Guillain-Barre syndrome and posterior reversible encephalopathy syndrome: two cases of a rare association

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Background

Posterior Reversible Encephalopathy Syndrome (PRES) and Guillain Barrè syndrome (GBS) are rare and potentially life-threatening neurological conditions that require a prompt diagnosis for a better prognosis.

Case series

An hypertensive, diabetic and cardiopatic 78-year-old man presented to the emergency department for ongoing seizures. He presented gait disturbance with frequent falls in the previous week. Brain MRI showed a picture suggestive of PRES (Fig.1 A, B). Aggressive i.v. antihypertensive and antiseizure therapy were promptly started. The next day a rapidly progressive strength deficit in the 4 limbs became evident. Electroneurographic examination (ENG) documented an acute inflammatory demyelinating polyneuropathy (AIDP), therefore treatment with i.v. immunoglobulins (Ig) was started. Clinical picture rapidly worsened with respiratory failure and the need for orotracheal intubation in intensive care unit. At the end of Ig treatment, extubation was possible and the patient showed a progressive recovery of the strength deficit.

The second case is 60-year-old woman who presented with balance disorder and paresthesias in the feet and a mild flu-like syndrome in the previous week. Lumbar puncture revealed normal cell count and protein value, with negative film-array. ENG was normally, but despite this, a trial of i.v. Ig was started. In the next 36 hours the clinical condition worsened with dry mouth, severe tetraparesis, bilateral ophthalmoplegia and VII, IX, and X nerve palsies. She was ataxic and hyporeflexic with power of 3/5 in all limbs. Brain MRI showed a picture suggestive of PRES with left parieto-occipital subarachnoid hemorrhage (Fig. 1 C, D). She was also investigated for botulism as she had eaten homemade aubergines in oil, but a sample of stool and food was negative for Clostridium botulinum toxin. ENG after 5 days was suggestive for AIDP. Patient was transferred in the Rehabilitation ward and progressively recovered.

Conclusion

Co-occurrence of GBS and PRES has been rarely reported in literature, but the relationship between them is not better explained. PRES could be considered a consequence of GBS dysautonomia or Ig treatment, but a pathogenetic explanation when PRES onset precedes the acute radiculopathy has not been found yet. A continuum between these conditions could be hypothesized.

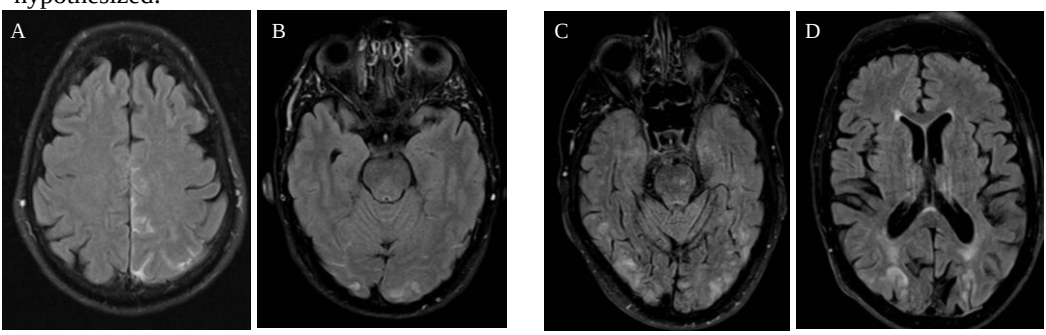

Fig.1 Brain MRI of patient 1 (A, B) and patient 2 (C, D) showing a picture typical and suggestive of PRES.

Reference

1. *Nabi S et al., BMJ Case Rep. 2016. ; 2. Joshi S et al., BMJ Case Rep. 2020; 3. Belgrado E et al., Front Neurol. 2022.*