

Clinical Reports

Leptospirosis presenting as a flaccid paraplegia

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Summary: A fatal case of leptospirosis in a 64 year old farm worker is described. The dramatic neurological presentation with a rapidly evolving flaccid paraplegia associated with biochemical evidence of renal and hepatic dysfunction is discussed. Attention is drawn to the wide range of neurological symptoms reported in leptospirosis, and to the possibility that this infectious disease may present neurologically.

Introduction

Neurological involvement in leptospirosis is rare. Both central and peripheral neurological manifestations of the disease may be seen, but usually in the context of advanced infection, where the diagnosis has already been established. We report a fatal case of leptospirosis which presented with prominent neurological symptoms.

Case report

A 64 year old farm worker was admitted to a general surgical ward complaining of inability to pass urine, abdominal pain and difficulty walking due to leg weakness. This combination of symptoms led the admitting house surgeon to suspect acute spinal cord compression, and an urgent neurological opinion was sought.

On close questioning, the patient gave a 72 hour history of increasing leg weakness associated with pain in the lower limbs. He had also been anuric for a similar period of time. As a complicating aspect to the history, he had been painting in an enclosed space one week before admission, following which he immediately developed generalized muscle aches and malaise. The patient himself was adamant that this was the cause of his symptoms.

On initial examination the patient was alert but mildly disorientated in time and place. He was afebrile, 36.5°C. There was a suspicion of scleral jaundice with marked conjunctival suffusion. There was generalized abdominal tenderness.

The bladder was not felt and rectal examination was normal.

Neurological examination of cranial nerves and the upper limbs was unremarkable. Examination of the lower limbs showed a flaccid paraplegia with areflexia and downgoing plantars. All sensory modalities were preserved, indeed, trivial stimulation to either leg produced intolerable painful paraesthesiae.

A working diagnosis of an acute symmetrical painful motor neuropathy of unknown cause was made. In view of the history of recent exposure to paint, a toxic aetiology was thought most likely.

Initial laboratory investigations showed a neutrophil leucocytosis with neutrophil toxic granulation, thrombocytopenia and the presence of burr cells. Haemoglobin 13.1 g/dl, white cell count $16.5 \times 10^9/l$, 94% neutrophils, platelets $34 \times 10^9/l$. Serum creatinine was elevated: 748 $\mu\text{mol/l}$ (range 30–90 $\mu\text{mol/l}$). Liver function was deranged: bilirubin 241 $\mu\text{mol/l}$ (range up to 17 $\mu\text{mol/l}$), gamma glutamyl transferase (gamma GT) 67 IU/l (range up to 50 IU/l) and alanine aminotransferase (ALT) 64 IU/l (range up to 40). Alkaline phosphatase and serum amylase were normal. Creatine kinase (CK) was elevated: 969 IU/l, (range 24–195 IU/l).

Transurethral catheterization had been performed on admission with negligible yield of urine despite fluid challenge. Urine testing was strongly positive for bilirubin and negative for urobilinogen.

An upper abdominal ultrasound was normal, with no intrahepatic or extrahepatic duct dilation. No focal liver abnormality was seen. Chest, abdominal and spinal X-rays were normal. Nerve conduction studies showed normal motor and sensory conduction velocities in both upper and lower limbs.

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On receipt of the biochemical data suggesting both renal and hepatic dysfunction, and in view of the patient's occupation as a farm worker, Weil's syndrome due to leptospirosis was considered a probable unifying diagnosis. Treatment with benzyl penicillin 1.2 g i.v. four times a day was commenced and the patient was transferred to the local renal unit for dialysis.

On transfer to the renal unit he was found to be deeply jaundiced, pyrexial and confused. Neurologically the areflexia had extended to involve the upper limbs. He underwent acute haemodialysis via a right subclavian line without any complications. Later that day he suffered an asystolic cardiac arrest from which he could not be resuscitated.

Positive serological tests for leptospirosis became available shortly after death: microscopic agglutination test positive 1 in 40, IgM ELISA positive 1 in 640, the latter figure confirming recent leptospiral infection.

A post-mortem examination was performed. Ischaemic changes with a mild lymphocytic infiltrate were seen in the myocardium, and the lungs were congested. The liver was markedly enlarged (3200 g) and pale, with a nutmeg pattern visible macroscopically on cut surface. Microscopic examination revealed focal acute hepatitis. Both kidneys were enlarged (right 240 g, left 320 g) with histological evidence of acute tubular necrosis, interstitial haemorrhage and lymphocytic infiltrate. The brain and spinal cord were unremarkable on macroscopic and microscopic examination. There was no evidence of structural spinal cord pathology or haemorrhage.

Discussion

Diverse central nervous system (CNS) abnormalities have been reported in leptospirosis, including encephalitis, coma, convulsions, hemiplegia, myelitis and movement disorders. Peripheral nerve abnormalities reported include transient lower limb weakness, areflexia, paraesthesiae, neuropathies and the Guillain-Barré syndrome.¹⁻³

The neurological manifestations result from the effect of the organism on the central nervous system, and the host's immune reaction to the organism. Following infection, leptospirae reach the CNS rapidly. The spirochaetes can usually be cultured from the CSF for the first week of clinical illness, but not following the appearance of specific antibodies. Changes in the cerebrospinal fluid contents are a later manifestation, and it has therefore been suggested that some of the inflammatory neurological manifestations of leptospirosis result not from the organism itself but from the

antibody reaction to it.^{3,4}

The present case had an ascending polyneuropathy in the absence of sensory loss. The neurological picture was progressive, and reminiscent of the Guillain-Barré syndrome. The probable cause was meningeal and radicular inflammation as a result of antibody production. Post-mortem examination showed no evidence of other CNS complications such as intraparenchymal bleeding or spinal extradural haematoma. Both of these are recognized in leptospirosis, and could arguably have produced a similar clinical picture.

The striking feature of this case is that the initial presentation was with neurological symptoms. Indeed, such was the severity of the neurological disability, that the immediate medical concern was to exclude primary disease of the nervous system. Although subtle clues to the overall diagnosis were present, these were overlooked until biochemical and haematological abnormalities were identified.

The presentation of leptospirosis as a neurological syndrome is unusual. In the immune phase of the illness an aseptic meningitis is common, with up to 90% of cases having a cerebrospinal fluid pleocytosis. About half of these cases will have symptoms of meningeal irritation. However, these features are usually incidental to an established and generalized illness, with the symptoms of the initial leptospiraemic phase of the illness having passed some days previously. The majority of florid neurological features which are seen occur when the diagnosis has already been made, and treatment may be in progress. Cerebral symptoms in the early leptospiraemic phase of the illness probably occur in fewer than 25% of patients.⁵

The biochemical and haematological abnormalities seen in the present case are all consistent with the diagnosis. A neutrophil leucocytosis is common, though occasionally leucopenia is seen. Thrombocytopenia is a less consistent, but well-recognized finding during the illness. The platelet count may fall sufficiently to give rise to bleeding (less than 30×10^9 platelets per litre).⁶ No pathological bleeding was identified in the present case.

There has been recent media attention to a changing epidemiological pattern of leptospirosis infection in the United Kingdom. Whereas previously this was a condition of farm and sewer workers who were exposed to rats or rats' urine, the incidence of infection is now increasing in teenagers and young adults, who enjoy recreational pastimes such as swimming, wind-surfing and water skiing on inland waterways.^{7,8}

The present case illustrates that leptospirosis may present as a primary neurological problem. General physicians and neurologists should be aware of the varied manifestations of leptospirosis, and the larger population now susceptible.

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