Case Report on Extranodal Presentation of Follicular Lymphoma.

ABSTRACT

Aims: This study aims to present a case of follicular lymphoma with an unusual extranodal presentation in the lower limb and discuss the diagnostic and therapeutic implications.

Presentation of Case: A 55-year-old woman presented with a painless, slowly growing mass in the right thigh. MRI showed a 63mm mass with well-defined boundaries. Excisional biopsy and histopathology confirmed grade 1 follicular lymphoma, but reviewed histopathology favor the diagnosis of follicular lymphoma, grade 3A, with areas of progression to diffuse large B-cell lymphoma. Bone marrow biopsy and PET scan were normal. The patient underwent six cycles of RCHOP chemotherapy. No significant abnormalities were detected post-treatment.

Discussion: Follicular lymphoma typically presents with nodal involvement, and extranodal presentation is uncommon. The MRI findings and excisional biopsy were crucial for diagnosis. The absence of B symptoms and normal bone marrow biopsy indicated a localized disease. The RCHOP regimen is standard for treating follicular lymphoma, and the patient responded well to this treatment. This case underscores the importance of considering follicular lymphoma in the differential diagnosis of extranodal masses and the role of comprehensive diagnostic workup in guiding treatment.

Conclusion: Extranodal follicular lymphoma is a rare clinical presentation. This case emphasizes the necessity of considering follicular lymphoma in the differential diagnosis of extranodal masses and highlights the importance of histological diagnosis for guiding treatment.

Keywords: Extranodal Lymphoma; Histological Diagnosis; Soft Tissue Mass; Chemotherapy Treatment

1. INTRODUCTION

Follicular lymphoma is the most common indolent B-cell lymphoma in adults and the second most common non-Hodgkin lymphoma. It typically presents with nodal involvement and primarily affects adults, with a median age at diagnosis of 65 years. The etiology of follicular lymphoma is not well understood, but factors such as immunosuppression, autoimmune diseases, certain viruses (Epstein-Barr virus, human T-cell lymphotropic virus type I, and human herpesvirus 8), and exposure to pesticides and herbicides have been associated with increased risk. Histologically, follicular lymphoma consists of malignant B-cells derived from germinal centers and often exhibits the translocation t(14;18), leading to overexpression of the anti-apoptotic protein BCL2.[3,4]

This case report describes an unusual extranodal presentation of follicular lymphoma in the lower limb, highlighting the importance of thorough diagnostic evaluation and appropriate therapeutic management.

2. PRESENTATION OF CASE

A 55-year-old woman with a medical history of anxiety, unilateral oophorectomy, and surgery for ectopic pregnancy, presented with a painless, slowly enlarging mass in the right thigh. She was medicated with Citalopram 20mg daily and Victan 1mg as needed.

Upon clinical evaluation, a non-tender mass was noted in the right thigh, with no associated B symptoms (fever, night sweats, weight loss). MRI revealed a 63mm ovoid mass in the anterior-medial aspect of the proximal to mid-thigh, with well-defined borders, relatively hypointense on T1 and hyperintense on T2, and showing homogeneous enhancement with contrast. The mass had a clear plane of separation from adjacent muscular structures and was closely related to the vascular territory of the internal saphenous vein.

Excisional biopsy was performed, and histopathological examination revealed involvement by a small B-cell peripheral lymphoma, consistent with grade 1 follicular lymphoma. Reviewed histopathology favor the diagnosis of follicular lymphoma, grade 3A, with areas of progression to diffuse large B-cell lymphoma, with an immunophenotype CD20+, CD3-, CD5-, cyclin D1-, CD10-, CD23+, Bcl-6+, Bcl-2- and proliferative index (Ki-67) estimated at around 40 to 50%. Bone marrow biopsy was normal, and a post-surgical PET scan showed no abnormalities. The patient was subsequently treated with six cycles of the RCHOP chemotherapy regimen (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone). No significant abnormalities were detected post-treatment.

3. DISCUSSION

Follicular lymphoma (FL) is recognized as the most common indolent B-cell lymphoma among adults, typically presenting with lymphadenopathy. However, extranodal presentations, such as the one observed in this case involving a lower limb mass, are rare and pose diagnostic challenges.[1] This case study highlights the atypical presentation and the diagnostic journey of a 55-year-old woman with a slowly growing, painless mass in her right thigh, later identified as follicular lymphoma.

The initial presentation of a painless mass without accompanying systemic symptoms (fever, night sweats, and weight loss) or lymphadenopathy suggested a differential diagnosis that included benign soft tissue tumors, sarcomas, or other malignancies. The magnetic resonance imaging (MRI) findings further complicated the clinical picture, demonstrating a well-defined mass with specific signal characteristics and vascular associations but no significant lymph node involvement.

Histological examination post-surgical excision confirmed the diagnosis of FL, with areas of progression to diffuse large B-cell lymphoma. This case underscores the importance of histopathological analysis in the diagnosis of soft tissue masses, which is crucial for guiding appropriate therapeutic strategies. It also brings to light the importance of reviewing pathology slides in the medical field, because it ensures accurate diagnosis and treatment planning.

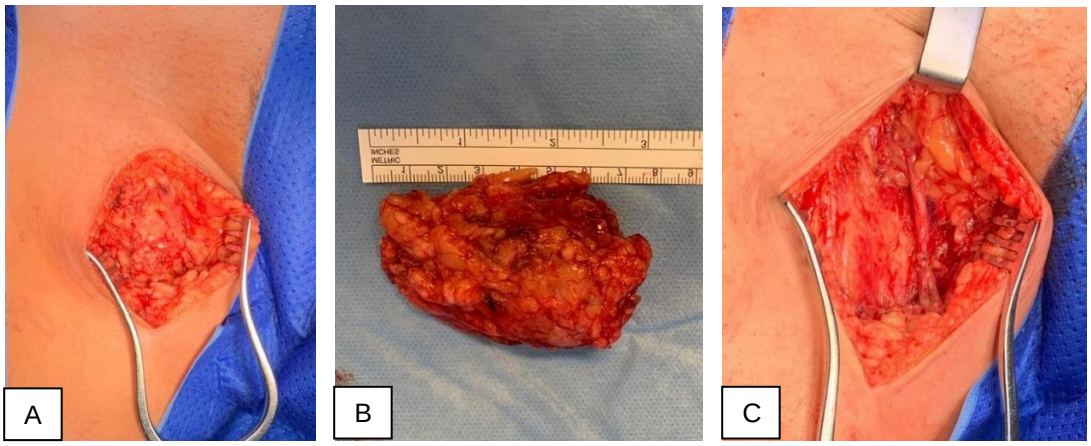

Fig. 1. Intra-operative images
A: Lesion In Situ During Surgery. This image shows the follicular lymphoma lesion in situ within the right thigh of the patient. The mass is well-defined and encapsulated. B: Excised Lesion. The excised follicular lymphoma mass, removed from the right thigh. The lesion appears well-circumscribed and homogenous. C: Surgical Site Post-Excision. The surgical site post-excision, demonstrating the area from which the follicular lymphoma mass was removed. The surrounding tissue and vascular structures are visible.

The subsequent management of this patient involved referral to oncology for chemotherapy with the R-CHOP regimen, a standard treatment for this disease. This case exemplifies the treatment course for localized FL and highlights the effectiveness of the R-CHOP protocol in inducing remission in patients without advanced disease.

FL accounts for approximately 20-30% of all non-Hodgkin lymphomas (NHL) in Western countries. Its etiology remains largely unclear, but several risk factors, including immunosuppression, autoimmune diseases, and viral infections (such as Epstein-Barr virus), have been associated with an increased risk of developing FL. Genetic abnormalities, particularly the t(14;18)(q32;q21) translocation leading to the overexpression of BCL2, play a significant role in the pathogenesis of FL by inhibiting apoptosis and contributing to the accumulation of malignant cells.[5]

The clinical behavior of FL is characterized by its indolent course with periods of spontaneous remission and relapse. While it is generally considered incurable with current therapies, recent advancements have significantly improved survival outcomes. The introduction of monoclonal antibodies, such as rituximab, has been particularly transformative, increasing the overall survival rates for FL patients.[2,8]

Despite its indolent nature, FL can undergo histological transformation to a more aggressive lymphoma, most commonly diffuse large B-cell lymphoma (DLBCL). This transformation occurs at a rate of approximately 2-3% per year and is associated with a poor prognosis. It underscores the need for vigilant long-term follow-up in patients with FL, even in those who achieve complete remission after initial treatment.[1]

The FL International Prognostic Index (FLIPI) and its revised version (FLIPI2) provide valuable tools for risk stratification and prognostication in FL. Factors such as age, disease stage, hemoglobin levels, number of nodal sites involved, and serum lactate dehydrogenase levels are integral to these indices and help guide treatment decisions.[7] However, the FLIPI was developed before the widespread use of rituximab and may not fully reflect the prognosis of patients treated with contemporary regimens.[7]

MRI remains the imaging modality of choice for evaluating soft tissue masses due to its superior contrast resolution and ability to delineate tumor extent and involvement of adjacent structures. In this case, MRI findings were pivotal in planning the surgical approach and ensuring complete excision of the mass. However, the definitive diagnosis relied on histopathological examination, highlighting the complementary roles of imaging and pathology in the diagnostic process.[6]

This case emphasizes the importance of considering FL in the differential diagnosis of soft tissue masses, particularly in adults. Although rare, extranodal FL should be part of the diagnostic consideration, especially in patients presenting with atypical symptoms. Multidisciplinary collaboration among surgeons, radiologists, pathologists, and oncologists is essential in managing such complex cases to achieve accurate diagnosis and optimal treatment outcomes.

4. CONCLUSION

In conclusion, this case report illustrates an unusual extranodal presentation of follicular lymphoma, manifesting as a slow-growing mass in the lower limb without systemic symptoms. The diagnosis was established through surgical excision and histopathological analysis, underscoring the critical role of tissue biopsy in the evaluation of soft tissue masses. The patient received standard chemotherapy with R-CHOP, reflecting current therapeutic practices for FL.

This case highlights several key points: the necessity of considering FL in the differential diagnosis of extranodal masses, the indispensable role of histopathology in confirming the diagnosis, and the importance of a multidisciplinary approach in managing complex cases. Moreover, it underscores the advancements in FL treatment, particularly the impact of monoclonal antibodies on improving patient outcomes.

Further research is warranted to enhance our understanding of FL's pathogenesis, identify reliable prognostic markers, and develop more effective treatment strategies. Vigilant long-term follow-up is crucial due to the potential for histological transformation and disease recurrence. This case contributes to the growing body of literature on the diverse clinical presentations of FL and provides valuable insights into its diagnosis and management.

ACKNOWLEDGEMENTS

The authors would like to thank the medical and surgical teams at the Department of General Surgery and Oncology, Unidade Local de Saúde da Guarda, for their support in managing this case.

COMPETING INTERESTS

The authors declare that no competing interests exist.

AUTHORS' CONTRIBUTIONS

Author A designed the study, wrote the first draft of the manuscript and managed the literature searches. All authors read and approved the final manuscript.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

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