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CASE REPORT

Cutaneous involvement by a mantle cell lymphoma

Envolvimento cutâneo por linfoma do manto

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Abstract

We describe the case of a 73-year-old female patient with priors of a mantle cell lymphoma (MCL), diagnosed 9 months ago, treated with chemotherapy, and currently with maintenance rituximab therapy, and a total knee replacement on the left leg 1 year ago. The patient presented to the Dermatology Department with patches on the left leg, with 3 months of evolution.

On examination, the patient presented infiltrated, confluent erythematous-violaceous patches and plaques, on the anterior surface of the left knee and leg, along the arthroplasty scar. No complaints associated. Blood work showed anaemia, neutropenia and an elevation of lactate dehydrogenase (LDH). A skin biopsy revealed occupation of the skin by a diffuse lymphoid proliferation, with intermediate-sized cells, with scarce cytoplasm, hyperchromatic nuclei and irregular borders. At immunohistochemistry, the cells were a cluster of differentiation (CD) 20+, CD5+, B-cell lymphoma 2 (Bcl-2) +, cyclin D1+ and CD3–. These findings are compatible with cutaneous involvement by MCL. The patient underwent radiotherapy on the left leg with regression of the lesions and is currently under chemotherapy. Mantle cell lymphoma (MCL) is a B-cell non-Hodgkin lymphoma (NHL). Cutaneous involvement is rare, secondary and means lymphoma dissemination. This case is also interesting for the site of skin involvement on a scar of knee replacement surgery.

Keywords: Cutaneous involvement. Cutaneous lymphoma. Lymphoma. Mantle cell lymphoma.

Resumo

Doente do sexo feminino, 73 anos, com antecedentes pessoais de linfoma do manto diagnosticado há 9 meses, para o qual fez quimioterapia, atualmente em tratamento de manutenção com rituximab, e artroplastia total do joelho esquerdo há um ano.

A doente vem à consulta de Dermatologia por manchas na perna esquerda com 3 meses de evolução. Ao exame dermatológico observavam-se manchas e placas eritematovioláceas, confluentes, infiltradas, na face anterior do joelho e perna esquerdas, ao longo da cicatriz de artroplastia. Sem queixas associadas. Sem lesões no restante tegumento cutâneo. Analiticamente apresentava anemia, neutropenia e aumento da LDH. A biópsia cutânea revelou pele ocupada por proliferação linfóide de padrão difuso, constituído por células de tamanho intermédio, escasso citoplasma, núcleo hipercromático e

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contornos irregulares. Ao estudo imunohistoquímico as células eram CD20+, CD5+, BCL2+ e Ciclina D1+ Achados compatíveis com envolvimento cutâneo por linfoma do manto. A doente realizou radioterapia com regressão das lesões e encontra-se atualmente sob quimioterapia. O linfoma de células do manto é um linfoma não Hodgkin, de células B. O envolvimento cutâneo é raro, secundário e sinaliza disseminação da neoplasia. Este caso é interessante pelo local onde ocorreu o envolvimento cutâneo, na cicatriz de artroplastia do joelho.

Palavras-chave: Envolvimento cutâneo. Linfoma. Linfoma cutâneo. Linfoma do manto.

Introduction

Mantle cell lymphoma (MCL) is a B-cell NHL that develops from malignant B-lymphocytes within a region of the lymph node known as the mantle zone. MCL represents only 6% of all NHL[1].

Many affected individuals have widespread disease at diagnosis, with involved regions often including multiple lymph nodes, the spleen, and, potentially, the bone marrow, the liver and the gastrointestinal tract. Skin involvement is rare and is associated with progressive disease[1].

Our case report describes one of the rare cases of cutaneous involvement by an MCL.

Clinical significance

We describe the case of a 73-year-old female patient with priors of an MCL and a total left knee replacement 1 year ago.

Around 9 months ago, the patient started experiencing asthenia and undesired weight loss. Blood work showed anaemia and neutropenia, and clinical examination detected cervical lymphadenopathies. Biopsy of a lymph node diagnosed an MCL. Bone marrow biopsy showed involvement of the bone marrow and a positron emission tomography/computed tomography scan revealed multiple lymphadenopathies, thus an MCL stage IVB.

The patient underwent six cycles of chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone, with disease control, followed by maintenance rituximab therapy, currently on the second cycle.

The patient presented to the dermatology department with patches on the left leg, with 3 months of evolution.

On examination, the patient presented with infiltrated, confluent, shiny erythematous to violaceous patches and plaques on the anterior surface of the left knee and leg and on the arthroplasty scar (Fig. 1), with no associated complaints and no other relevant cutaneous lesions.

Blood work showed aggravation of anaemia, neutropenia and an elevation of LDH *de novo*.

A skin biopsy revealed dense occupation of the skin by a diffuse patterned lymphoid proliferation, with intermediate-sized cells, scarce cytoplasm, hyperchromatic nuclei and irregular borders (Figs. 2 and 3). In immunohistochemistry, the cells were positive for CD20, CD5, cyclin D1 and Bcl-2, and negative for CD3 and CD10 (Figs. 4 and 5). These findings are compatible with cutaneous involvement by MCL.

Maintenance therapy was stopped and the patient underwent radiotherapy on the left leg with regression of the lesions. The patient is currently under chemotherapy with R-BAC (rituximab, bendamustine, and cytarabine).

Discussion

Mantle cell lymphoma (MCL) is an uncommon form of NHL, accounting for 5–7% of all cases of NHL. It develops from malignant B-lymphocytes within a region of the lymph node known as the mantle zone[1]. Around 90% of patients with MCL have a mutation that leads to the overproduction of a protein called cyclin D1 in the lymphoma cells[2].

It affects mostly men who are usually 60–70 years old[1].

The disease is typically widespread at diagnosis. Extranodal involvement, especially of the gastrointestinal tract, spleen and bone marrow, is fairly common in MCL[3].

In the classification of World Health Organization, MCL is listed as an extracutaneous lymphoma secondarily involving the skin[2]. Primary involvement of the skin with MCL is controversial and extremely rare. Secondary involvement is described to occur in 2% of all MCL cases and in 17% of cases with stage IV MCL, with about 30 cases reported[4-6]. This case was also a stage IV MCL.

Most cases report skin involvement of the trunk, followed by the face, upper limbs and less commonly, the lower limbs and abdomen. Also, the primary lesions are usually described as erythematous nodules but can also be papules and plaques[2,4-6]. In this case, the patient presented with patches and plaques in only one lower limb at the site of a previous orthopaedic surgery which has never been described to our knowledge.

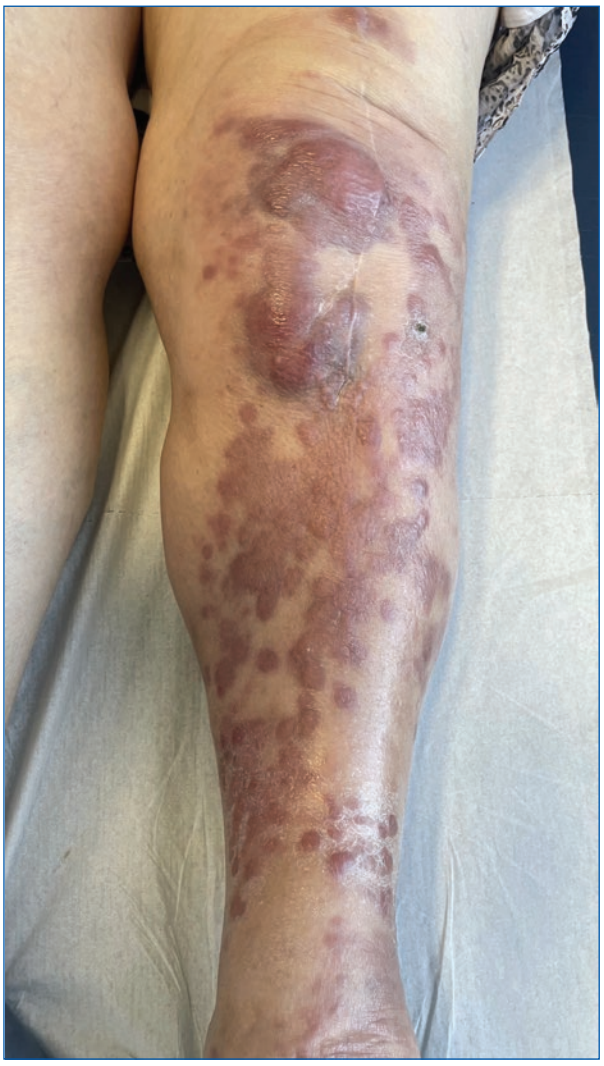

Figure 1. Erythematous-violaceous patches and plaques on the anterior surface of the left knee and leg.

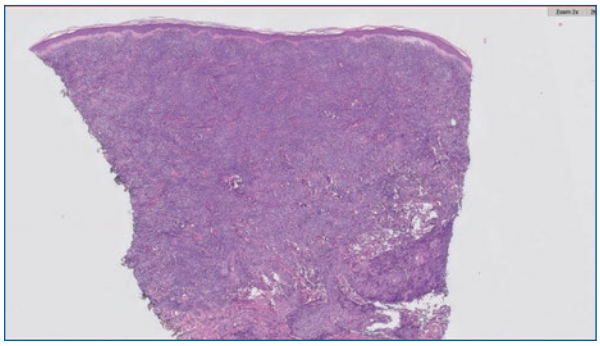

Figure 2. Skin biopsy with the dense occupation of the skin by a diffuse patterned lymphoid proliferation, sparing only a straight subepidermal band (grenz zone).

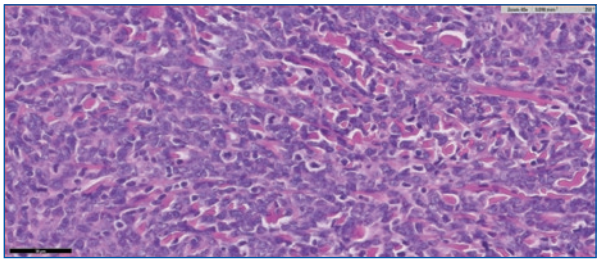

Figure 3. Lymphocytes with intermediate-sized cells, with scarce cytoplasm, hyperchromatic nuclei, and slightly irregular nuclear borders. Some mitotic figures can be seen.

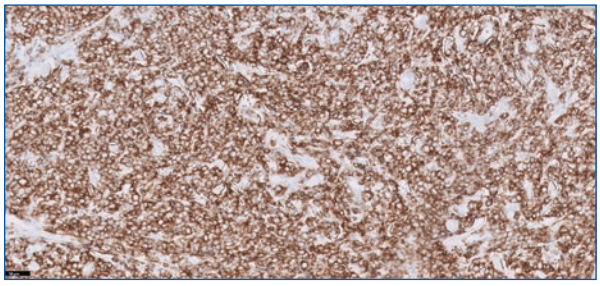

Figure 4. In immunohistochemistry, the cells were positive for CD20, therefore, from the B-lymphocyte lineage.

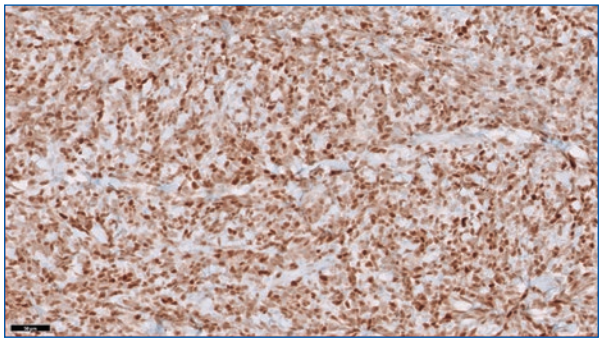

Figure 5. The cells are also positive for cyclin D1, a characteristic of MCL.

The immunohistochemical profile of the MCL of our patient was comparable to that of published cases. According to the guidelines, the diagnosis of MCL should be established on the basis of morphological examination and immunophenotyping with the detection of cyclin D1 protein overexpression. Histologically MCL is composed of diffuse or nodular proliferations of B lymphocytes positive for B-cell markers, like CD79a, CD19, CD20, CD22 and CD5 and usually negative for CD10, CD23 and bcl-6[6]. In this case, the skin biopsy revealed a diffuse lymphoid proliferation with tumour cells positive for B-cell markers—CD20 and CD5, negative for CD3 and CD10 and overexpression of cyclin D1.

Patients who develop the cutaneous disease with widespread MCL typically have a progressive disease and, therefore, poor prognosis[4].

Although rare, skin involvement can be the first complaint of MCL in some cases and therefore, its first manifestation. Since this type of lymphoma is associated with a poor prognosis, a quick diagnosis and rapid treatment are crucial for survival.

Funding

None.

Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work centre on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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