Abstract: PB1953

Title: TWELVE CASES OF MULTICENTRIC CASTLEMAN DISEASE WITH ELEVATED SERUM IGG4 LEVELS AND INFILTRATING IGG4 POSITIVE PLASMA CELLS PATHOLOGICALLY FROM A SINGLE CENTER.

Abstract Type: Publication Only

Session Title: Chronic lymphocytic leukemia and related disorders - Clinical

Background:

Multicentric Castleman's disease (MCD), classified as Tumor-like lesions with B-cell predominance, is a rare systemic and progressive disease with lymphadenopathy involved multiple lymph nodes. Patients presenting elevation of serum IgG4 levels and infiltrating IgG4 positive plasma cells in lesions should be carefully differentiated from IgG4-related disease (IgG4-RD) in diagnosing MCD.

Aims:

This study aimed to describe a 12 patients' cohort with the diagnosis of MCD, which could also fulfil the comprehensive diagnostic criteria for IgG4-RD.

Methods:

A retrospective medical record review was conducted on all patients diagnosed with MCD in our hospital from January 2010 to December 2022. Inclusion criteria of the cohort: (1) serology: IgG4 concentration> 1350 mg/L, and (2) pathology: number of IgG4+ cells/high-power field ≥10. Clinical manifestations, laboratory examinations, pathological evidence, and treatment details of the cohort were provided.

Results:

Twelve MCD patients were identified, 3 (25%) females and 9 (75%) males. Median age of diagnosis was 46.5 years (range, 21 - 72 years) with 10 (83.3%) patients being younger than age of 60 years. The most frequent manifestation was recurrent fever (temperature > 38°C) presented in 5 (41.7%) patients; other symptoms included night sweats (25%), weight loss (16.7%), and fatigue (66.7%). Violaceous papules were investigated in 2 (16.7%) patients. Anemia (hemoglobin <100 g/L) was found in 11 (91.7%) patients, thrombocytopenia/thrombocytosis (platelet count <150 k/mL or >400 k/mL) in 8 (66.7%), hypoalbuminemia (albumin <3.5 g/dL) in 10 (83.3%), proteinuria (total protein >10 mg/100 ml) in 3 (25%), elevated C-reactive protein (>10 mg/L) in 8 (66.7%) and interleukin-6 in 11 (91.7%). The median serum IgG4 level was 5 585 mg/L (range 1 690 – 33 000 mg/L). Pathological diagnosis of IgG4-RD preferred were presented in 5 (41.7%) patients. Glucocorticoids monotherapy was given in 5 (41.7%) patients before they diagnosed with MCD. Three (25%) patients received Tocilizumab therapy for MCD. Other therapies consisted of TD, CTX + dexamethasone, TCP/TCD, Rituximab and BD. The median follow-up of all patients was 19.8 (range, 1.47-84.13) months. All patient were alive at the time of data collection.

Summary/Conclusion:

While pathological diagnosis was vague, clinical manifestation and response to glucocorticoids monotherapy were valuable for differentiated from IgG4-related disease (IgG4-RD) in diagnosing MCD. In this small cohort, most MCD manifested anemia (91.7%) and elevated interleukin-6 (91.7%). The treatment for MCD was heterogeneous with a favorable outcome in this study.

Keywords: IL-6, Clinical data, Castleman's disease