up to 16% of patients with WG [9]. Others have found a higher prevalence of 23%, nevertheless noting that it occurs less frequently and is less severe than in the other ANCA-associated vasculitides [10].

In one descriptive study of 128 cases of Wegener granulomatosis, patients who developed PNS manifestations were more often males, had a significantly older age at onset of WG (53 versus 44 years-old), a larger disease extent and showed higher titers of ANCA antibodies [11].

Our patient developed an acute motor axonal neuropathy (AMAN), which is the axonal form of Guillain Barré syndrome (GBS).

It has been stated that AMAN is rare in Western countries, but not uncommon in Eastern Asia [12]. A recent study evaluating the geographic variations of GBS, found a higher prevalence in Bangladesh for both the axonal subtype and pure motor clinical form of GBS, as compared to European/American and Asian countries [13]. AMAN is also known to occur more frequently than the classic demyelinating form of GBS in China, especially during yearly summer outbreaks of GBS [14].

The clinical picture of AMAN is similar to that of classic GBS, however muscle atrophy becomes evident relatively early in the axonal form [8]. Also, patients with AMAN tend to have more severe symptoms at onset, but the general prognosis does not differ significantly between the two [15].

Development of AMAN has been associated with antibodies against gangliosides GM1, GD1a, GalNac-GD1a and GD1b [16]. Infectious agents, especially *Campylobacter jejuni*, *Cytomegalovirus* or *Haemophilus influenzae* are recognized as potential triggers (through the process of molecular mimicry), by expressing lipooligosaccharides on their surface. These are very similar to the gangliosides found in peripheral nerves and are subsequently targeted by the host's immune system [17].

Regarding therapy, vasculitis neuropathies usually require immunosuppressive treatment of the underlying disorder. Regimens usually begin with corticotherapy. Combined treatment with other immunosuppressive agents, such as cyclophosphamide or rituximab, is useful in refractory or fulminant cases and may also help reduce the dose of corticosteroids [2,8].

CONCLUSION

In our patient, intravenous immunoglobulins were required, as would have been the treatment of any acute inflammatory polyneuropathy.

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