RARE ASSOCIATION BETWEEN HEMOPHAGOCYTIC HISTIOCYTOSIS AND A LYMPHOPROLIFERATIVE SYNDROME.

Laura Sofroni[1], Doramina Ghiorghiu[1], Cristina Burcoveanu[1], Angela Dascalescu[2], C. Danaila[2]
1 Regional Institute of Oncology,
2 UMF "Gr.T.Popa"Iasi

Hemophagocytic lymphohistiocytosis is a syndrome of pathologic immune activation characterized by clinical signs and symptoms of extreme inflammation. It is an aggressive, life threatening disease that frequently affects children and young adults, but can occur at any age. It is known in two forms: primitive, which is due to a genetic defect and secondary, which appears as a pathological manifestation that occurs secondary to other diseases (viral infection, autoimmune disease, malignant proliferation - T lymphomas, acute leukemias). The pathogenetic mechanism consists of an uncontrolled proliferation of macrophages, explained by the decreased functionality of NK lymphocytes and cytotoxic T lymphocytes (CD8 +) caused in their turn by mutations in the gene encoding perforin synthesis.
This paper presents the case of a 79 years old female who submitted to the hospital with febrile syndrome occurred 6 weeks prior to the submission. The following clinical and biological investigations excluded a possible infectious process, raising the suspicion of autoimmune hepatitis, and she was directed to an infectious disease clinic, where they were all removed: meningitis, endocarditis, a viral infection, autoimmune hepatitis (all this time the patient was febrile under extended spectrum antibiotics with negative urine cultures and hemocultures). A positive Quantiferon test indicated a possible tuberculosis, suspicion that was ruled out in a pneumology hospital.
Meanwhile we noticed a decrease in haematological constants (that were near normal initially) to the occurrence of cytopenias. In the haematology clinic, following the lack of response to broad-spectrum antibiotics, and the thorough investigations conducted, which excluded other pathology, the suspicion of hemophagocytic histiocytosis occurred, later confirmed by the presence of 5 criteria out of 8, sufficient for the diagnosis according to guidelines HLH 2004. We initiated specialized treatment, resulting in remission of febrile syndrome, improvement of general health and haematological constants. Thereafter, osteomedulara biopsy result was suggestive for follicular lymphoma.
In conclusion, prompt initiation of chemotherapy is essential for survival, but an early diagnosis was a challenge because of the rarity of the disease, variable presentation and the time required to perform diagnostic testing.