

Lhermitte's sign in multiple sclerosis: a clinical survey and review of the literature

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SUMMARY Lhermitte's sign was experienced by 33.3% of 114 patients with multiple sclerosis, and in 16% it occurred in the first episode. Eleven patients with subacute combined degeneration of the cord due to pernicious anaemia also were reviewed, and one reported Lhermitte's sign. Review of the literature suggests that the symptom rarely occurs in other conditions except subacute combined degeneration of the cord, neck trauma, radiation myelitis and prolapsed cervical disc.

Lhermitte's sign is described in most standard neurology textbooks.¹⁻³ All authorities agree that it is most common in multiple sclerosis but the prevalence of the symptom in this disease is unknown. We have tried to determine its prevalence in an unselected group of patients with multiple sclerosis and reviewed the literature on the other causes commonly mentioned in textbooks.

McAlpine⁴ appears to have been the first to use the term Lhermitte's sign in print although, in modern medical usage, the phenomenon would be called a symptom, "the consciousness of a disturbance in a bodily function".⁵ We have used the term since it is widely accepted and conveniently indicates the sensation with its precipitation by movement of the spine and characteristic radiation.

HISTORICAL REVIEW

The first description of the symptom was given by Marie and Chatelin in 1917⁶ . . . "le blessé a la sensation qu'un courant électrique parcourt ses bras et même ses jambes . . . lorsque le blessé baisse fortement la tête sur la poitrine" . . . Their patient had been shot in the head. The following year, Babinski and Dubois⁷ described a similar symptom in four soldiers of whom one had been stabbed in the neck. . . . "chaque fois qu'il fléchit la tête, qu'il étternue ou qu'il touse, une sensation de décharge électrique, partant de la nuque et s'irradiant très

violente le long de bras et du membre inférieur droits". Thus, electric shock-like sensations radiating down the body and limbs were first recorded. Ribeton⁸ described 12 cases, all following head or neck trauma. He thought that the lesion responsible was spinal, even in the head injured cases, and must involve the posterior columns because of its distribution in traumatic Brown-Sequard lesions.

The first reference to the symptom in multiple sclerosis was by Beriel and Devic in 1918.⁹ Lhermitte, Bollak and Nicholas¹⁰ described a woman with multiple sclerosis who experienced the symptom in her first attack. They commented on the similarity of the symptom to Tinel's¹¹ observation of tingling sensations on percussion or stretching of partly-damaged peripheral nerves. Tinel found demyelination with preserved axons in such nerves, and they referred to experimental work by Lhermitte, Claude and Roussy on "concussion" of the spinal cord which had produced demyelination.

Patrick,¹² while reporting a patient with multiple sclerosis, claimed that Lhermitte's symptom occurred in tumours of the cord and Pott's disease of the spine, but gave no details. This was followed in 1933 by two reports^{13 14} of the symptom occurring in subacute combined degeneration of the cord. In 1944 Walshe¹⁵ drew attention to the frequent occurrence of symptoms and signs of cervical cord damage in patients who had suffered violent neck flexion at the time of a head injury. He noted that these patients were often accused of malingering because of sensory complaints, including Lhermitte's sign, which were out of proportion to the severity of the head injury and apparently unrelated to it. He noted the interval

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of a few weeks between the injury and the appearance of symptoms which Ribeton had described.

The occurrence of electric shock-like sensations in patients who had received radiation to the cervical cord was first reported by Boden in 1948.¹⁶ He described the symptom both as a transient manifestation and as a prelude to permanent myelopathy and noted a latent interval following radiation before the symptom appeared.

A paper critical of Lhermitte's conclusions by Alajouanine, Thurel and Papaïoanou¹⁷ appeared in 1949. Without reference to the cases of Walshe, Boden, Olkon or Hassin, they reported six patients with the "electric symptom" who did not have multiple sclerosis (two with atlanto-axial subluxation, two with tumours, one with a prolapsed cervical disc and one with tuberculous arachnoiditis). Their main disagreement with Lhermitte was his insistence on demyelination as the most important feature. Their cases had only "mechanical irritation" of the posterior columns and they felt an extrinsic cause should be excluded before demyelination was assumed. They also stated that poking the exposed posterior columns during operation under local anaesthetic produced a similar symptom . . . "l'excitation punctiforme du cordon postérieur détermine une douleur en éclair, localisée en une zone circonscrite de la partie sous-jacente homolaterale du corps".

A definitive account of Lhermitte's sign in radiation myelopathy was published in 1964.¹⁸ Jones reviewed the literature and discussed the mechanism of the sign in detail. He pointed out that mechanical factors alone could not be responsible as Alajouanine had suggested but a medullary lesion must be present, since neck flexion, which triggered each sensation, was operating during the latent period after trauma and radiation as well as when the symptom appeared. In 1973 Gautier-Smith¹⁹ reported that 11 of 44 patients with subacute combined degeneration of the cord had described Lhermitte's sign.

Method

All patients admitted to hospital or attending the out-patient clinic with a clinical diagnosis of multiple sclerosis were studied. In addition, discharged patients listed in the departmental diagnostic index as having multiple sclerosis or subacute combined degeneration of the cord were recalled to the clinic.

On review of the history, clinical examination and laboratory tests, the patients were classified as definite, probable or possible multiple sclerosis by the criteria of Rose *et al*,²⁰ and subacute combined degeneration of the cord on the basis of clinical features and the presence of vitamin B12 malabsorption or anti-intrinsic factor antibodies.

Patients considered eligible for study were asked, without using leading questions but with prompting where necessary, about the occurrence of unusual sensations. Lhermitte's sign was considered to have occurred if electric shock, tingling, rippling or other feelings had travelled rapidly along the spine. If the symptom had occurred, details of its nature and distribution, precipitating and relieving factors and the association of its onset with the onset or exacerbation of multiple sclerosis or onset of subacute combined degeneration of the cord were recorded.

Results

One hundred and fourteen patients with multiple sclerosis were studied. There were 36 men and 78 women. At the time of interview their ages ranged from 17-71 years, mean 43.7. Thirty-eight reported the occurrence of the symptom at some time and the details are given in tables 1 and 2.

In 19 patients the symptom occurred during the first attack of multiple sclerosis and in two patients it was the very first symptom. It tended to disappear over a period of 4-6 weeks. A few patients continue to experience the symptom from time to time, even years after the first attack. This often happened at the time of a relapse, even when the new lesion was clearly in some other part of the nervous system. Thirty patients spontaneously used the term "electric shock" and the descriptions of the others are listed in table 3. The radiation of the sensation is given in table 4 and precipitating movements in table 5.

Table 1 *Prevalence of Lhermitte's sign in multiple sclerosis*

	No	Lhermitte's sign present
Definite	69	26 (37.6%)
Probable	25	7 (28%)
Possible	20	5 (25%)
Total	114	38 (33.3%)

Table 2 *Age at time of occurrence of Lhermitte's sign in multiple sclerosis (Range 10-62 years. Mean 35.7 years)*

Under 20	21-30	31-40	41-50	51 and over
3	10	12	11	2

Table 3 *Description of sensation in multiple sclerosis*

"Electric Shocks"	30
"Pain"	2
"Tingling, pins and needles"	2
"Ripple or shiver"	2
"Hair standing on end"	1
"Fizzing"	1

Table 4 *Pattern of spread in multiple sclerosis*

Down spine only	11	Up and down spine	7
+ both legs	7	Down unilateral arm and leg	1
+ arms and legs	4	Variable	5
+ arms only	3		

Table 5 *Movements precipitating the sign in multiple sclerosis*

Neck flexion	28
Limb movements	3
Bending down	3
Coughing/sneezing	1
Laughing	1
More than one movement	4
Spontaneous	2

Patients reported that each paroxysm lasted for a very short time. Some were able to count aloud for us while experiencing the sensation and it usually took less than two seconds to travel the length of the body. In two cases a typical sensation occurred without a precipitating movement, and no patient reported persistence of any abnormal sensation after the first shock. Eight patients had noticed that after five or six neck movements the shock no longer occurred. None of the patients experiencing the symptom at the time of interview asked for treatment to relieve it, though all commented that it was unpleasant.

Twelve of the patients who reported Lhermitte's sign had never disclosed their experience to a doctor before we interviewed them.

Eleven patients with subacute combined degeneration of the cord were studied; one had suffered Lhermitte's sign in the first few weeks of his illness. He was 76 years old and presented with paraes-

thesia in the feet and ataxia due to loss of position sense. Lhermitte's sign disappeared when B12 therapy was given. Another patient with subacute combined degeneration of the cord complained of increased tingling in her feet whenever she bent her head forward, but no sensation travelled along her body. All these patients were over 58 years and the oldest was 77 years of age.

Discussion

The prevalence of Lhermitte's sign in a group of patients with multiple sclerosis attending a general hospital neurological clinic has been determined. We observed a striking uniformity in the description of the symptom, both in patients who had described it to doctors before and in those reporting it for the first time.

Similar terms were used by patients 50 years ago. Why "electric shock" is used when so few people have ever experienced one is a puzzle; perhaps the rapidity with which the sensation travels and its brevity influence the description. A higher proportion of patients with definite multiple sclerosis reported the symptom, suggesting that some of the probable and possible cases have still to experience it.

The prevalence of the symptom has been determined in radiation myelopathy¹⁶ and subacute combined degeneration of the cord¹⁹ but not in any other condition. A review of standard textbooks and the literature on the symptom produced a number of anecdotal reports and a few reviews of larger numbers of patients which have been listed in table 6. From this it can be seen that subacute combined degeneration, trauma, radiation and disc prolapse

Table 6 *Other reported causes of Lhermitte's sign*

Condition	2	—	
Condition	No with signs	No studied	Authors
Atlanto-axial subluxation	2	—	Alajouanine <i>et al</i> ¹⁷
Fractured odontoid	1	6	Greenberg ²¹
Prolapsed cervical disc	1	—	Alajouanine <i>et al</i> ¹⁷
	3	8	O'Connell ²²
Cervical spondylosis	1	55	Brooker <i>et al</i> ²³
TB Arachnoiditis	1	—	Alajouanine <i>et al</i> ¹⁷
Spinal tumour	2	—	Alajouanine <i>et al</i> ¹⁷
Subacute combined degeneration of the cord	1	—	Hassin ¹⁸
	1	—	Olkon ¹⁴
	11	44	Gautier-Smith ¹⁰
Radiation myelopathy	6	145 (transient)	Boden ¹⁶
	4	16 (progressive)	
	1	7 (progressive)	
	7	—	Palmer ²⁴
Syringomyelia	1	100	Jones ¹⁸
	(post-traumatic)		Barnett <i>et al</i> ²⁵
Trauma	2	—	Marie and Chatelin ⁶
	4	—	Babinski and Dubois ⁷
	4	—	Walsh ¹⁵
	30	—	Rieder (US Army) ²⁶

are the other conditions in which the symptom has most often been reported. In our small series of patients with subacute combined degeneration we did not find so high a proportion experiencing Lhermitte's sign as did Gautier-Smith.

Jones¹⁸ states that Lhermitte's sign is common in cervical spondylosis, but gives no details; it is not mentioned in the reviews by Pallis *et al*²⁷ or Brain *et al*,²⁸ which report in detail on a total of 95 patients with cervical spondylosis. Brooker and Barter²³ describe one patient with Lhermitte's sign as a result of spondylosis. Both Bickerstaff² and Northfield²⁹ state that Lhermitte's sign can occur in tonsillar ectopia but it is not recorded by Appleby *et al*³⁰ in their review of 17 patients with Arnold-Chiari malformations or by Barnett *et al*²⁵ in their patients with tonsillar ectopia and syringomyelia.

Lhermitte¹⁰ suggested that sensory fibres stripped of their myelin sheaths were unduly excitable to percussion or elongation. Smith and McDonald³¹ examined sensory fibres traversing demyelinated lesions made by the microinjection of lysophosphatidyl-choline in cat spinal cord. They found spontaneously active fibres and on deforming the dorsal columns with a glass rod observed an increase in the firing rate. Fibres were most mechanically sensitive in the demyelinated region and deformations of less than 1 mm were effective. They demonstrated that the activity arose within the lesion. Whether these action potentials arose from demyelinated intact axons or by ephaptic spread from severed axons could not be determined.

Severed fibres have been described in multiple sclerosis lesions³² and also in subluxation injury of the cord.³³ However, myelin damage preceding axonal disruption has been reported in experimental radiation myelopathy³⁴ and Wallerian degeneration³⁵ in rat spinal cord as well as in experimentally induced vitamin B12 deficiency in Rhesus monkeys.³⁶ In both experimental radiation myelopathy and Wallerian degeneration in central fibres the earliest changes found are widening of the nodes of Ranvier and para-nodal myelin breakdown. Godwin-Austen *et al*³⁷ found myelin damage with apparently normal axons in three patients who died with radiation myelopathy.

Agamanolis *et al*³⁸ produced B12 deficiency in Rhesus monkeys and showed that the histology and localisation of lesions in the monkeys were indistinguishable from subacute combined degeneration in humans. Electron microscopy of early spinal lesions³⁶ revealed distension of myelin sheaths, with separation and stripping of myelin lamellae leaving denuded but intact axons surrounded by astrocytic processes and a cystic honeycomb appearance in the myelin sheath. Later total axonal degeneration was

found in lesions. In a detailed study of the myelopathy of pernicious anaemia, Pant *et al*³⁹ found distension of myelin sheaths and felt that the myelin was more severely involved than the axons in the early lesions; but they found no case which did not have distal tract degeneration and they concluded that there was little if any focal demyelination with preservation of axons. However, they did not study the ultrastructure of the cord and their material was from patients dying from subacute combined degeneration whereas Agamanolis *et al* examined material obtained after various periods of B12 deficiency.

These early changes in the myelin sheaths in Wallerian degeneration and radiation may explain the occurrence of sensory symptoms in conditions in which axonal degeneration eventually predominates. If the myelin changes found in Rhesus monkeys with B12 deficiency are assumed to occur in humans with subacute combined degeneration, then the fact that Lhermitte's sign is so much more common in multiple sclerosis and subacute combined degeneration suggests that demyelination with axonal preservation is more important for the generation of spontaneous activity than the presence of severed axons. The spread of the sensation noted by patients could indicate ephaptic activation of adjacent demyelinated axons in the posterior columns.

The spine is most mobile in the neck. During flexion the cervical cord changes in length by several centimetres and the denticulate ligaments tighten.⁴⁰ This produces much more deformation of the cord than Smith and McDonald used in their experiments.

Oppenheimer⁴⁰ examined the spinal cords of 14 patients with multiple sclerosis and noted that plaques were twice as common in the cervical region as in the rest of the cord. The plaques mainly occurred in the dorsal columns in relation to the point of attachment of the denticulate ligaments. He felt that this mechanical stress was in some way responsible for the more frequent occurrence of plaques at that point and for their shape. For this reason, Oppenheimer suggested that neck flexion has a detrimental effect in patients with multiple sclerosis, especially those with Lhermitte's sign.

In conclusion, Lhermitte's sign does occur in other conditions but it is so uncommon in conditions other than multiple sclerosis and subacute combined degeneration that its development in a young person, with no history of trauma to the neck or radiation, is of great diagnostic importance. Ekbom⁴¹ has reported that carbamazepine relieves the symptom, but only occasionally is it intolerable enough to warrant therapy.

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