Pediatric Rheumatology

Poster presentation

Circulating endothelial cells and endothelial progenitor cells in childhood primary angiitis of the central nervous system

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from 15th Paediatric Rheumatology European Society (PreS) Congress
London, UK. 14–17 September 2008

Published: 15 September 2008

Pediatric Rheumatology 2008, **6**(Suppl 1):P276 doi:10.1186/1546-0096-6-S1-P276

This abstract is available from: http://www.ped-rheum.com/content/6/S1/P276

Background

Primary angiitis of the central nervous system in children (cPACNS) is an inflammatory vasculitis that solely affects the CNS vessels in the absence of a systemic inflammatory process. Circulating endothelial cells (CECs) are increasingly described as biomarkers for tracking vascular injury [1]. Additionally, bone marrow-derived endothelial progenitor cells (EPCs) are thought to play a pivotal role in the regeneration of damaged endothelium. We describe the relationship of CECs and EPCs to clinical and/or radiological disease progression in cPACNS.

Materials and methods

16 children, median age 7 years old (range 1.8–17); 9 males with cPACNS were studied. Two groups were identified, according to radiological and/or clinical progression, or non progression at >6 months from diagnosis. CECs were isolated from whole blood using immunomagnetic bead extraction. EPCs were detected using flow cytometry and were defined as mononuclear cells triple positive for CD34/CD133/CD144 and CD34/CD133/VEGFR2.

Results

Median CEC count in progressive cPACNS was significantly raised to 480/ml (176–1152) compared to 36/ml (0–168) in non-progressive disease ($p = 0.0007$), 32/ml (0–152) in child control ($p = 0.0050$) and 24/ml (16–141) in patients with non inflammatory cerebrovascular pathology ($p = 0.0016$). CD34+CD133+CD144+ cells were significantly raised in patients with progressive disease compared to child controls ($p = 0.005$) and patients with non progressive disease ($p = 0.03$). There was a similar but non significant trend for EPCs expressing CD34/CD133/VEGFR2.

Conclusion

CECs can be used to track vascular injury due to cPACNS and differentiate progressive versus non-progressive cerebral vasculitis. We also demonstrated an increase in EPCs in progressive cPACNS, perhaps indicative of a compensatory reparative vasculogenic response.

References

1. Clarke LA, Shah V, Arrigoni F, Eleftheriou D, Hong Y, Halcox J, Klein NJ, Brogan PA: **Quantitative detection of circulating endothelial cells in vasculitis: comparison of flow cytometry and immunomagnetic bead extraction.** *J Thromb Haemost* 2008, **6:**1025-1032.