nosticadas entre o período fevereiro de 2016 a julho de 2019 na cidade de Guanambi-BA. Os dados foram coletados de prontuários oriundos do serviço Centro de Hematologia de Guanambi. Os diagnósticos confirmados verificados foram: leucemias agudas e crônicas; mieloma múltiplo; neoplasias mieloproliferativas crônicas (trombocitemia essencial, policitemia vera, mielofibrose primária e secundária e leucemia mielóide crônica). **Resultados:** Foram diagnosticados 104 casos de doenças onco-hematológicas num raio de 159,8 km nas mesorregiões centro-sul e extremo-oeste da Bahia, tendo Guanambi como centro, e resultaram em uma população total de 535.237 mil pessoas. Na população estudada, houve predomínio no sexo feminino, indivíduos acima de 60 anos e a maior ocorrência de casos foi no ano de 2018. Aproximadamente 52% dos pacientes eram do sexo feminino e 48% do sexo masculino, com idades entre 1 e 92 anos, enquanto o pico de incidência sucedeu acima dos 60 anos de idade. O Mieloma Múltiplo foi a doença mais frequente (27,8%), seguida pelas Leucemias Agudas (25,96%), Leucemias Linfocíticas Crônicas (17,3%), Trombocitemia Essencial (13,1%) Mielofibroses (6,5%), Policitemia Vera (4,9%), enquanto a menos comum foi a Leucemia Mieloide Crônica (1,6%). **Discussão:** Considerando dados das estimativas disponibilizadas pelo IBGE e pelo INCA dos anos de 2017 a 2019 para a incidência de Leucemias no Nordeste e na Bahia, os resultados do estudo foram confrontados a essas informações por meio de proporção populacional, considerando o número de habitantes dos municípios cobertos pelo serviço de saúde. Assim, percebemos que para o ano de 2017, o esperado para a região segundo os dados oficiais seriam de 20 novos casos, enquanto apenas 7 foram registrados pelo centro de hematologia. Para 2018, o esperado seria de 23 novos casos, sendo que um total de 41 diagnósticos foram feitos. Por fim, em 2019 eram estimados 23 casos, sendo que o serviço registrou 17 casos em um período de apenas 7 meses. **Conclusão:** O estabelecimento da situação epidemiológica dessas doenças no Brasil requer estudos em diferentes regiões do país, porém os dados existentes são pouco detalhados, dão ênfase à macrorregiões, há escassez de dados microrregionais e para determinadas doenças há ausência de dados. Portanto, a epidemiologia da região abordada não se encontra limitada aos dados estabelecidos no presente estudo, devido aos empecilhos em sua obtenção, sendo estes de cunhos regionais, sociais e governamentais. Entretanto, percebe-se que mesmo diante dos empecilhos, há uma tendência que os dados sejam superiores aos índices estimados nacionalmente, levando à consideração de que podem haver fatores predisponentes específicos da região estudada que justificariam o aumento nos diagnósticos.

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PITFALLS TO DIAGNOSE AND TREAT LIGHT CHAIN AMYLOIDOSIS IN A UNIVERSITY REFERENCE CENTER: 10 YEARS OF EXPERIENCE IN A PUBLIC HEALTH SYSTEM

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Background: Amyloidosis is caused by tissue deposition of misfolded protein aggregates, leading to organ dysfunction. Light chain amyloidosis (AL) is the most common subtype. Epidemiological data are scarce in Latin America. **Objectives:** To describe clinical and laboratory characteristics and treatments of patients with systemic AL amyloidosis in a reference center. **Methods:** Retrospective cohort study of patients with biopsy-proven systemic AL amyloidosis diagnosed from 2009 to 2018 at the Hospital das Clinicas, University of Sao Paulo. Primary endpoint was overall survival (OS). **Results:** From 83 cases revised, 75 patients met the eligibility criteria. 53% were male with median age of 62 years. Before diagnosis 56% were seen by ≥3 physicians. Referrals to the hematologist were made mainly by nephrologists (46%), cardiologists (23%) and general practitioners (14%). Median time between symptom onset and diagnosis was 9.1 months. In 75% of patients ECOG was ≤ 2 at diagnosis. Initial clinical presentations were: 61% renal disorders, 43% consumptive syndrome, 39% heart disease, 25% gastrointestinal symptoms, 20% neuropathy, 19% fatigue, 15% skin lesions, 12% macroglossia, 11% hepatic disorders and 5% periorbital purpura. Mean number of biopsies performed per patient was 2.5. In 67% a method to subtype amyloid on biopsy was performed: 68% indirect immunofluorescence, 36% immunohistochemistry and mass spectrometry in 1 case. Free light chain was assessed in 49% of cases. 75% were λ subtype. The mean number of organs involved was 2.8 (11% 1 organ, 37% 2, 52% ≥ 3). Main affected organs were: 81% heart, 63% kidney, 52% soft tissue. 34% had coexisting multiple myeloma. Standard Mayo Clinic (SMC) staging was evaluated in 56% of cases: 59% stage III, 31% stage II, 10% stage I. Stage III patients were assessed by European staging: 32% IIIa, 48% IIIb, 20% IIIc. Revised Mayo Clinic staging was available in 21% of patients: 25% in each stage I to IV. Renal staging showed 81% stages I/II, 19% stage III. 81% of patients were treated with chemotherapy (54% melphalan, 43% cyclophosphamide, 10% bortezomib and 18% thalidomide). Median number of cycles was 4. 12% underwent autologous stem cell transplantation (ASCT). 3 patients received doxycycline, 12% only supportive measures and 1 patient underwent kidney transplantation. 40 patients had hematological response assessed: 30% PR, 12.5% VGPR, 17.5% CR, 25% no response and 15% disease progression. Median follow-up time of survivors was 66.3 months and estimated OS was 17% in 5 years. Statistical difference was observed in median OS of SMC stage I-II and III: 51.6 and 14.4 months respectively (p = 0.023). **Discussion:** The broad clin-

ical spectrum of AL presentation reflects the complexity of diagnostic approach: patients are seen by different specialists, require more than 2 biopsies and are diagnosed late in advanced stages with markers of poor prognosis. Moreover, proteasome inhibitor is not widely available in public system and few patients are eligible for ASCT. Altogether may explain poor outcomes of AL patients in our center. **Conclusions:** Diagnosis of systemic AL amyloidosis is a challenge in Brazil. Medical education, better tools for diagnosis, establishment of a multidisciplinary team and a registry, availability of disease-modifying drugs and ASCT may improve outcomes.

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POMALIDOMIDE, BORTEZOMIB, DEXAMETHASONE AFTER 1 PRIOR LINE OF THERAPY IN RELAPSED OR REFRACTORY MULTIPLE MYELOMA: SUBANALYSIS OF OPTIMMISM BY AGE, PRIOR TRANSPLANT, AND HIGH-RISK CYTOGENETICS

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Goals: Upfront Lenalidomide (LEN) until disease progression is a standard treatment (Tx) for newly diagnosed multiple myeloma, regardless of patient (pt) age and transplant eligibility. However, pts who have exhausted the benefit of LEN at first relapse are a growing population in need of effective Tx options. In the OPTIMISMM trial (NCT01734928), pomalidomide (POM), bortezomib (BORT), and dexamethasone (DEX; PVd) significantly improved PFS vs Vd (median, 20.7 vs 11.6 mos; hazard ratio = 0.54 [95% CI, 0.36-0.82]; p = .0027) in an analysis of pts at first relapse. Here we report the efficacy and safety of PVd by age, prior stem cell transplant (SCT) status, and presence of high-risk cytogenetic abnormalities (HR CAs; defined as del[17p], t[4;14], or t[14;16]) in pts treated after 1 prior line of therapy (LOT). **Methods:** Pts received PVd or Vd (1:1) in 21-day cycles (C). POM 4 mg/d on d 1-14 (PVd arm only); BORT 1.3 mg/m^2 on d 1, 4, 8, and 11 of C 1-8 and on d 1 and 8 of C 9+; and DEX 20 mg/d (10 mg/d for pts aged > 75 yrs) on days of and after BORT. PFS was the primary endpoint. **Results:** 226 of 559 pts (40%) enrolled in OPTIMISMM had 1 prior LOT: 100 pts aged ≤ 65 yrs (49 PVd, 51 Vd) and 126 aged > 65 yrs (62 PVd, 64 Vd). In pts aged ≤ 65 yrs (PVd vs Vd), 55.1% vs 51.0% were LEN refractory and 83.7% vs 72.5% had prior BORT. In pts aged > 65 yrs, 59.7% vs 60.9% were LEN refractory and 41.9% vs 46.9% had prior BORT. After 1 prior LOT, PVd significantly improved PFS in pts aged ≤ 65 yrs (median, 22.0 vs 13.1 mos; HR = 0.49 [95% CI, 0.26-0.92], p =.0258) and those aged > 65 yrs (median, 17.6 vs 9.9 mos; HR = 0.57 [95% CI 0.34-0.97], p =.0369) vs Vd; data cutoff was 26 Oct 2017. In pts aged ≤ 65 yrs, ORR was 89.8% vs 54.9% (p <.001; ≥ VGPR 65.3% vs 17.6%) and in pts aged > 65 yrs, ORR was 90.3% vs 54.7% (p <.001; ≥ VGPR 58.1% vs 26.6%). Significant improvements in PFS and ORR with PVd vs Vd were also observed in pts with prior SCT (56 PVd, 54 Vd; median PFS, 22.0 vs 13.8 mos, p =.0241; ORR, 91.1% vs 57.4%, p < .001) or without prior SCT (55 PVd, 61 Vd; median PFS, 16.5 vs 9.5 mos, p =.0454; ORR, 89.1% vs 52.5%, p < .001). Pts with HR CAs had a median PFS of 14.7 mos with PVd (n = 18) vs 9.9 mos with Vd (n = 14); ORR was 94.4% vs 57.1% (p = .027), ≥ VGPR was 72.2% vs 35.7%. The most common grade 3/4 treatment-emergent adverse events (PVd vs Vd) were neutropenia (49.0% vs 6.3%), infections (system organ class; 30.6% vs 14.6%), and thrombocytopenia (26.5% vs 18.8%) in pts aged ≤ 65 yrs and neutropenia (25.8% vs 12.9%), infections (27.4% vs 16.1%), and thrombocytopenia (14.5% vs 22.6%) in pts aged > 65 yrs. **Discussion:** In pts with LEN-pretreated RRMM at first relapse, PVd reduced the risk of progression or death by 51% in pts aged ≤ 65 yrs and 43% in those > 65 yrs vs Vd, and led to significantly improved ORR and deeper responses. Similar outcomes were observed in pts regardless of prior SCT. Although limited by the number of pts, the high ORR and depth of response seen with PVd in pts with HR CAs are promising. The safety of PVd was consistent with the known profiles of POM, BORT, and DEX. **Conclusions:** These results support the use of PVd after first relapse in pts previously treated with LEN, regardless of age, prior SCT status, and presence of HR CAs.