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Letters to the editor

OVERVIEW

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Transverse myelitis: a diagnostic challenge

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Editor – We read with interest the article published by Notghi and colleagues.[1] The authors described a case of transverse myelitis, with symptoms manifesting 7 days post-administration of first dose of the AstraZeneca COVID-19 vaccination. The patient had a history of inactive pulmonary sarcoidosis. Due to the temporal association with the vaccine, lack of evidence supportive of active sarcoidosis on systemic imaging and the poor response to steroid therapy, the authors postulated that the myelitis is post-vaccination rather than a vaccine triggered relapse of isolated neurosarcoidosis. Due to major implications, distinction is crucial.

Post-vaccination immune-mediated response has been implicated in the pathogenesis of several neurological conditions including Guillain–Barré syndrome, acute demyelinating encephalomyelitis and transverse myelitis.[2] Similarly, vaccine administration may trigger a flare-up of pre-existing inflammatory conditions. Recent reports have described a role of vaccination in triggering a relapse of chronic immune-mediated conditions including rheumatological diseases, minimal change disease and microscopic polyangiitis.[3–5] Taking this point into account would make the temporal link not useful in discriminating between a purely vaccine-induced disease and a vaccine triggered relapse of inactive disease.

From reading the article, we have also noted that the authors felt that lack of active systemic features of sarcoidosis make neurosarcoidosis unlikely. While patients with neurosarcoidosis often have other systems involvement, several studies have reported isolated neurosarcoidosis.[6] We have also noted that some crucial investigations have not been performed in this case, including cerebrospinal fluid (CSF) angiotensin converting enzyme (ACE) and CD4/CD8 ratio. Although insensitive, CSF ACE may be reasonably specific for neurosarcoidosis.[7] Additionally, combined elevation of CD4/CD8 ratio and CSF lymphocytosis may provide a specificity of 95% for neurosarcoidosis.[8] Measuring CSF interleukins may also play a useful role in this clinical context. Finally, the observed clinical improvement is likely due to steroids starting to take effect rather than plasma exchange that would not be expected to exert a theraputic response by day 2 of the treatment cycle.

We suggest close monitoring of this category of patients for features suggestive of re-emergence of quiescent disease, and we strongly advocate discussing such complex cases at specialised neuro-inflammatory multidisciplinary meetings to guide further management. ■

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Haemophagocytic lymphohistiocytosis in pregnancy

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Editor – The annual confidential enquiries into maternal mortality in the UK, published by MBBRACE, state that we should 'Treat pregnant and postpartum women the same as non-pregnant women unless there is a clear reason not to'.[1] Consecutive MBRRACE reports include numerous maternal deaths occurring after standard of care treatment was withheld or delayed due to concerns about its safety in pregnancy. In the paper by Jha *et al*, the authors state that 'the safety and efficacy of … cyclosporin, rituximab and immunoglobulin during pregnancy and lactation are yet to be established.[2] Evidence-based statements supporting the use in pregnancy of cyclosporin, intravenous immunoglobulins and, for life-threatening maternal disease, rituximab

have been produced by the American College of Rheumatology, European League Against Rheumatism and British Society for Rheumatology.[3–5] Therefore, these treatments should not be withheld from pregnant women due to concerns about fetal harm. This is particularly important for women with severe maternal disease such as haemophagocytic lymphohistiocytosis (HLH), as untreated disease could be harmful to both mother and baby.

The HLH label should be viewed as we see sepsis, as a starting point to initiate management while striving to identify an underlying source/trigger. The immunological changes of pregnancy can trigger HLH, with delivery being curative, but HLH in pregnancy can also occur secondary to infection, autoimmunity/autoinflammation and haematological malignancy.[6] Jha *et al* state that 'Biopsy of liver, spleen or lymph nodes could not have revealed any extra information to change the diagnosis', but we would advise caution with this assumption.[2] Steroids are often employed early in HLH to control hyperinflammation but can mask or delay the identification of triggers such as lymphoma.[7] Our practice is to undertake repeated histological examination of abnormal organs, lymph nodes and bone marrow alongside the use of steroid sparing agents, where possible. Managing a pregnant woman with HLH is best done within a multidisciplinary team (MDT) setting, including rheumatology, haematology, infectious diseases, obstetrics and obstetric medicine and with advice from an HLH MDT, in order to best judge the timing of investigations and delivery. ■

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Conflicts of interest

Taryn Youngstein is on industry-sponsored advisory boards for Sobi, Novartis and Roche. Ian Giles is a speaker, has advisory board fees and an unrestricted research grant from UCB Pharma.

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