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Case report

Longitudinally extensive transverse myelitis with seropositive chikungunya

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SUMMARY

Chikungunya viral (CHIKV) fever is often a self-limiting febrile illness associated with severe debilitating arthralgia. Neurological complications associated with CHIKV, although rare, have been reported in literature; however, longitudinally extensive transverse myelitis (LETM) is rarely associated with it. We present a case of a middle-aged man with a 1-week history of low-grade fever and arthralgia followed by urinary retention and quadriplegia. A sensory level was noted at T2. On subsequent investigations, he was diagnosed with LETM. Although LETM is commonly seen in patients with neuromyelitis optica, the other possible etiologies are inflammatory and parainfectious. To date, only two cases of LETM are reported worldwide in association with CHIKV fever and this is the first case from Pakistan. With frequent chikungunya outbreaks, neurological complications are increasingly seen in clinical practice. The knowledge of these associations will result in their early diagnosis and treatment.

BACKGROUND

Chikungunya viral (CHIKV) fever is generally considered a self-limiting febrile illness associated with severe debilitating arthralgia. Although CHIKV-associated neurological complications have been reported in literature, they are uncommon and,¹ longitudinally extensive transverse myelitis (LETM) is very rare and has only been reported in two previous cases worldwide.^{2,3}

CASE PRESENTATION

A 40-year-old man presented with a 1-week history of low-grade fever associated with mild arthralgia. Two days prior to admission, he developed bilateral leg weakness and urinary retention. On the day of admission, he became drowsy and was disoriented with no signs of meningeal irritation. Pupils were bilateral, 4 mm in diameter and reactive to light. Fundus examination was normal. Extraocular movements were normal and there was no facial asymmetry. Muscle strength was Medical Research Council (MRC) grading 4/5 in bilateral upper limbs and 2/5 in bilateral lower limbs in all muscle groups with increased deep tendon reflexes in all four limbs and bilateral extensor plantar responses. No clonus was observed. He had a sensory loss in all modalities below T2 level.

INVESTIGATIONS

Cranial MRI was unremarkable, while MRI of the spine revealed diffuse patchy abnormal T2-hyperintense signals within the cervical and thoracic spinal cord without contrast enhancement involving more than three consecutive vertebrae along with mild swelling of the cervical cord (**figure 1**). Cerebrospinal fluid (CSF) analysis was performed on day 2 (**table 1**). Serum IgM antibodies against CHIKV were detected. Serum dengue serology was negative. Malarial parasite was not seen on peripheral film examination and malarial antigen were not detected by the rapid immunochromatographic test. Aquaporin-4 antibodies tested by ELISA for neuromyelitis optica (NMO) disorder were also negative. Therefore, the diagnosis of encephalitis and LETM secondary to CHIKV was made. CSF-CHIKV antibodies were not available, which is a limitation. We also did not investigate for CSF oligoclonal bands and other coinfections including Zika virus and HIV.

DIFFERENTIAL DIAGNOSIS

Based on the history and examination, the differential diagnosis of transverse myelitis, compressive myelopathy and acute disseminated encephalomyelitis were considered. After MRI spine, we initially considered NMO spectrum disorder along with infectious and parainfectious causes.

TREATMENT

The patient was initially administered ceftriaxone 2 g two times a day, vancomycin 750 mg every six hours and acyclovir 750 mg every eight hours for

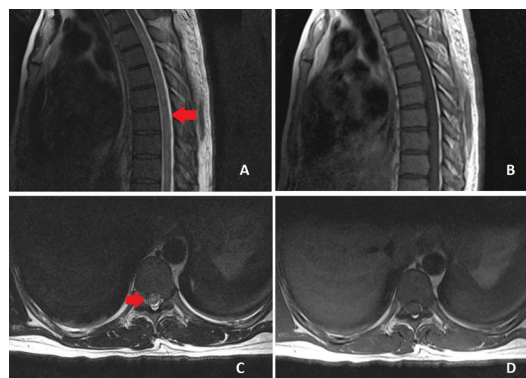


Figure 1 MRI of the spine showing diffuse patchy abnormal T2-hyperintense signals from T2-L1 vertebrae (red arrows in panels A and C) without contrast enhancement on T1-post contrast images (panels B and D).



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Table 1 Cerebrospinal fluid (CSF) analysis

| | |
|---|--|
| CSF leucocyte count | 68/uL (range 0–5/uL) |
| CSF protein | 70 mg/dL (range 15–40 mg/dL) |
| CSF glucose | 75 mg/dL (serum glucose was 114 mg/dL) |
| CSF PCR for HSV 1 and 2, HHV 6, VZV, CMV, enterovirus, Neisseria meningitidis, Listeria monocytogenes, Streptococci pneumonia and Cryptococcus neoformans | Negative |
| Gene Xpert for <i>Mycobacterium tuberculosis</i> | Negative |
| CSF culture | Negative |

CMV, cytomegalovirus; HHV, human herpes virus; HSV, herpes simplex virus; VZV, varicella zoster virus.

1 day but later discontinued after the CSF results and positive chikungunya serology. He was further managed with methylprednisolone (1 g/day, intravenous) for 3 days followed by oral steroids in tapering doses for 4 weeks and rehabilitation.

OUTCOME AND FOLLOW-UP

The patient improved with the treatment. His consciousness level improved significantly. He was more awake and oriented by day 3. Muscle strength in bilateral lower limbs improved to MRC grade 3/5 on discharge (on day 6) and to 4/5 on the first outpatient follow-up (on day 24). We planned for a repeat MRI imaging of the spinal cord to assess the radiological improvement in the next follow-up outpatient visit but unfortunately, the patient did not follow-up.

DISCUSSION

LETM is described as intramedullary T2-hyperintense cord signals extending over three or more consecutive vertebrae. It is commonly associated with NMO but different autoimmune, infective, inflammatory, paraneoplastic and vascular etiologies have been proposed.⁴ In our case, the temporal association of fever and arthralgia, positive CHIKV serology, radiological and clinical features of LETM make CHIKV the most likely aetiology. Other arboviruses, dengue virus and Zika virus, have also been linked with LETM in literature.⁵ Irrespective of the aetiology, LETM results in severe disability.⁴

CHIKV is an RNA arbovirus transmitted by the bites of *Aedes* mosquitoes. The word chikungunya originates from African language and means ‘which bends up’, so named due to the severe debilitating arthralgia. CHIKV-associated neurological complications are rare. Encephalopathy is the most common CHIKV-associated neurological complication; however, cases of acute disseminated encephalomyelitis, Guillain-Barre syndrome, myeloradiculitis, myelitis and neuro-ocular diseases (uveitis, retinitis, optic neuritis) have also been reported.¹ CHIKV-associated LETM is very rare and only two cases have been reported in literature.^{2,3} We are reporting the first case of CHIKV-associated LETM from Pakistan and we have previously reported a case series of CHIKV-associated Guillain-Barre syndrome presenting in our institution.⁶

Due to the rarity of this condition, no significant data regarding the management of CHIKV-associated LETM is available. The previous case of CHIKV-associated LETM was managed with high dose intravenous steroids with good functional outcome.² In our case, a similar positive outcome was seen with the steroids. It is inconclusive whether steroids hasten recovery in these cases or the symptoms resolve spontaneously with time as other viral features do. Further studies are needed to evaluate the response to steroids in this condition.

Our case report and previous experience with CHIKV⁶ also highlight the fact that CHIKV fever is not a benign condition, as previously considered. It is associated with diverse neurological dysfunctions. With frequent CHIKV outbreaks, these associations should be considered in the management of the patients.

Learning points

- ▶ Chikungunya virus (CHIKV) is not always a benign condition and may present with variable neurological complications, including Guillain-Barre syndrome, encephalitis and transverse myelitis.
- ▶ Infectious and parainfectious causes, including arboviruses, should always be included in the differential diagnoses of longitudinally extensive transverse myelitis (LETM).
- ▶ Steroids have shown good results in the management of CHIKV-associated LETM.

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