Pediatric Rheumatology

Poster presentation

Infantile Onset Panniculitis with Uveitis and Systemic Granulomatosis: immunohistochemical findings

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Infantile-Onset Panniculitis with Systemic Granulomatosis is a recently described clinicopathologic entity, considered part of the spectrum of pediatric granulomatous inflammatory diseases.

Through the International Registry of Pediatric Granulomatous Arthritis (PGA), we now identified 5 children with this disorder, all manifesting from very young age panniculitis, fever, hepatosplenomegaly, arthritis, uveitis and acute phase response. Underlying infections, immune deficiency and autoimmune disease, were excluded. No *CARD15* or *CIAS1* mutations were found. Histologically, the subcutaneous nodules showed a non-vasculitic non-cytophagic lobular panniculits. Giant and epitheloid cell granulomas were found in liver (pt 1, 5), synovium (pt 2), lymph node (pt 3, 5), colon (pt 3), subcutaneous fat (pt 3), dermis and lung (pt 4). Immunohistochemical study of the granulomas revealed the presence of abundant CD68+ macrophages, numerous CD4+ T lymphocytes and few CD8+ cells. TNF stainings were only weakly positive, conversely abundant IL-6 staining was apparent, especially in the corona of lymphocytes.

Despite steroid and cyclosporin treatment, the course was progressive in patient 1 with severe lung involvement and death from respiratory insufficiency at age 14. In patients 2, 3, 4 and 5, therapy with anti-TNF MoAbs allowed better disease control.

Infantile onset Panniculitis with Systemic Granulomatosis may be a potentially fatal granulomatous disease in children. Although the response to anti-TNF MoAbs in four of our patients is of note, the paucity of TNF with overwhelming presence of IL-6 *in situ* in granulomas suggest the implication of alternative immune-inflammatory pathways. The role of the Th17 pthway is currently under investigation