment. However, due to the variability of the initial symptoms, it is difficult for clinicians to distinguish between the two diseases during the initial clinical visit.

Although DSA is the gold standard for the diagnosis of SDAVF, its invasiveness often causes physicians to be hesitant in performing the test unless robust clinical evidence points to a possible diagnosis of SDAVF. Therefore, it is important to explore other non-invasive yet sensitive clinical tests to diagnose SDAVF. Given that both SDAVF and LETM patients commonly present with symptoms of lower motor neuron lesions, EMG is usually ordered by the physician. Our study shows that EMG is potentially useful in aiding the differentiation of LETM and SDAVF when used in combination with symptomatology and imaging. Our study reveals that both LETM and SDAVF patients are likely to show EMG abnormalities, and the abnormalities of needle EMG can distinguish SDAVF patients from LETM patients. If clinicians can combine the clinical manifestations, imaging findings and EMG features of patients, it will greatly improve the diagnostic accuracy of SDAVF.

Key points

- Our study systematically compares the demographics, symptomatology, and disease progression of SDAVF and LETM.
- These findings can help the physician to empirically differentiate SDAVF from LETM.
- Our study compares the MRI characteristics of lesions of SDAVF and LETM. Lesions of SDAVF tend to appear at lower lumbar and sacral segments, whereas lesions of LETM more frequently appear at cervical and upper thoracic segments.
- Our study is the first to date to explore the diagnostic value of EMG in differentiating SDAVF from LETM. Evidence of neurogenic damage is a key characteristic of SDAVF that is often missing in LETM.
- Therefore, our findings suggest the clinical implication of EMG in the diagnosis of SDAVF.

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Availability of data and materials

The raw data and materials supporting the conclusions of this article will be made available by the authors, without undue reservation.

Author contributions

DHH and WPG designed the programme. JZ, YL, SYX and MHL contributed to the data collection and discussed the results. JZ drafted the manuscript. All authors contributed to important editorial changes in the manuscript. All authors read and approved the final manuscript. All authors have participated sufficiently in the work and agreed to be accountable for all aspects of the work.

Ethics approval and consent to participate

This study was approved by the Institutional Ethics Committee of Chinese PLA General Hospital (approval no. S2023-587-01). Written informed consent was obtained from each participant.

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Conflict of interest

The authors declared that they have no conflicts of interest in this work.

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