Siltuximab treatment in a Patient with multicentric Castleman’s disease

D. Maltezas, E. Evmorfiadis, N. Koufos, P. Repousis

Metropolitan Hospital, Pireaus, Greece

Case description: A 59-year-old male received surgery for a right inguinal hernia. An enlarged lymph node was removed during the surgical procedure and the biopsy revealed mixed type Castleman’s disease. 10 months earlier he had been treated with antibiotics because of fever and pleural effusion of the right lung. 3 months before surgery he had noticed enlarged lymph nodes in the right inguinal region. The patient was then referred to the hematology department for further diagnostic and treatment. A PET-CT scan revealed diffuse lymphadenopathy on both sides of the diaphragm along with a lesion of high SUV in the right pulmonary lobe. A biopsy of this lesion was performed showing non-specific inflammatory, benign findings. Bone marrow biopsy was without relevant findings. Laboratory tests were negative for HHV-8 and HIV. The patient fulfilled two major (histological findings, lymphadenopathy) and two minor (elevated ESR, polyclonal hypergammaglobulinemia) Castleman’s disease diagnostic criteria, establishing the diagnosis. We initiated treatment with Siltuximab, a monoclonal antibody indicated for the treatment of adult patients with multicentric Castleman’s disease who are negative for HIV and HHV-8. Dosage was 11 mg/kg every three weeks. The patient tolerated treatment without any side effects. After 5 cycles of treatment the patient is presenting a significant decrease of the lymph nodes’ size and improvement of the initially abnormal laboratory test values.

Discussion: Castleman’s disease is a rare benign lymphoproliferative disorder with different subtypes. The clinical and histological findings may vary substantially, imitating other malignant or benign diseases. Therefore, the diagnosis can be quite challenging. Our patient fulfilled the specific diagnostic criteria. In the disease severity evaluation, the patient was rather categorized in the non-severe group. The presence of the pulmonary lesion could have been interpreted as pulmonary involvement, however there were no specific findings related to Castleman’s disease in the biopsy performed. Treatment with Siltuximab was chosen, as it is the first line treatment for non-severe disease. After 5 cycles of treatment the patient has shown significant improvement. A new PET-CT scan is scheduled after the 6th cycle to further evaluate response and decide about future course.

Conclusion: Castleman’s disease is challenging to diagnosis and treat. A individual approach is necessary taking under consideration each patient’s situation and needs in order to make the best choice from the available treatment options.