Autoimmune disease

Autoimmune diseases arise from an abnormal immune response of the body against substances and tissues normally present in the body (autoimmunity.) This may be restricted to certain organs (e.g. in autoimmune thyroiditis) or involve a particular tissue in different places (e.g. Goodpasture's disease which may affect the basement membrane in both the lung and the kidney).

The treatment of autoimmune diseases is typically with immunosuppression—medication that decreases the immune response.

A large number of autoimmune diseases are recognized. A major understanding of the underlying pathophysiology of autoimmune diseases has been the application of genome wide association scans that have identified a striking degree of genetic sharing among the autoimmune diseases.

Many diseases can present as tumefactive lesions and mimic neoplastic lesions.

Torres et al., aimed to determine the frequency of pseudotumoral CNS lesions referred to an Oncology center and the frequency of the tumor mimickers

In a retrospective study at the National Institute of Cancer, Rio de Janeiro, Brazil. Medical charts of patients admitted to the Neurosurgery and Pediatrics services from 2007 to 2011 were reviewed. Clinical and radiological features of cases initially diagnosed with primary CNS tumors but received a final diagnosis of pseudotumoral disease were recorded.

Among 891 patients referred as primary brain tumors, 38 cases had pseudotumoral lesions (4.3%). Most were adults (63%), with mean age of 29.4 years, and women (60.5%). Most frequent symptoms were headache (28.9%), motor signs (23.7%) and seizures (15.8%). Mean time from initial symptoms to diagnosis was 12.2 months. Lesions were single in 84.2% of patients, had contrast enhancement in 45.6% and surrounding edema in 17.4%. Twenty patients (52,6%) underwent biopsy. Systemic autoimmune diseases were the most frequent etiologies (28.9%), followed by idiopathic inflammatory demyelinating diseases, infections and vascular abnormalities (15.8% each). Good outcome with no major deficits was observed in 60.5% cases.

The frequency of pseudotumoral lesions in an oncology reference center was low. Young women were most affected, and lesions were associated more frequently to systemic autoimmune diseases. Prompt recognition is important to avoid unnecessary treatment since most patients had a good outcome [1)].

A study indicated a higher overall autoimmune disease prevalence in unilateral than in bilateral Moyamoya disease (MMD). Unilateral MMD may be more associated with autoimmune disease than bilateral MMD. Different pathogenetic mechanisms may underlie moyamoya vessel formation in unilateral and bilateral MMD [2)].

Lymphocytic hypophysitis is a relatively rare autoimmune disease defined by lymphocytic infiltration to the pituitary.

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Torres YC, Alves-Leon SV, Lima MA. Frequency of pseudotumoral CNS lesions in an oncology center. World Neurosurg. 2019 Jun 19. pii: S1878-8750(19)31635-3. doi: 10.1016/j.wneu.2019.06.083. [Epub ahead of print] PubMed PMID: 31228702.
2)
Chen JB, Liu Y, Zhou LX, Sun H, He M, You C. Increased prevalence of autoimmune disease in patients with unilateral compared with bilateral moyamoya disease. J Neurosurg. 2015 Sep 25:1-6. [Epub ahead of print] PubMed PMID: 26406790.