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Hemophagocytic Lymphohistyocytosis associated with Hoghkin lymphoma

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Hemophagocytic lymphohistiocytosis (HLH) is an aggressive and life-threatening syndrome of excessive immune activation and tissue destruction due to abnormal immune activation. This is a hyper inflammatory/dysregulated immune state caused by the uncontrolled proliferation of activated lymphocytes and histiocytic with massive release of inflammatory cytokines. Secondary HLH is associated with, and thought to be emerged by malignant and non-malignant diseases. Malignant disorders associated with secondary HLH include lymphoma, acute lymphocytic leukemia, acute myeloid leukemia, and syndrome. Many physicians in most cases make delayed diagnosis of HLH. HLH-associated cHL is an uncommon condition that can cause severe systemic symptoms acting as the perfect mimic of septic shock, deviating the clinical eye toward treating with antibiotics and not addressing in a timely manner the real aetiology of the patient's condition.

Patient, 45 years old male presented with tremor, weight loss, swollen fingers manifested 4 month ago and treated with NSAI. After 3 months the fever, weakness and fatigue were observed in the patient. The CT scan, biochemistry test, full blood count and electro neuro miography were performed. The examinations revealed mild lymphadenopathy (mediastinal, para aortal, para caval lymph nodes-1,5-1,8-2,0cm), splenomegaly (spleen sizes-16,5cm), Urea-20.22mmoml/L, Creatinine>500mmol/l, Anti-CCP>500U/ml, Rheumatoid factor 92.53IU/ml, C-reactive protein-302.63mg/L, GGT-102U/L, low WBC count-2.80x109/l, low PLT count-132.8x109/L. The patient suspected of having rheumatic disorders as he had a history of hereditary rheumatism (his mother suffered from rheumatism). The clinical diagnosis of periarteritis nodosa was suspected. The histological examination of tissue samples did not confirm the diagnosis of periarteritis nodosa. However, the collagenase wasn't been diagnosed on a basis of Anti-CCP results. Patient was sent to the nephrology department for nephrobiopsy. The renal disfunction was described as a complication of dehydration and use of NSAI and nephrobiopsy was not performed. Patient received Metylprednisolone 500mg per one day and 1000mg on the following 2 days after Metylprednisone of 24 mg was prescribed. After being treated with Corticosteroid and infusion therapy creatinine level went down, however on a second day of Metylpredisone administration the fever reappeared. Patients' general condition was getting worse, fever was persisted. The results of common blood count showed progressive pancytopenia.

The deterioration of liver function and coagulopathy were observed. Due to pancytopenia patient was hospitalized at Hematology Center. The X-Ray, USD examination revealed enlarged lymph nodes in axillar, cervical and supraclavicular zone and the spleen size of 15,5cm. Bone marrow biopsy was done, the aspirate was hypo cellular. Bone marrow immunophenotyping showed CD79a-2%, CD34-3%, CD14-14%, CD5-21%, CD3-50%, CD20-62.1%, CD15-65%, CD11c-66%. Ferritin level>2000ng/ml, Vitamin B12>1546pg/ml. EBVIgM-0.178, EBVIgG-0.774. The pancytopenia, splenomegaly, high ferritin level, fever let us suspect HLH. The triglycerides level was checked, the result was 3.8mmol/l (n=<3.0mmol/l), Fibrinogen level was 1.0g/l. No hemophagocytes were detected in bone marrow aspirate. Bone marrow biopsy with histological and immunohistochemistry examination was prescribed In order to confirm the diagnosis of acquired HLH the calculation score for HLH diagnostic criteria was used. 5 criteria from 8 were positive for HLH. The HLH was verified by the calculation score with the accuracy of 89%. HLH-94 protocol was started before getting histological and immunohistochemistry results. After starting corticosteroid therapy and first Etoposide administration the fever went down and patient's clinical results were improved. The immunohistochemistry revealed classic Hoghkin lymphoma mixed cellular subtype.

After receiving the immunohistochemistry results HLH-94 treatment protocol was switched to ABVD scheme. The dynamic improvement of patient's clinical condition and laboratory features were observed. After 4-th course the control bone marrow biopsy and CT were performed. No signs of disease were observed. Patient received total 6 courses of ABVD. Now the patient is in clinical hematological remission and under follow-up at Hematology center.

The long term follow up results will be provided in our second article where we will try to describe the possible pathogenesis mechanisms of the process.

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