Commentary

Guillain-Barré syndrome (GBS) and posterior reversible encephalopathy syndrome (PRES) are rare conditions and their co-occurrence in a small number of cases does suggest that there may be an etiological link between the two conditions, especially when there is a close temporal relationship. Reaching the correct diagnosis of either of these disorders in remote practice areas without specialist electrophysiological and neuroradiological support raises considerable challenges. Since prompt diagnosis and intervention for both conditions is likely to improve the prospects of good recovery, early recognition is particularly important. The current clinical case by Basavaraj *et al*. in this issue of the journal lucidly describes such a scenario and highlights pertinent diagnostic and management issues.[1]

PRES is a descriptive term for a clinical constellation of neurological abnormalities generally including

headache, visual loss, nausea, confusion and seizures, underpinned by characteristic radiological features of posterior signal hyperintensities on T2-weighted magnetic resonance imaging (MRI).[2] The widespread advent of MRI accounts for its increased recognition; indeed in the absence of MRI it would be hard to establish the diagnosis with certainty. Whilst systemic hypertension is characteristic, it is not always present, adding further diagnostic doubt in the absence of imaging. One can readily imagine the clinical uncertainties and a wide differential diagnosis when faced with a patient concurrently manifesting features of both GBS and PRES, in the absence of any supportive investigations. Equally, access to diagnostic electrophysiology and radiology rapidly pinpoint the problem. Recognizing that the two diseases can be associated is clearly the first step in the right direction to establishing a diagnosis, hence the value of case reports such as this.

GBS is principally characterized by an acute symmetrical ascending paralysis with some degree of somatic sensory involvement. Autonomic dysfunction is a relatively common but frequently over-looked complication of the disease, manifesting as sympathetic or parasympathetic over-activity, or deficiency, or a combination of both.[3]

PRES has been described in association with a very wide range of disorders including eclampsia, systemic lupus erythematosus, use of immunosuppressive and cytotoxic drugs and renal failure with hypertension. The pathophysiology is not precisely known, but it is thought that rapid rises in blood pressure overcomes the normal autoregulation of blood supply to the brain resulting in dilation and leakage from arterioles, which results in vasogenic edema, primarily of the white matter. How this relates to GBS is speculative, but might concern the autonomic dysfunction often seen in GBS.

There are numerous published case reports highlighting the association between GBS and PRES. In this issue of the Journal of Rural Neurosciences in Rural Practice, another such patient is described who presented somewhat unusually with features of acute hypertensive encephalopathy and subsequently developed flaccid tetraparesis. The MRI findings were consistent with PRES and the nerve conduction studies showed features of demyelination, leading to a diagnosis of GBS. PRES has been described in patients with GBS treated with intravenous immunoglobulin (IVIg), leading authors to conclude that PRES might be a complication of IVIg, rather than GBS itself. The first such case was reported by Voltz *et al*. in 1996 in which a patient with GBS developed clinical and radiological features consistent with PRES 3 days after completing a course of IVIg treatment.[4] PRES has also been reported in patients who were treated with IVIg for the diseases other than GBS. However, all these patients were also known to have clear risk-factors for development of PRES such as renal failure and chemotherapy.[5] There are also several reports of patients developing PRES before the onset of GBS or IVIg treatment. These cases all had sustained hypertension at the time of presentation followed by marked fluctuation in the blood pressure, which was thought to be due to underlying dysautonomia.[6-8] Any patient who presents with PRES in the absence of any obvious etiological factor should prompt vigilance toward monitoring for the development of GBS.

This case and others highlight three particularly interesting features of the association between GBS and PRES. At first, the temporal relationship is generally ordered such that PRES onset follows GBS; however, as seen in this case the evolution of clinical presentation of both disorders can be concurrent. Secondly, previous reports have highlighted a temporal relationship in which the onset of PRES follows IVIg therapy; in the current case and some other reports PRES onset clearly preceded IVIg therapy. Therefore, IVIg could be considered an additive risk-factor for the development of PRES in GBS; it clearly cannot be the sole driving force. This suggests that IVIg can be safely used to treat patients who develop PRES and GBS. Thirdly, they highlight the fact that acute hypertension as a result of dysautonomia in the setting of GBS acts as a possible risk-factor for PRES.

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