

Guillain Barre syndrome as a manifestation of neurological melioidosis

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Abstract

Neurological melioidosis is a very rare and very few cases have been reported from India. Presentation is an extremely varied and as this disease is associated with high mortality, high index of suspicion is needed to diagnose and treat. In this context, we report a patient presenting as Guillain Barre syndrome evaluated as melioidosis.

Key Words

Guillain Barre' syndrome, infection, melioidosis

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Introduction

Guillain Barre syndrome (GBS) is a post infectious poly radiculoneuropathy and many infections have been reported to be associated with it.^[1] The various infections preceding GBS that have been described in case control studies or large case series include Cytomegalovirus (CMV), Epstein-Barr virus (EBV), varicella zoster, *Campylobacter jejuni*, *Mycoplasma pneumoniae*.^[2] GBS associated with *Burkholderia pseudomallei* is very rare and has been reported only once.^[3]

B. pseudomallei is transmitted by percutaneous inoculation and inhalation. It is a predominantly found in Australia, South-East Asia^[4] and preferentially affects diabetic, alcoholic and chronic kidney disease patients.^[5] Though, the commonly affected organs are skin and lung it can affect almost all systems including nervous system. Neurological melioidosis is very rare and most of the cases have been reported from Northern Australia.^[6] Very few cases have been reported from India.^[7] In this context, we report a patient presenting as GBS evaluated as melioidosis.

Case Report

A 53-year-old gentleman, known diabetic of 8 years duration presented with a pain in both lower limbs of 15 days duration and weakness in the form of ascending quadriparesis involving proximal followed by distal group of muscles of 12 days duration. There were no cranial nerve and respiratory symptoms. There was history of malaise and low grade fever preceding this illness.

On examination at admission, vital signs were normal. General physical examination was normal. Higher mental functions and cranial nerve examination were normal. There was hypotonia in all four limbs with Medical research council (MRC) power of grade 4/5. The deep tendon reflexes were absent and he had bilateral flexor plantar response. Cerebrospinal fluid examination revealed albumino-cytological dissociation (Glucose 70 mg%, protein 88 mg%, no cells). Nerve conduction studies were suggestive of demyelinating polyradiculoneuropathy. He was detected to be HBs Ag positive and was seronegative for human immunodeficiency virus infection. Apart from elevated glycosylated hemoglobin A_{1c} level of 7.8%, serum biochemical parameters including potassium levels were normal. He had neutrophilic leukocytosis (13,400/cmm, 88% neutrophils), blood cultures were sent and was empirically started on ceftriaxone. IV immunoglobulins of 0.40 g/kg/day were given for 5 days. During the course of the hospital stay, his blood sugars were kept under control with insulin.

On 2nd day of admission, patient developed bilateral facial weakness, power deteriorated to MRC grade 3/5. After 4 days of treatment, the patient developed respiratory distress requiring ventilatory support. Arterial blood gases report showed metabolic acidosis with raised lactate levels and hypoxia. Later,

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patient developed refractory septic shock and succumbed to death. Cultures grew *B. pseudomallei* and the diagnosis of melioidosis presenting as GBS on a background of diabetes mellitus was considered.

Discussion

We report a diabetic patient presenting with an acute radiculoneuropathy requiring ventilator support.

Diabetic neuropathy is a microvascular complication of diabetes and commonly presents as slowly progressive length dependent neuropathy, mononeuropathy or mononeuritis multiplex. Diabetic neuropathy presenting as acute quadriplegia is uncommon. Rarely acute quadriplegia may occur because of dyselectrolytemia,^[8] and diabetic amyotrophy.^[9] Further diabetic neuropathy manifests as pure sensory or sensorimotor neuropathy. However, predominant or pure motor neuron involvement including paralysis or weakness of extremities with generalized areflexia is suggestive of GBS. Albumino-cytological dissociation and nerve conduction studies are diagnostic for GBS.

Although, there are no reports showing high risk of GBS in diabetic patients, a few cases of GBS associated with diabetic ketoacidosis have been described.^[10-12] However, in the present case, the admission blood sugars were not very high (164 mg%) and were under control throughout the hospital course.

GBS with an underlying diabetic neuropathy is uncommon, but could possibly be a serious cause of persistent paralysis or weakness of limbs, although, this condition can be overlooked on presentation.

All forms of GBS are autoimmune diseases, due to an immune response to foreign antigens (such as infectious agents) that is targeted at host nerve tissues by a phenomenon called molecular mimicry.^[13] The targets of such immune attack are thought to be gangliosides compounds naturally present in the large quantities in human peripheral nerve tissues. GBS in approximately 70% of cases is preceded by some upper respiratory or gastrointestinal infection, the cause of which is mostly unidentified. In a case control study, multivariate analysis showed that infections with *C. jejuni* (32%), cytomegalovirus (13%), and EBV (10%) were significantly more frequent in GBS patients than in controls.^[14] The other infections, which may be associated with GBS include H. influenza, varicella, herpes, mumps and *Mycoplasma*. However, 60% of cases do not have a known cause.

B. pseudomallei is saprophytic bacteria in soil and stagnant water, which infects animals and humans. Septicemia, septic arthritis, pneumonia and soft-tissue infections are the clinical manifestations noted in case reports from India.^[15] The clinical presentation of neurological melioidosis in various reports include features of aseptic meningitis, brain abscesses, brainstem encephalitis with cranial nerve palsies and acute flaccid paralysis.^[3] Other CNS manifestations of melioidosis include, focal encephalitis, encephalomyelitis, cerebral abscesses, meningitis, and osteomyelitis of skull.^[16] Probable mechanism of neurological melioidosis is direct invasion of the

brain and spinal cord by the bacteria^[17] and an exotoxin-induced neurological syndrome, with profound neurological disease occurring in the absence of apparent direct infection of the CNS. Cerebrospinal fluid (CSF) analysis shows mononuclear pleocytosis mimicking tuberculosis. Culture in the Ashdown's medium, a gentamicin containing liquid transport broth grows *B. pseudomallei*.^[18] Treatment consists of IV ceftazidime (40 mg/kg) or imipenam (20 mg/kg) thrice daily for 10 days^[19] followed by trimethoprim/sulfamethoxazole (8/40 mg/kg) twice daily for 3-6 months.^[20]

In our patient as blood culture grew *B. pseudomallei*, the possibility of GBS secondary to melioidosis is considered. The disease could have been due to an exotoxin mediated neurological disease or the infection could have triggered an autoimmune process. However, an alternate hypothesis of a coexistent *B. pseudomallei* infection with an unrelated GBS cannot be ruled out. The rarity of melioidosis however argues against this latter hypothesis.

Conclusion

Infections with the many agents have been reported preceding GBS most common being viral infections. Rarely GBS may be a manifestation of neurological melioidosis, which is caused by the gram negative bacillus *B. pseudomallei*. The exact mechanism of pathogenesis is unclear. In patients with immune-compromised states like diabetes presenting with GBS-like picture and fever, uncommon infective causes such as melioidosis may also need to be considered for appropriate timely management.

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