**PB1989 LIGHT CHAIN AMYLOIDOSIS (AL): THE EXPERIENCE OF A TUNISIAN CENTER**

**Topic:** 14. Myeloma and other monoclonal gammopathies - Clinical

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**Background:** The AL amyloidosis is one of the most common types of amyloidosis that is derived from the immunoglobulin light-chain. The clinical presentation varies widely depending on which organs are involved. In this work, we report the experience of the hematology department of Sfax in the management of patients followed for AL amyloidosis.

**Aims:** Our study aims to evaluate the clinico-biological, therapeutic and evolutionary characteristics of al amyloidosis in our patients.

**Methods:** Our study is retrospective, it concerned AL amyloidosis patients, followed in the hematology department of Hedi Chaker Sfax hospital between January 2010 and December 2021.

The treatment was based on different chemotherapy protocols: the Cybor-D protocol (Bortezomib – Cyclophosphamide – Dexamethasone), the BorTD protocol (bortezomib – thalidomide – dexamethasone) and the M-DEX regimen (melphalan – Dexamethasone).

**Results:** Our series included 20 patients with a male predominance (sex-ratio = 1.8) and a mean age of 58 years. 85% of the patients were followed for a multiple myeloma which was then complicated by amyloidosis. The initial symptomatology was polymorphic: cutaneous-mucosal involvement, cardiac symptomatology and neurological symptomatology were noted in 50%, 30% and 25% of cases respectively. The diagnosis was made on 2 different sites in 11 patients (55% of cases). Anemia was found in 55% of cases, and renal failure was reported in 50% of theses cases. Monoclonal gammopathy was found in all patients with demonstration of the Lambda and kappa chain in 50% of cases each. Transthoracic ultrasound revealed signs of myocardial amyloid infiltration, such as wall thickening greater than 15 mm, in 11 patients. Cardiac involvement, labial amyloidosis and renal involvement were diagnosed in 60%, 55% and 25% of cases respectively. According to the classic Mayo Clinic prognostic score, 4 patients were at stage I, 4 others were at stage II and 12 patients were at stage III.

Regarding the treatment, 9 patients (45% of cases) received the Cybor-D protocol, 6 patients were treated with the BorTD protocol, 4 patients received the M-Dex regimen, and 1 patient died before starting treatment. This first-line treatment resulted in objective response rate (VGPR and CR) at 42% of cases (n=8), partial response at 16% (n= 3), Progression at 16% (n=3), toxic death at 10% (n=2) and 3 patients were lost to sight before assessment. This objective response was obtained by protocols based on Bortezomib. For patients with amyloid cardiomyopathy (12 patients), 5 were progressing at the end of treatment despite the fact that 4 of them were in hematological CR, and 2 died from cardiac decompensation.

A relapse occurred in 6 patients out of 11(CR+VGPR+RP) after a median time of 29 months.

Median overall survival was 18 months.

**Summary/Conclusion:** Through this work, we notice the polymorphism of the amyloidosis clinico-biological presentation as well as the concept of multiorgan involvement. The light chain was Lambda in 50% of cases, whereas it is predominant (80%) in the literature (1), and cardiac involvement was the leading cause of morbidity and mortality as similar as what is reported (1). Treatment is based on Bortezomib combined with other chemotherapy.
molecules (2). With this protocol, we obtained an objective response rate and a median survival rate a bit lower than the rates described in literature (1).