



Seán J Slaght (pictured)¹,
Ian Galea¹,
Susan O'Connell²,
Simon Hammans¹

1: Wessex Neurological Centre,
Southampton General Hospital,
Tremona Road, Southampton,
SO16 6YD.

2: Health Protection Agency Lyme
Borreliosis Unit, Southampton
General Hospital, Tremona Road,
Southampton, SO16 6YD.

Correspondence to:
sean.slaght@uhs.nhs.uk

This case report won the ACNR
sponsored prize for Best Case
Report at the ABN meeting in
Newcastle, October 2011.

Help, I've become shorter than my wife! Camptocormia due to Lyme Neuroborreliosis

Camptocormia (or bent spine syndrome) is the abnormal flexion of the thoracolumbar spine, which increases with standing or walking and resolves on lying down. Camptocormia is particularly associated with advanced Parkinson's disease, but is also described in a number of other (predominantly neurodegenerative) central nervous system disorders, as well as disorders of the peripheral nerve, muscle and psychiatric conditions. For a recent review of camptocormia see Finstere and Strobl.¹

The only infection previously reported to have camptocormia as part of its clinical course is Viliuisk encephalomyelitis.² This is a rare and devastating disorder of the central nervous system, seen only in the Yakut population of Eastern Siberia, which invariably leads to death. Viliuisk encephalomyelitis is transmitted horizontally, but the causative organism has not yet been identified.³

Lyme neuroborreliosis (LNB) is the major complication of Lyme borreliosis (LB), which is due to infection with the spirochaete *Borrelia burgdorferi*.⁴ Prospective, population based studies show that around 3% of patients with LB develop LNB.⁴ The most common presentations are with facial palsy (unilateral or bilateral), meningoencephalitis or polyradiculopathy.⁵ Neuroborreliosis can be successfully treated with either oral doxycycline or intravenous ceftriaxone.⁶

We believe this is the first case of camptocormia due to Lyme neuroborreliosis to be reported in the medical literature.

The case

In October 2009, the patient sought medical help as he had become shorter than his wife. He was a 57-year-old ex-smoker with well-controlled hypertension and type-2 diabetes mellitus. He was otherwise fit and well and a keen amateur ornithologist and wildlife photographer (Figure 1a).

About four to six weeks prior to presentation he had noticed a slightly itchy rash behind his left knee. Shortly after this, he developed pain in his left knee, which gradually progressed up his left leg to his hip. His left leg became weak and over

the next four to six weeks posture started to stoop. This continued to the point that he became shorter than his wife.

Examination revealed a stooped posture consistent with camptocormia (video and Figure 1b). Muscle tone was normal. He had mild weakness of the neck flexors, truncal and abdominal muscles, as well as in the left arm and leg. He was areflexic. There was a suspended sensory level from T8-T10 on the left hand side.

Routine blood screen was unremarkable, as was MRI of his spine. Lumbar puncture showed clear cerebrospinal fluid with an opening pressure of 15 cmH₂O. There were no red cells, but 100 white blood cells: 66% neutrophils and 32% lymphocytes. The protein was 1884 mg/L and the glucose 5.9 (matched serum 10.9). Unmatched cerebrospinal fluid oligoclonal bands were seen. Western blot for antibodies against *Borrelia burgdorferi* were positive in blood and cerebrospinal fluid (Figure 2).

He was treated with a two-week course of IV ceftriaxone, with rapid resolution of his leg pain. The strength in his leg and his posture recovered back to normal over the next few months (video). In March 2010, he was able to take part in a sponsored 10 km walk (Figure 1c).

Discussion

We believe this is the first reported case of camptocormia caused by an identified infectious agent: *Borrelia burgdorferi*. As with most cases of Lyme neuroborreliosis, our patient made a full recovery after treatment with antibiotics. The camptocormia appeared to be caused by paraspinal muscle weakness secondary to a polyradiculopathy. This assumption is backed by a recent case of camptocormia caused by chronic inflammatory demyelinating polyradiculopathy, which responded to treatment with intravenous immunoglobulins.⁷

Lyme disease classically presents with erythema migrans; an expanding area of erythema associated with the point of entry of the spirochaetes.⁴ Eighty-nine per cent of patients with LB in a large prospective population based

study in Germany had erythema migrans.⁸ Although, in a laboratory based study in Devon only 36% of patients with LBN had a preceding erythema migrans and 27% had no memory of a bite, rash headache, fever or arthralgia/myalgia.⁵ Our patient did have an itchy rash prior to developing his camptocormia, but this only came to light following direct questioning. The patient is a keen wildlife photographer and had spent time in Lyme infested areas prior to his illness, although he did not have any memory of a tick bite, in common with 45% of patients with Lyme neuroborreliosis.⁵

Most cases of camptocormia are due to degenerative disorders such as Parkinson's disease and develops several years after the diagnosis has been made.¹ The rapid onset of camptocormia in our patient would be unusual for a degenerative cause and in such cases a search for reversible causes should be sought.

Lyme neuroborreliosis should be considered in the differential diagnosis of camptocormia. ♦



Figure 1: Photographs of the patient prior to his illness (A), at presentation (B) and following his recovery (C).

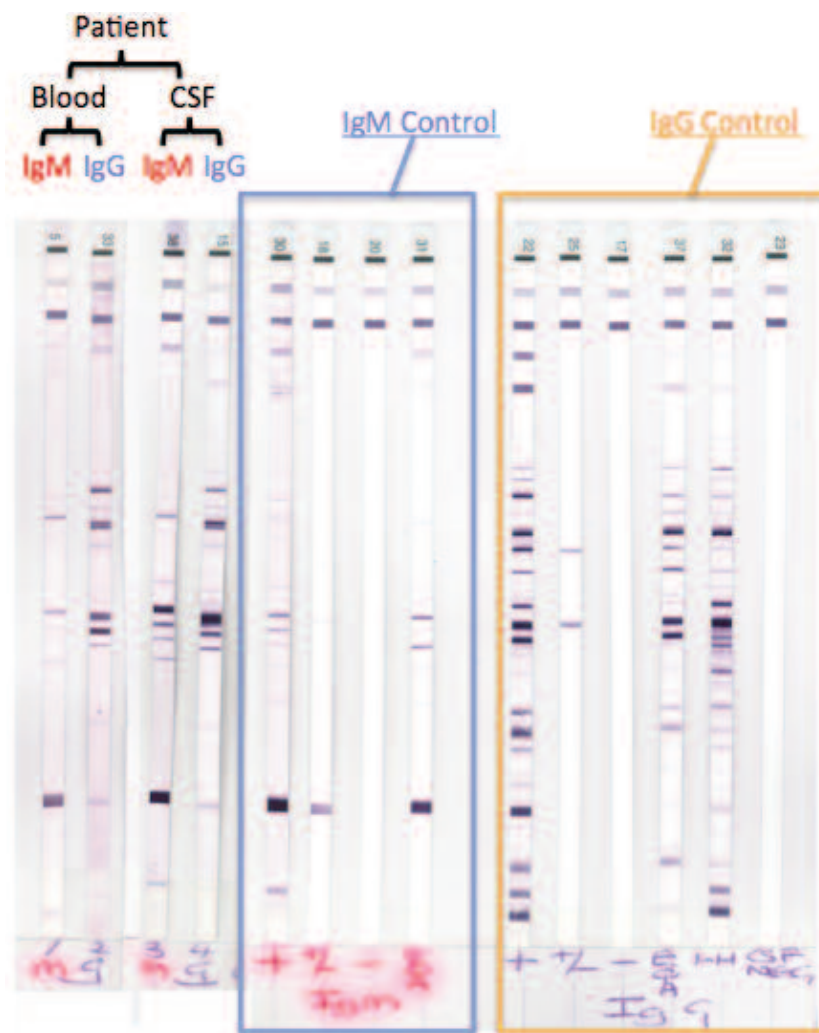


Figure 2: Western blots from the patient (first 4 lanes) from blood and CSF for both IgM and IgG as well as positive and negative controls for IgM (within blue square) and IgG (within orange square). The positive bands to IgG and IgM in the patient's CSF and blood confirm the diagnosis of Lyme neuroborreliosis.

REFERENCES

1. Finstere J and Strobl W. *Presentation, Etiology, Diagnosis, and Management of Camptocormia*. *Eur Neurol* 2010;64:1-8.
2. Goldfarb LG and Gajdusek DC. *Viliuisk Encephalomyelitis In The Yakut People Of Siberia*. *Brain* 1982;115:961-78.
3. Goldfarb LG, Vladimirtsev VA, Platonov FA, Lee H-S, McLean CA and Masters CL4. *Viliuisk encephalomyelitis in Eastern Siberia – analysis of 390 cases*. *Folia Neuropathologica*, 2009;47:171-81.
4. Stanek G, Wormser GP, Gray J, Strle F. *Lancet* 2011. *Lyme borreliosis*. Published online September 7, 2011 DOI:10.1016/S0140-6736(11)60103-7
5. Lovett JK, Evans PH, O'Connell S and Gutowski NJ. *Neuroborreliosis in the South West of England*. *Epidemiol. Infect.* 2008;136:1707-11.
6. Ljøstand U, Skogvoll E, Eikeland R, Skarpaas T, Berg A, and Mygland A. *Oral doxycycline versus intravenous ceftriaxone for European Lyme neuroborreliosis: a multicentre, non-inferiority, double-blind, randomised trial*. *Lancet Neurology* 2008;7:690-5.
7. Terashima M, Kataoka H, Sugie K, Horikawa H and Ueno S. *Coexistence of chronic inflammatory demyelinating polyneuropathy and camptocormia*. *J Neurol Neurosurg Psychiatry* 2009;80:1296-7.
8. Huppertz HI, Böhme M, Standaert SM, Karch H, Plotkin SA. *Incidence of Lyme borreliosis in the Würzburg region of Germany*. *Eur J Clin Microbiol Infect Dis*. 1999;18:697-703.