of kidney, heart, peripheral nerve , liver and other organs (digestive, bone marrow) were found in 28 (35%), 14 (17.5 %), 26 (32.5%), 4 (5%) and 8 (10%) cases, respectively. The stage of the disease was rated according to Mayo Stage 2012 obtaining the following results stage I: 25%, stage II: 25 % Stage III : 17.5% and stage IV: 42.5% The hematological responses (HR) were: CR 22.72%, VGPR 29.54%, PR 13.63%, SD 20.45%, NR (not evaluable) 13.63% The organ response was 50% (n = 22). The OS was 35 months (95% CI 4.2-65.7). The patients were classified according to first line treatments based on bortezomib: A) CyBordex (cyclophosphamide, bortezomib and dexamethasone, n = 23, B) VTD (bortezomib, thalidomide and dexamethasone) n = 2, C) VMP (bortezomib, melphalan and prednisone) n = 2, D) BD (bortezomib and dexamethasone) n = 12, and other regimens A) MP (melphalan and prednisone) n = 4 and B) CP (cyclophosphamide and prednisone) n = 1. Hematological responses (at least VGPR) of CyBordex, VTD, VMP, BD were of: 60.89%, 100%, 100% and 41.66% respectively; and organ response was : 47.82%, 100%, 25% and 41.66% respectively. The rate of premature death within the first six months was 22.72%. **Conclusions:** A retrospective study in which the majority of AL patients have an average age of 65 years, who are diagnosed in advanced stages and with a small number of cases that are HSCT candidates. It should be noted that almost the same number of responses were achieved from the organ response as well as from the HR (including CR + VGPR): 50%. Regimens based on bortezomib in newly diagnosed AL amyloidosis show results that seem comparable to those previously published in the literature. To improve these outcomes it is necessary to define the role of proteasome inhibitors the role of new therapies. It's still yet to define the role of proteasome inhibitors in the first line in combination with new molecules in order to improve responses. Educational interventions are important.

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Other Plasma Cell Disorders and Amyloidosis

SP-223

MGCS : an IgG-lambda monoclonal gammopathy associated with renal failure, lung disease and cutis laxa

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Renal diseases are frequently associated with monoclonal gammopathies. Heavy chain deposition disease (HCDD) is an extremely rare condition, one of the 3 entities of monoclonal immunoglobulin deposition disease (MIDD). It is characterized by the presence of nodular glomerulosclerosis and glomerular and tubular deposition of monoclonal HC without associated light chains (LC). Here, we reported a case of HCDD associated with other manifestations, e.a. pulmonary fibrosis and cutis laxa. A 38-year-old Polish woman was referred to the emergency room for a 2-month history of fatigue, 10 kg weight gain and NYHA II dyspnea. She has been treated in 2014 by corticosteroids for a nephrotic syndrome responsible to acute renal failure, attributed to a minimal change glomerulonephritis, evolving with time to a progressive decrease of her renal function. In addition, she presented a 3-year history of skin changing appearance with premature ageing of the face, neck, chest, arms and feet. On physical examination, she looked much older than her age, with a sagging of the skin and a lack of elasticity that predominates in the neck, axillary regions, back and abdomen. She had peripheral edema and a blood pressure at 160/90 mmHg. Initial lab tests revealed a terminal stage chronic renal failure (creatinine, 10.24 mg/dL) with microcytic anemia (hemoglobin, 7.2 g/dL), hypogammaglobulinemia (IgG, 5 g/L) with a discreet IgG Lambda M-protein and low C3 levels. Both kappa and lambda light chains were elevated with a normal kappa/lambda ratio, and associated to a significant proteinuria (4.78 g/L) with excess lambda LC and micro-hematuria. Kidney biopsy confirmed the presence of nodular glomerulosclerosis with peritubular anti-IgG staining and 'powdery punctate' deposits along the inner aspect of the tubular basement membranes, confirming the diagnosis of HCDD. IgG and C3 deposits were also reported on skin biopsies, in addition to elastic fibers degenerative changes responsible for cutis laxa. Functional pulmonary evaluation revealed a marked restrictive syndrome with a decreased diffusion capacity suggesting an interstitial lung disease, probably related to the elastolysis seen in cutis laxa. Bortezomib-based chemotherapy was started, with the prospect to collect stem cells later on, and in case of complete hematological response, proceed to kidney transplantation. Monoclonal gammapathies are rarely found in patients under the age of 40, and can be associated with numerous clinical manifestations such as MGRS. HCDD is an extremely rare condition, in which patients presented with nephrotic syndrome, hematuria and hypertension, develop progressive renal failure, but can be successfully treated with chemotherapy.

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