Cytomegalovirus Retinitis in a Patient with Waldenstrom Macroglobulinemia

Felipe Tavares Rodrigues¹, Alexandre de Carvalho Paiva²
Catherine da Cal Valdez², Cleonice Alves de Melo Bento³, Regis Mariano de Andrade³

¹Graduate Student of Escola de Medicina e Cirurgia do Rio de Janeiro - UNIRIO.
²Physician at Hospital Universitário Gafré e Guinle - UNIRIO.
³Professor of Universidade Federal do Estado do Rio de Janeiro – UNIRIO, Brazil.

A 62-years-old woman presented with microcytic hypochromic anemia, diffuse adenopathy, and hepatosplenomegaly, was diagnosed as having Waldenstrom Macroglobulinemia by blood serum electrophoresis which detected a IgM-KAPPA monoclonal gammopathy, besides the results of immunophenotypic characteristics of neoplastic B-lymphocytes present in bone marrow positive for CD19, CD20, FMC7, CD79; and not reactive for CD23, CD5 and CD38. MYD88 mutation was absent. The patient underwent a week intermittent chemotherapy (oral scheme: Prednisone 60 mg and Chlorambucil 2 mg during 5 months) and then, without adequate therapeutic response, it was reset including intravenous anti-CD20 monoclonal immunoglobulin (Rituximab, for 4 months). The patient had a good response to the therapy. However, almost a year after Rituximab scheme started, patient complained of sudden decrease of visual acuity (20/200) according to the Snellen chart with floaters, only in the left eye. CD4 T cell count was 205 cells/mm³, relatively reduced when compared to a health person because of the chronic use of immunosuppressant drugs and the down modulation due to the inflammatory stress state seen at a hematological lymphoproliferative disease. Fundoscopy findings were compatible with CMV retinitis (Figure 1), confirmed by CMV-DNA PCR in plasma samples with a high viremia (3.10² copies/mL). Plasma cytokines were measured to see the inflammatory response (Figure 2).

Intravenous Ganciclovir 500mg every 12 hours for 2 weeks was administered. Granulocyte colony-stimulating factor was used to manage neutropenia. Visual acuity improvement was observed (20/40). Drugs only suppress viral replication, not eliminating virus, in some cases of advanced retinitis and mainly when detection is delayed, retina impairment is irreversible [2]. CMV retinitis can seldom occur in those who have undergone chemotherapy treatment to transplantations, autoimmune diseases and malignancies [3].

To our knowledge, this is the first case of CMV retinitis in a patient with WM, after Rituximab use.

Informed consent: Informed consent was obtained from all individual participants included in the study.
Figure 1: The “frosted branch” showing white, fluffy, cotton irregular lesions surrounding the retinal vessels.

Figure 2: The cytokines patient’s profile. We can see all the disruption of immune system homeostasis. INF-alfa is related to innate response to virus and cancer. INF-gama is related to Th1 lymphocyte phenotype and adaptative response to virus and cancer. IL-17 is known to participate in autoimmune diseases mediating Th17 formation. IL-1B, IL-6 and TNF-alfa are the triad inflammatory cytokines. The increasement of IL-10 is an attempt to control inflammatory process.

REFERENCES

