P.26. **REGRESSION OF CARDIAC AMYLOID DEPOSITS AFTER CHEMOTHERAPY IN ONE PATIENT WITH AL AMYLOIDOSIS: A CASE REPORT.**

***Sorina Bădeliţă*[1], *Inga Botnariuc*[1], *Manuela Crişan*[1,2], *Monica Dogaru*[1], *Camelia Dobrea*[1,2], *Oana Diana Agatinei*[1] , *Alexandra Ghiaur*[1], *Daniel Coriu*[1,2]**

[1]Center of Hematology and Bone Marrow Transplantation, Fundeni Clinical Institute, Bucharest

[2]University of Medicine and Pharmacy "Carol Davila", Bucharest

Free light chain type of amyloidosis (AL amyloidosis) represents a neoplastic disease due to deposits of light chains as amyloid fibrils in different organs. The prognosis is poor and depends on the number and severity of organ involvment especially cardiac involvement. According to the published data, the presence of cardiac involvement at diagnosis and/ or enlarged interventricular sept (> 15mm) represents a poor prognosis factor with a median survival of 6 months.

We report a case of 57 year old woman admitted in March 2008 for progressive dyspnea, angor, oedema of legs and hyposalivation, symptoms with abrupt onset and evolution. The clinical examination revealed medium general status, no signs of orthostatic hypotension and ascendent paresthesia to both legs.
The lab results showed Hb=12.6 g/dl, Hct=37.7 %, Wbc=9100/mmc, Plt=282.000/mmc, (Segmented=74, Eosinophyls=1, Basophyls=1, Lymphocytes=18, Monocytes=6), no Jolly bodies; coagulation tests were in normal ranges with the exception of serum level of FX= 57%. The serum protein electrophoresis did not revealed any monoclonal component; serum levels of Ig were in normal ranges; free kappa= 16 mg/l, free lambda= 62.4 mg/l, raport free kappa/ lambda=0.25 (VN=0.26-1.65). serum protein immunofixation revealed compact band identified with anti- lambda light chain antiserum; proteinuria was 6 grams/24 hours. The bone marrow exam (aspiration and trephine): normocellular bone marrow with 5% lymfoplasmocytes infiltration. The abdominal fat tissue aspirate was Congo red positive. The cardiac assessment showed that there were ECG and cardioechogram signs of infiltrative cardiomiopathy (SIV= 17) NT-proBNP 781 pg/mls; EMG: sensitive polineuropathy; Fibroscan: F3 Metavir.

The clinical setting and lab results were suggestive for lambda light chain type primary amyloidosis cu multiple organ involvement (cardiac, renal, liver and peripheral nervous system).

The patient received 7 cycles of MP regimen and at the end of the treatment the lab results were Hb= 12 g/dl, Hct =36%, MCV= 102 fl Wbc=5370/mmc, Plt=106.000/mmc, (Segmented=55, Eosynophyle=2, Basophyle=1, Lymphocytes=32, Monocytes=10); raport free kappa/lambda= 1.48; cardioechography: diastolic disfunction type delayed relaxation, SIV= 12.5).

At 16 months after end of treatment, the patient presented progression of sensitive neuropathy. The patient received 5 cycles of MP regimen (Melphalan was given intravenously) with remission of neuropathy.
The evaluation from 2013 (after 6 years since diagnosis) showed: Hb=9.3 g/dl, Hct= 28.7 %, MCV=116 fl, Wbc=3200/mmc, Plt=50.000/mmc, , (Segmented=74, Eosinophyls=1, Lymphocytes=17, Monocytes=8), frequent macrocytes, granulocytes with hypersegmented nucleus.

Free kappa=14 mg/l, free lambda=11.1 mg/l, rapport free kappa/lambda =1.27; proteinuria was absent. The bone marrow exam (aspiration and trephine) showed normocellular marrow; granulocyte line was normal, hypogranular myelocites and neutrophils with pelger nucleus; hyperplasic erythrocyte line with basophilic megaloblasts; small size megakaryocytes.
Cardioechography: SIV=13, FE= 60%; Fibroscan: F3 Metavir;

The clinical setting was suggestive for late trilineal dysplasia due to chemotherapy and the lab tests confirmed the suspicion.

This case report ilustrates a case of lambda light chain type of systemic amyloidosis with important cardiac involvement which had a good response with intravenous Melphalan but in the same time, developed multilineal dysplasia as late side effect.

This work was supported by the grant CEEX 74/2006 from the Romanian Ministry of Research and Technology.