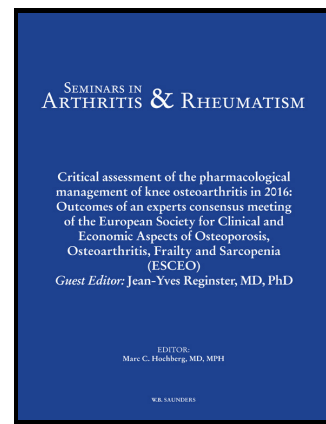


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Intra-cranial Granulomatous Disease in Common Variable Immunodeficiency: Case Series and Review of the Literature

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Abstract:

Background/Purpose:

Common variable immunodeficiency (CVID) is typically characterized by hypogammaglobulinemia and often but not always recurrent infections. Paradoxically, 8-22% of patients with CVID develop granulomatous disease. Granulomata have been described in many organs including the lungs, skin, liver, spleen, kidneys, eyes, lymph nodes and intestines. Data about central nervous system (CNS) involvement in CVID are extremely rare. We aim to describe a case series and include an extensive literature review of CNS involvement in CVID to understand the different features and patterns of the disease.

Methods:

We searched the English Pubmed database for relevant articles between 1950 and 2014 using the Key Words "common variable immunodeficiency", "granulomatous disease", "brain", "sarcoidosis", and "sarcoid-like syndrome". Data from all case series, surveys, systematic reviews, and individual case reports, as well as retrospective studies were extracted. A total of 15 patients were reported in the literature. We combined our experience with four additional patients from The Cleveland Clinic between 2009 and 2014. Demographics, clinical features, laboratory and imaging findings, treatment and follow-up were extracted for the 19 patients and summarized descriptively.

Results:

Female sex and Caucasian race represented 63.2% (12/19), and 80% of the patients respectively. The mean age of CVID diagnosis was 24 years; mean age when the CNS disease was diagnosed was 21.5 years. 68.4% of the patients (13/19) had granulomas involving ≥ 2 organs including the central nervous system, 31.6% (6/19) had CNS granulomas only. Associated granulomatous diseases occurred in lungs (72.7%), lymph nodes (27.2%), spleen (27.2%), eyes (18.1%), liver (18.1%), parotid glands (9%), and skin (9%).

Fifty-three percent (10/19) of the patients had documented recurrent infections, all of them being upper respiratory tract infections.

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