GvHDHub

aGvHD, cGvHD

A review of CNS involvement in GvHD from case reports and published literature

Sara Valente | Jul 23, 2018

In October 2017, findings from reported cases and a literature review on CNS involvement in graft-versus-host-disease (GvHD) were published in *Medicine* by Mathilde Ruggiu, Saint-Louis Hospital, France, and colleagues. The authors aimed to review data from reports of CNS-related GvHD in order to better understand this rare complication of allogenic hematopoietic stem cell transplantations (allo-HSCT).

Overview

- 7 cases of CNS GvHD were reported from the Saint-Louis Hospital and 32 cases were found from a PubMed literature review between 1990 and December 2016
- Patients included in the review had received allo-HSCT and 34 were transplanted for hematologic malignancies and 5 were transplanted for non-malignant hematopoiesis disorders
- Reports of CNS symptoms were associated with biological or imaging abnormalities that had no other diagnosis. The authors noted that in order for chronic CNS GvHD to be reported, according to the Consensus Conference, there must be an occurrence of chronic GvHD affecting other organs. They reported that no diagnosis criteria were documented in the literature for acute GvHD

Key Findings

- Median onset of symptoms was 385 days after allo-HSCT (range, 7–7320)
    - Median onset of neurological symptoms for patients without chronic GvHD was 81.5 days (range, 7–1095) and for patients with chronic GvHD was 549 days (range, 11–7300), $P = 0.001$
- Clinical features reported included; encephalitis (n = 14) stroke-like episodes (n = 7), lacunar syndrome (n = 3), multiple sclerosis-like presentation (n = 7), acute demyelinating encephalomyelitis-like presentation (n = 4), mass syndrome (n = 1) and non-specific (n = 3)
- Histological data in 17 patients were obtained from biopsies and reported; neurological vasculitis (n = 7), demyelinating lesions (n = 5), immune-mediated encephalitis (n = 5) and non-caseating granuloma (n = 1)
- A significant number of patients experienced encephalitis more frequently if they had early involvement of CNS after allo-HSCT and no chronic GvHD symptoms, $P = 0.07$
- 34 patients with CNS-GvHD were treated with immunosuppressive therapy, including corticosteroids (n = 31).
    - Other treatments included intravenous immunoglobulin, plasmapheresis, cyclophosphamide, calcineurin inhibitors, mycophenolic acid, methotrexate and etoposide
    - 10 patients achieved a complete response and 15 patients achieved a partial response

The authors concluded that CNS involvement in GvHD is rare and difficult to diagnose but due to the findings of this study there may be evidence that GvHD targets the CNS. The authors recommended that magnetic resonance imaging and cerebral spinal fluid analysis is conducted to rule out other causes of the effects of the CNS disorders. They reported that despite good response to the immunosuppressive therapy given as a treatment for CNS GvHD, it is associated with a poor prognosis.

References

Ruggiu M. *Et al*. Case report: Central nervous system involvement of human graft versus host disease: Report of 7 cases and a review of literature. *Medicine* (Baltimore). 2017 Oct;96(42):e8303. doi: 10.1097/MD.0000000000008303