intersticiais e perivasculares com cadeias leves kappa e lambda de imunoglobulina apresentando relação dentro dos limites da normalidade. Afastada a presença de mieloma múltiplo, a paciente iniciou terapia com Dexametasona 4 mg (10 comprimidos ao dia por 4 dias), Melfalana 2 mg por 4 dias e profilaxia de Pneumocistose com tratamento com Sulfametoxazol + trimetoprima 800+160 mg 3x por semana. Após tratamento não houve queda nas cadeias leves livres e nem melhora das funções cardíaca e renal e foi encaminhada para o serviço de transplante de medula óssea. **Discussão:** A ACL é uma doença rara caracterizada pelo depósito de cadeias leves monoclonais em diversos órgãos, mais prevalente em homens e diagnosticada em média aos 58 anos. Plasmócitos clonais produzem excessivamente cadeias kappa ou lambda, com metade dos casos associada ao mieloma múltiplo e 20% à gamopatia monoclonal de significado indeterminado. A disfunção renal e a síndrome nefrótica são comuns, enquanto envolvimento hepático e cardíaco são menos frequentes, incluindo cardiomiopatia e insuficiência cardíaca. O diagnóstico é confirmado por biópsia com coloração pelo vermelho do congo e espectroscopia de massa. O tratamento visa suprimir a produção de cadeias leves, destacando-se o regime com daratumumabe, bortezomibe, ciclofosfamida e dexametasona e o transplante autólogo de células-tronco, que apresenta boa eficácia e taxas baixas de mortalidade. A resposta hematológica é um importante preditor de sobrevida, com taxa de 53% em dez anos para pacientes com resposta completa ao tratamento. **Conclusão:** A ACL apresenta desafios diagnósticos e terapêuticos substanciais. Este caso ilustra a importância de um diagnóstico precoce e de um tratamento intensivo, visando aumentar a sobrevida e o bem-estar do paciente, mesmo diante de um prognóstico reservado.

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MAPPING AMYLOID SUBTYPES IN BRAZIL THROUGH MASS SPECTROMETRY: RESULTS OF A DIAGNOSTIC SUPPORT PROGRAM

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Objectives: Amyloidosis results of tissue deposition of misfolded proteins aggregates, leading to localized or systemic forms. Over 40 proteins are amyloidogenic, the main 2 being immunoglobulin light chain (AL) and transthyretin (ATTR). Mass spectrometry (MS) is the gold-standard for subtyping amyloid. Despite MS commercial availability in Brazil since 2019, amyloidosis remains underdiagnosed with nearly a years delay, underscoring the need for ongoing medical education and specialized referral centers. Limited access to MS due to regional disparities and health system heterogeneity led to the implementation of a diagnostic support program (DSP) operated by a pharmaceutical company. This study aims to describe the occurrence of amyloid subtypes in Brazil using DSP data. **Materials and methods:** This observational, cross-sectional study analyzes data from DSP database between January 2022 and May 2024. Eligible patients were ≥18 years, with biopsy-proven amyloidosis confirmed by Congo red staining and consented to data collection/publication. Amyloid was extracted by laser microdissection, processed by protein denaturation, trypsin digestion and resulting peptides analyzed by MS (QExactiveHF-X mass spectrometer coupled with NanoUltimate liquid chromatography). Protein identification used bioinformatics (MaxQuant v.4.2.1, FragPipe v.13 platforms against UniProt/SwissProt database). The techniques were conducted at Grupo Fleury Laboratory. Sociodemographic characteristics, biopsy sites and MS results were analyzed using descriptive statistics. **Results:** 403 patients were included. Median age was 65 years (56-74), 52% were male, and 65% from the southeast region. AL was the predominant subtype in Brazil (76%), followed by ATTR (7%), both present in in all regions of the country. AA and AHL types were reported only in the southeast region. Only one case of AA amyloidosis was observed, in the gastrointestinal tract. Abdominal fat and kidney were the most common sites of biopsy (23% and 22%), followed by bone marrow (15%). This pattern was consistent across most regions, except in the south, where abdominal fat and bone marrow were equally used (24.1% each), and heart biopsies were the third most common (17.2%). AL amyloidosis was mainly identified in the kidney and bone marrow (29%; 14%), and ATTR in abdominal fat, and heart (35%; 30%). **Discussion:** This study is the first to analyze a Brazilian national cohort with amyloidosis, providing a comprehensive overview of amyloidosis subtypes and their geographical distribution across different country regions. AL is the most common subtype in Brazil, followed by ATTR, consistent with findings in Europe and the United States. Abdominal fat is the main biopsy site nationwide, following international recommendations for diagnosing amyloidosis and avoiding invasive organ biopsies. **Conclusion:** The widespread use of MS on a national scale, facilitated by the creation of a DSP, has proven to be a relevant tool in the diagnosis of amyloidosis in Brazil. It allows for accurate amyloid subtyping, contributes to epidemiological information, improves the diagnostic process and enables the correct treatment of patients. Additionally, it strengthens the establishment of a reference laboratory with expertise in the field in Brazil, with the potential to become a reference center in Latin America. **Financial disclosure:** The development of this study was funded by Janssen-Cilag Ltda

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PRACTICAL CHALLENGES IN AMYLOID SUBTYPING IN BRAZIL: A COMPARATIVE STUDY BETWEEN MASS SPECTROMETRY AND A CLINICAL-LABORATORY MODEL

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Background: Systemic amyloidosis is a protein misfolding deposition disease often under-recognized in middle-income countries. The precise identification of the precursor protein is a key factor in the diagnosis challenging clinical practice, especially outside of reference centers. **Objectives:** To subtype amyloid by mass spectrometry (MS), the gold standard method for diagnosis, and to compare it with the clinical-laboratory model (CLM) for amyloidosis diagnosis when the gold standard is not available. **Methods:** This is a retrospective, observational, unicentric study. Patients with a histologic diagnosis of systemic amyloidosis from 2009 to 2018 at the Hospital das Clínicas of USP, a Brazilian public university hospital, were included. Patients were identified through database searches and an active search of biopsies positive for amyloid in the pathology laboratory archives. Stored tissue samples were selected by availability to perform MS. All medical records were retrospectively reviewed. Amyloid specimens were analyzed by MS coupled with liquid chromatography following laser microdissection, thermochemical processing, and trypsin digestion. Amyloid subtypes were defined by the CLM using combined data on clinical presentation, monoclonal gammopathy, pyrophosphate scintigraphy, immunohistochemistry, immunofluorescence, and genetic mutations. When exact criteria couldn't be met to define the amyloid subtype, clinical judgment was used based on available data. **Results:** From the 138 patients with biopsy-proven systemic amyloidosis, 57 biopsies were excluded (17 external diagnoses, 13 unavailable samples, 8 insufficient samples for laser microdissection, 10 insufficient for MS, 9 had no remaining amyloid deposit in the stored block). MS was performed on 81 biopsies from 13 different organs: 30% kidney, 23% heart, 14% subcutaneous fat, 7% gastrointestinal tract, 6% nerve, 5% respiratory tract, 4% bone marrow, 3% muscle, and 1% each lymph node, gingiva, liver, salivary gland and tongue. The following amyloid subtypes were detected: AL (58%, n = 36), ATTR (23%, n = 14), AA (8%, n = 5), AFib (3%, n = 2) and AH (2%, n = 1). In 2 cases amyloid co-depositary proteins were not identified, and another 2 were inconclusive (1 weak detection of fibrinogen with a dispersed pattern and 1 detection of both fibrinogen and transthyretin). Results are still pending for 19 cases. The CLM correctly subtyped amyloid in 100% of AA and AFib subtypes, 94% of AL, and 79% of ATTR cases. In 2 AL and 1 ATTR cases, the CLM was not able to determine the subtype, and patients received only supportive treatment. ATTR was misdiagnosed as AL in 3 patients, who were treated with anti-plasma cell agents. The patient with AH was diagnosed as AL by the CLM. **Discussion:** Although the CLM showed good performance in subtyping amyloid in our cohort, it failed in 3 patients, resulting in a misdiagnosis that led to inappropriate treatment with chemotherapeutic agents. Additionally, 3 other patients did not receive specific treatment due to the models limitations in identifying the amyloid subtype. **Conclusion:** Our study emphasizes the role of mass spectrometry as the gold standard method for accurately subtyping amyloid and the importance of establishing referral centers for amyloidosis. This approach can prevent potential harm from misdiagnosis and improve patient care.

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OS IMPACTOS DOS CUIDADOS PALIATIVOS NA QUALIDADE DE VIDA DE PACIENTES IDOSOS COM MIELOMA MÚLTIPLO

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Introdução: O mieloma múltiplo (MM) é uma neoplasia hematológica maligna, caracterizada pela proliferação clonal de plasmócitos originários da medula óssea, resultando em comprometimento sistêmico do paciente. Com milhares de novos casos registrados anualmente, essa doença afeta predominantemente homens após a quinta década de vida. Associada a um prognóstico desfavorável, os indivíduos com mieloma múltiplo apresentam uma sobrevida média de 5 anos e podem manifestar uma variedade de sintomas, incluindo dor óssea, complicações renais, hipercalcemia, anemia, infecções recorrentes, além de impactos psicológicos, sociais e espirituais. A importância dos cuidados paliativos (CP) em doenças crônicas é amplamente reconhecida, independentemente do estágio da doença. Os CP visam aliviar o sofrimento e a dor do paciente quando não há mais esperança de cura, proporcionando um tratamento adequado e humanizado para o paciente e familiares, visando à prevenção de ansiedade, depressão e suicídio. **Objetivo:** O objetivo é analisar criticamente o conhecimento atual sobre mieloma múltiplo em idosos em CP. **Materiais e métodos:** Trata-se de uma revisão bibliográfica da literatura integrativa, utilizou-se uma abordagem qualitativa com propósito de explorar o tema. Foram utilizadas como bases as seguintes plataformas: BMJ journals, Ministério da Saúde, Organização Mundial de Saúde, PubMed e SciELO. A partir do recorte temporal de 2002 a 2022, nos idiomas inglês e português. **Resultados:** Os pacientes que receberam tratamento hematológico tradicional tiveram indicadores inferiores de qualidade ao final da vida em comparação aos pacientes que receberam CP com início em até 8 semanas após diagnóstico. Estes apresentaram sintomas mais brandos de dor e diferentes combinações dos indicadores, como a falta de necessidade de intubação, hospitalização e realização de ressuscitação cardiopulmonar, além da dispensabilidade do tratamento anti-mieloma nos últimos dias de vida. **Discussão:** As intervenções dos CP baseiam-se na avaliação da condição biopsicossocial do paciente, considerando o grau de estadiamento do tumor, a rede de apoio presente, gênero e idade, buscando manejar os sintomas e informar o paciente e seus entes sobre a condição, dando