

vistos, em maior frequência, na coluna torácica, coluna lombar e fêmur, respectivamente. Apenas sete casos sob a forma extramedular (PEM) foram diagnosticados no período do estudo, entre eles: seio maxilar da face, laringe, faringe, parede torácica, pulmão, mama e omento. Em relação às lesões de órgãos alvo, mais da metade dos pacientes já apresentavam outras lesões osteolíticas ao diagnóstico, 35% apresentavam anemia, 22% apresentavam alteração da função renal e a minoria apresentava hipercalcemia. A maioria dos casos não tinha estudo da medula óssea descrita no prontuário. Foram identificados 38 estudos medulares, e metade destes resultados apresentavam contagem de mais de 10% de plasmócitos clonais. **Discussão:** O PS é raro e definido como um tumor sólido proveniente da proliferação anormal de linfócitos tipo B maduros, sem evidência de proliferação sistêmica. São classificados de acordo com sua localização em PSO e PEM. De acordo com a literatura, 70% são PSO e acomete principalmente a coluna vertebral e fêmur. O PEM é mais comum na região de cabeça e pescoço (seios da face, naso e orofaringe), seguido de acometimento gastrointestinal e pulmonar. A topografia dos PS identificados em nossa população foi semelhante ao encontrado na literatura, porém, identificamos um caso raro de plasmocitoma em mama e omento. **Conclusão:** Pacientes que apresentam PS tem maior risco de desenvolver Mieloma Múltiplo, portanto, requerem tratamento adequado e acompanhamento médico. São necessários novos estudos clínicos para definir a melhor forma de seguimento após o diagnóstico e tratamento de PS, com o objetivo de controlar precocemente a doença e melhorar a sobrevida.

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CUTANEOUS INVOLVEMENT IN MULTIPLE MYELOMA IN THE ERA OF PROTEASOME INHIBITORS: HAS THE DISMAL PROGNOSIS CHANGED?

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Material and method: We present a 3-case series of MM with cutaneous involvement and a wide range of clinical manifestations and dismal prognosis after cutaneous involvement. **Results:** Case 1- A 55-year-old man presenting with symptomatic MM, IgG- kappa, Durie-Salmon IIIA stage, and ISS = 3. He was treated with CyBORD, 4 cycles with complete response. Subsequently, he underwent HSCT whose conditioning was performed with MEL 200 mg/m² and infusion of 12 x 10⁶ CD34/kg cells. After HSCT, he remained in complete response and maintenance with bortezomib. After 4 months, he developed erythematous infiltrated plaque that evolved to an exophytic tumor on the left posterior axillary line. In addition, sudden spinal cord compression on T12 level whose radiological investigation demonstrated bone plasmacytoma was also observed. Decompressive laminectomy and skin lesion biopsy

were performed, which confirmed the presence of cutaneous infiltration by MM. Hence, the patient received a third-line treatment with DVd. The patient's renal function deteriorated and his skin lesions showed progressive growth. Eventually he became septic and died. Case 2- A 76-year-old man diagnosed with IgA/lambda secreting MM with Durie Salmon IIIA and ISS II. His initial treatment consisted of an association of melphalan, prednisone and bortezomib, which resulted in complete remission. A few months later, the patient relapsed leading to further treatment with lenalidomide and dexamethasone. At that moment, the patient showed no response and underwent a regimen containing KRD, which was unsuccessful. The association of dara-VMP was used after he developed renal failure, cardiovascular impairment and skin lesions over his legs were noted. A fine needle aspiration was performed and microscopic analysis showed signs of cutaneous plasmacytosis confirming cutaneous infiltration by MM. The patient developed multiple organ failure after sixty days of the onset of such cutaneous lesions. Case 3- A 50-year-old woman with a diagnosis of IgA secretory MM, Durie Salmon IIIB, ISS 3. Initial therapeutic approach was performed with thalidomide and dexamethasone. A partial response with stabilization of heavy and light chain monoclonal protein. A few months later, her condition deteriorated. Thus, a protocol with CYBORD was introduced. Despite treatment, she developed purplish plaques and nodules on the left anterior chest wall and over both arms, which evolved with rapid growth. Biopsy of the cutaneous lesions was performed, which revealed an extramedullary plasmacytoma. Afterwards, a more aggressive therapeutic regimen was applied containing melphalan, vincristine, cyclophosphamide and methylprednisolone with no clinical response. Ultimately she died of sepsis. **Discussion:** Cutaneous plasmacytic infiltration in multiple myeloma (MM) is a rare clinical condition, usually associated with a high burden of malignant cells and a poor prognosis. The clinical management of the secondary infiltration of the MM does not have a standard optimal therapeutic regimen. There are some physiopathologic hypothesis that arise that cutaneous infiltration is of more aggressive forms of myeloma usually with characteristics of plasmablasts and clones with high prevalence of high-risk molecular markers. **Conclusion:** In our experience, the use of these drugs has not translated into additional survival to patients, underscoring the role of skin involvement as a poor prognostic factor.

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SÍNDROME DE POEMS COM PADRÃO BICLONAL IGG/KAPPA E IGA/KAPPA

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Introdução: A síndrome POEMS é uma doença rara definida por polineuropatia periférica, organomegalia, endocrinopatia, distúrbio monoclonal de células plasmáticas e alterações cutâneas. De acordo com classificação da Organização Mundial da Saúde, a síndrome segue como uma neoplasia de células plasmáticas