

Lhermitte's Sign: The Current Status

Supreet Khare, Deeksha Seth¹

Armed Forces Medical College, Pune, ¹Kasturba Medical College, Mangalore (Manipal University), India

Abstract

Lhermitte's sign was described by Marie and Chatelin and named after Jean Lhermitte. This sign is mostly described as an electric shock like condition by some patients of multiple sclerosis. This sensation occurs when the neck is moved in a wrong way or rather flexed. It can also travel down to the spine, arms, and legs, and sometimes the trunk. Demyelination and hyperexcitability are the main pathophysiological reasons depicted for the Lhermitte's sign. Other causes for Lhermitte's sign include transverse myelitis, behçet's disease, trauma, etc. This article reviews the Lhermitte's sign, its history, and its etiopathophysiology. Very few studies are available on Lhermitte's sign and more research need to be done on the same to ensure its sensitivity and specificity.

Key Words

Demyelination, Lhermitte's sign, shock like sensation

For correspondence:

Dr. Supreet Khare, Virat Khand, Gomtinagar, Lucknow - 226 010, Uttar Pradesh, India.

E-mail: drsupreet.khare@gmail.com

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Introduction

Lhermitte's sign (also known as Lhermitte's phenomenon also referred to as the barber chair phenomenon is the name which describes an electric shock-like sensation that occurs on flexion of the neck. This sensation radiates down the spine, often into the legs, arms, and sometimes to the trunk. Lhermitte's sign was first described by Marie and Chatelin in 1917. The first reference to the symptom was described by Beriel and Devic in 1918 in multiple sclerosis (MS). In 1924, Lhermitte *et al.* described in detail a patient with MS and electric dysesthesias.^[1] Recently, Lhermitte's sign has been associated with the intensity-modulated radiotherapy (IMRT) for head and neck cancer patients. It is one of the late term effects following IMRT in these patients.^[2-4]

Pathophysiology and eliciting the sign

Lhermitte's sign is caused by miscommunication between the nerves that have become demyelinated. The pathophysiology of Lhermitte's sign was described as the stretching of the hyper excitable demyelinated dorsal column of

the spinal cord, particularly at the cervical level, thus triggering an electric shock-like sensation. Till date, hyper excitability is considered as the main pathophysiological mechanism for the occurrence of Lhermitte's sign. Also, from an etiological point of view, in the original paper by Lhermitte, the shock-like sensations were supposed to be caused by medullary lesions due to demyelination or trauma of the dorsal column.^[5] Neck movements are said to exacerbate Lhermitte's sign where Lhermitte's phenomenon is said to be induced by neck flexion while the reverse Lhermitte's phenomena is defined when symptoms are induced by neck extension. Reverse Lhermitte's phenomenon is said to be induced by extrinsic compression of the cervical cord and neck collar immobilization.^[6] A relatively rare form called inverse Lhermitte phenomenon can be described by upward moving paresthesia with neck flexion which can be a sign of myelopathy.^[7]

Causes

The causes for Lhermitte's sign are shown in the Table 1.^[8,9]

Validity

Two studies have measured the diagnostic accuracy of Lhermitte's sign which is found to be sensitive ranging from 3 to 17%, which is poor. One of these studies also found out that it had good specificity (97%) for non-specific compressive myelopathy.^[10]

Review

A study reported that Lhermitte's sign was experienced by 33.3% out of 114 patients of MS; and in 16%, it was reported to have been occurred in the first episode of MS. One out of eleven patients with subacute combined degeneration of the

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Table 1: The causes of Lhermitte's sign are listed below

Causes
Multiple sclerosis
Cervical spondylitis
Transverse myelitis
Behçet's disease
Sub-acute combined degeneration of the cord
Radiation myelopathy
Arnold-Chiari malformation
High-dose chemotherapy
Discontinuation syndrome associated with SSRIs Selective serotonin reuptake inhibitor
Trauma
Arachnoiditis
Syringomyelia
Vitamin B12 deficiency 6
Disc herniation
Spinal cord compression
Cisplatin neurotoxicity
Nitric oxide toxicity
Systemic lupus erythematosus
Parasitic invasion of spinal cord
Post-dural puncture headache
Herpes-zoster infection

cord due to pernicious anemia also reported the presence of Lhermitte's sign. The sign is said to occur commonly in conditions such as subacute combined degeneration of the cord, neck trauma, prolapsed cervical disc, and radiation myelitis. Lhermitte's sign has also been reported in cavernous angioma of the cervical spinal cord.^[11] A case reported that a 49-year-old woman diagnosed as having breast cancer and on treatment with cisplatin presented with Lhermitte's sign. An 80-year-old man previously operated on for adenocarcinoma colon, with no further treatment also presented with Lhermitte's sign. A 54-year-old man who was being treated for laryngeal cancer by radiotherapy presented with Lhermitte's sign. Lhermitte's sign is a non-specific sign, although in oncological patients a detailed history and clinical examination should be done for data regarding radiotherapy, chemotherapy, and spinal compression.^[12] A strong association between Lhermitte's sign and abnormalities of the cervical spinal cord has been seen on magnetic resonance imaging. The study presumed that Lhermitte's sign in MS is the result of a lesion in the cervical spinal cord and it was confirmed that a lesion in the posterior columns of the cervical spinal cord is the cause of Lhermitte's sign in MS.^[13] A case was reported where Lhermitte's sign occurred during yawning and was associated with congenital partial aplasia of the posterior arch of the atlas. Computed tomography (CT) myelography during yawning showed compression of the upper cervical cord due to the inward mobility of the isolated posterior tubercle. The symptoms completely disappeared following removal of the isolated posterior tubercle.^[14] Another case of a 34-year-old man suffering from herpes zoster was accompanied by Lhermitte's sign.^[15] A study was done to investigate the pathophysiology of the radiation-induced, chronic Lhermitte's sign on the basis of long-standing case histories with partial functional recovery. Positron Emission Tomography (PET) demonstrated increased fluorodeoxyglucose (FDG) accumulation and butanol

perfusion, but negligible methionine uptake in the irradiated spinal cord segments in the patients.^[16] A 29-year-old boy with an intrinsic, fusiform mass extending from C5 to C7, identified as low-grade ependymoma; developed Lhermitte's sign. Lhermitte's sign was most likely caused by tumor-induced distortion and demyelination of cervical dorsal column sensory axons.^[17] Another case reports of two patients who developed intrinsic cervical spinal cord damage as permanent complication of cervical epidural steroid injections which were administered while the patients were sedated. The patients were found to develop Lhermitte's sign.^[18]

Response to treatment

A study conducted treatment with extra-cranial picotesla range pulsed electromagnetic fields (EMFs) which was found to be effective in the management of various MS symptoms. Three MS patients in whom two brief applications of EMFs were done resulted in resolution of the Lhermitte's sign which emerged during a period of exacerbation of symptoms in one patient and during a prolonged phase of symptom deterioration in the other two patients with MS. As Lhermitte's sign is thought to result from the spread of ectopic excitation in demyelinated plaques in the cervical and thoracic regions of the spinal cord, it was hypothesized that the effects of EMFs were related to the reduction of axonal excitability via a mechanism involving changes in ionic membrane permeability.^[19] Neck movements are said to exacerbate Lhermitte's sign therefore, a brace can keep the patient from bending his neck too much which may be prescribed by a physical therapist to help with posture and positioning of the head in such cases. If a neck brace or collar is used, periodic monitoring is required to ensure that strength and range of motion is not compromised. An occupational therapist may offer progressive muscle relaxation techniques, deep breathing exercises, and active or passive stretching.^[20]

Conclusion

Lhermitte's sign is popularly described as a shock-like sensation by MS patients commonly. But recently several other causes for the Lhermitte's sign have been found where the most recent one includes the IRMT therapy for head and neck cancer patients. Lhermitte's sign has variants such as inverse and reverse Lhermitte's phenomena in which it can present itself. Diagnosis at the right time with proper management can help the patient cope up with the sign.

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