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CASE REPORT

Rare variant of Guillain-Barré syndrome after chikungunya viral fever

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SUMMARY

Chikungunya (CHIK) viral fever is a self-limiting illness that presents with severe debilitating arthralgia, myalgia, fever and rash. Neurological complications are rare. We present a case of a 36-year-old woman who presented with acute onset progressive difficulty swallowing and left arm weakness. She was diagnosed with CHIK viral fever 4 weeks prior to admission. After investigations, she was diagnosed with a pharyngeal–cervical–brachial variant of Guillain-Barré syndrome. In hospital, she required ventilator support. Her condition improved after five sessions of intravenous immunoglobulin with almost complete resolution within 6 months of symptom onset. With frequent CHIK outbreaks, the neurological complications are increasingly seen in the emergency department. The knowledge of these associations will result in early diagnosis and treatment.

BACKGROUND

Chikungunya (CHIK) fever is a self-limiting viral fever, that is, transmitted by the bite of *Aedes* mosquitoes, the same vectors for dengue fever.[1] Multiple outbreaks of CHIK fever have occurred in Asia and Africa in the last two decades affecting millions.[2] CHIK fever typically presents with fever, severe arthralgias, myalgias and skin rash. Atypical presentations with myocarditis, hepatitis and meningoencephalitis may also occur.[3] Cases of transverse myelitis and Guillain-Barré syndrome (GBS) secondary to CHIK fever have been reported.[4–6] Although these cases are rare, with frequent outbreaks of CHIK fever, they are increasingly being seen in clinical practice. Recognition of various neurological complications is important. We are reporting a case of a rare variant of GBS, the pharyngeal–cervical–brachial (PCB), secondarily to a CHIK infection.

CASE PRESENTATION

A 36-year-old woman presented to us with a 3-day history of difficulty swallowing followed by weakness in her left arm for 1 day. She had difficulty in swallowing both liquids and solids. It was progressive and associated with nasal regurgitation, voice changes and drooling of saliva. Her symptoms were continuous without a diurnal variation. The left arm weakness was noticed on awakening 1 day ago and was progressive. She denied symptoms of sore throat, fever, headache, neck pain, double vision, facial or limb sensory disturbances and hearing problems associated with her current symptoms. Four weeks prior to this admission, she had experienced low-grade fever, generalised body aches and moderate pain in her joints, with predominant involvement of knees, shoulders and ankle joints. Fever subsided within the next 5–6 days; however, the joint pains persisted although the severity had decreased. She was diagnosed with having CHIK viral fever on clinical grounds and was taking analgesics on as per needed basis. There was no history of chronic illnesses.

On physical examination, she was vitally stable, awake and oriented with intact higher mental functions. Extraocular movements were intact in all directions without nystagmus. She had a hypernasal speech. Pupils were bilaterally 3 mm equal and reactive to light and accommodation. She had a right-sided facial palsy of lower motor neuron type. Facial sensation was intact. Gag reflex was weak and uvula was deviated to the left side. Tongue was central on protrusion with no fasciculations. On motor examination, muscle bulk and tone were normal. Power in left upper limb was reduced. Medical Research Council (MRC) grade was 3/5 proximally and 4/5 distally with reduced deep tendon reflexes, while power and reflexes in the remaining limbs were normal. Sensory and cerebellar examination was normal.

INVESTIGATIONS

A baseline workup, including complete blood counts, serum electrolytes and thyroid profile, was unremarkable. IgM antibodies against CHIK virus were detected. She underwent MRI of the brain (on day 1) which did not reveal any acute abnormalities. Cerebrospinal fluid (CSF) examination (on day 2) was normal with no albuminocytological dissociation. On the same day (day 2), nerve conduction studies with electromyography (EMG) revealed non-specific findings, with prolongation of bilateral R1, R2 and contralateral R2 latencies on blink testing. On EMG, voluntary motor unit action potentials had normal morphology with decreased recruitment and a rapid firing rate. In the clinical setting, these were suggestive of an early neurogenic process, possibly demyelinating in nature and a diagnosis of the PCB variant of GBS was made. Testing for antiganglioside antibodies (IgG anti-GT1a antibodies) is not available which was a limitation for definitive diagnosis.

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DIFFERENTIAL DIAGNOSIS

Due to right-sided facial weakness, bulbar symptoms and left-sided motor weakness, our provisional diagnosis was of a posterior circulation stroke involving the brain stem. The other differentials were GBS, botulism, myasthenia gravis, postviral autoimmune demyelination or multiple sclerosis.

TREATMENT

Treatment with intravenous immunoglobulin (IVIG) was started on the second day of admission for a total of 5 days (total dose of 2 g/kg). On the same day (day 2), she became tachypneic with a respiratory rate of 34 breaths/min, and reduction of forced vital capacity to 500 from 1250 mL on the first day. Her single breath count was 12 with arterial blood gases (on 5 L oxygen supplementation) revealing a pH of 7.55, $paCO_2$ of 25.30 mm Hg, paO_2 of 167.30 mm Hg and bicarbonate of 21.80 mEq/L. She was intubated and mechanically ventilated.

OUTCOME AND FOLLOW-UP

Her symptoms partially improved after the five doses of IVIG. Rehabilitation was started. Her condition gradually improved and she was discharged after 10 days with regular physiotherapy. She had normal bulbar function with MRC grade 4+/5 power in the left upper limb at the sixth month of follow-up.

DISCUSSION

CHIK virus is an RNA virus that belongs to Togaviridae family. It is transmitted by the bites of mosquitoes, *Aedes aegypti* and *Aedes albopictus*.[3] The word Chikungunya is derived from an African language that means 'that which bends up', named due to the symptoms of severe debilitating joint pain.[7] Apart from fever, arthralgia, myalgia and skin rash, atypical presentations of hepatitis and myocarditis are also seen.[5] Neurological complications are rare. Less than 1000 cases of CHIK-associated neurological disease are reported in the literature with encephalopathy being the most common complication.[8] Peripheral neuropathy without central nervous system disease, including GBS, is even rarer.

GBS is a postinfectious autoimmune polyneuropathy that classically presents with an acute-onset of symmetrical, bilateral flaccid weakness. It has a median incidence rate of 1.1 per 100 000 in Europe and North America.[9] A twofold increase in the incidence of GBS was reported in the French West Indies during the 2014 CHIK outbreak.[10] GBS is a heterogeneous disorder with multiple subtypes. The common forms of GBS are acute inflammatory demyelinating polyneuropathy, acute motor axonal neuropathy (AMAN) and acute motor sensory axonal neuropathy. The uncommon forms of GBS include Miller Fisher syndrome, Bickerstaff's brainstem encephalitis and PCB variant.[11]

The PCB variant of GBS typically presents with the pharyngeal, neck and upper limb weakness with associated areflexia/hyporeflexia. Power in lower limbs is normal or mildly affected. Sensations are often normal.[12] Respiratory failure is not uncommon. In a study by Nagashima *et al*, 27% of the patients with PCB variant GBS required endotracheal intubation.[13] PCB variant is often misdiagnosed with brainstem stroke or myasthenia gravis. Electrophysiological studies and CSF albuminocytological dissociation (ie, high levels of protein with normal cell counts) help in the diagnosis but they can be normal in the early disease.[12 14] Nerve conduction studies usually show a localised pattern of axonal damage similar to AMAN. Further, the presence of IgG anti-GT1a antibodies is strongly associated with the PCB variant.[12]

GBS is a treatable condition. Plasma exchange and IVIG are equally effective in reducing the nerve damage and hastening the recovery if given early in the disease within the first 2 weeks. Patients should be observed in a critical care unit for dysautonomia and respiratory compromise. Mechanical ventilation may be needed.[15] With the current CHIK viral fever pandemic, the caregivers should be aware of neurological complications for their early diagnosis and treatment to decrease morbidity and mortality.

Learning points

- Chikungunya (CHIK) viral fever is associated with neurological complications, although rare.
- Cases of meningoencephalitis, transverse myelitis and Guillain-Barré syndrome in association with CHIK viral fever have been reported in the literature.
- High clinical suspicion, prompt diagnosis and treatment are important for a good neurological outcome.

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