Case Report

Primary diffuse large B-cell lymphoma of the corpora cavernosa presented as a perineal mass

González-Satué Carlos, Valverde Vilamala Ivanna, Tapia Melendo Gustavo[1], Areal Calama Joan, Sanchez Macias Javier, Ibarz Servio Luis

Department of Urology, [1]Pathology, Germans Trias i Pujol Hospital, Badalona, Barcelona, Spain

ABSTRACT

Primary male genital lymphomas may appear rarely in testis, and exceptionally in the penis and prostate, but there is not previous evidence of a lymphoma arising from the corpora cavernosa. We report the first case in the literature of a primary diffuse cell B lymphoma of the corpora cavernosa presented with low urinary tract symptoms, perineal pain and palpable mass. Diagnosis was based on trucut biopsy, histopathological studies and computed tomographic images.

Key words: Corpora cavernosa, lymphoma, perineum tumor. urinary obstruction

INTRODUCTION

Urologic extranodal lymphomas are infrequent, presenting less than 5% of primary extranodal lymphomas. Urologic lymphomas are mainly located in the kidney or the bladder.[1] In male, genital lymphomas are usually testicular, and rarely penile or prostatic. Corpora cavernosa lymphomas only appear in the literature as a secondary involvement of a primary lymphoma of the penis.[2] No evidence for any type of primary lymphoma to arise from the corpora cavernosa without penile involvement has been reported so far. Because extranodal lymphomas tend to behave aggressively, treatment of diffuse B-cell lymphomas is usually rituximab-based chemotherapy with local excision when possible.[3] Extern beam radiation may also be used in selected cases. Clinical presentation of perineal masses are not characteristic; therefore diagnosis is often performed too late.

For correspondence: Dr. González-Satué Carlos, Urology Department, Germans Trias I Pujol Hospital (9th floor). Crta canyet s/n, 08916, Badalona, Barcelona, Spain. E-mail: cgsatue@yahoo.es

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CASE REPORT

A 65-year-old man with pathological history of peripheral vasculopathy, Leriche syndrome, and bearing a cardiac pacemaker, presented a 3-month history of dysuria, weak urinary flow, progressive voiding discomfort, and perineal pain, when a perineal swelling that was slowly growing was detected. Physical examination showed normal external genitalia and a painful 4-cm perineoescrotal mass in contact with the bulbar urethra. Digital rectal examination detected a small and nonsuspicious prostate.

The patient was examined via ultrasonography. A 15-cc heterogeneous prostate was detected with 85 cc of postvoiding residual urine. Voiding cistourethrography showed a slight narrowing of the diameter of the bulbar urethra, but not a significant urethral stricture, suggesting an external compression of the urethra. PSA blood level was 0.56 ng/ml, erythrocyte sedimentation rate was 21 sec, and urinary culture was negative.

Multiple trucut transperineal biopsies of the mass were performed under 7 Mhz ultrasonography guidance. The histopathological examination of a core biopsy showed a diffuse, dense infiltrate of atypical large lymphoid cells with frequent mitotic figures [Figure 1]. Immunohistochemical studies showed the cells to be positive for B-cell marker (CD20) and negative for T-cell markers (CD3 and CD5), CD10, and BCL6. The proliferative index (Ki 67) was 70%. Based on these findings, a diagnosis of diffuse large B-cell lymphoma was delivered.

A thoracic-abdomino-pelvic computer tomography (CT) was

prescribed in order to search for a primary nodal origin of the lymphoma. Findings of the CT were as follows: absence of mesenteric, retroperitoneal, or pelvic significant nodes, at the CT sagital section, the anatomy of corpora cavernosa is replaced by a heterogeneous 6 × 4.5 cm mass [Figure 2].

In conclusion, the tumor was classified as IE stage diffuse B-cell lymphoma (single, extra-nodal, without B symptoms according to the Ann Arbor classification). A rituximab-combined chemotherapy R-CHOP (cyclophosphamide, adryamicine, vincristine, and prednisolone) was prescribed, to be administrated every 3 weeks for a total of six cycles. Surgical excision of the mass was not possible. Although the patient showed good clinical response, he died of cardiac toxicity after 8 months (six cycles of chemotherapy).

DISCUSSION

Primary malignant tumors of the corpora cavernosa are extremely rare. Only one case of perineal reticulum cell sarcoma can be found in the literature,[1] and it was described as a perineal mass with priapism. Malignancies of the corpora cavernosa often appear as a primary involvement of a tumour of the shaft of the penis, frequently advanced squamous carcinoma,[4] rarely lymphomas.[2] The common presentation in those cases is a palpable nodular mass in the shaft of the penis. Both advanced bladder[5] and prostate cancer may also display an involvement of the corpora cavernosa. However, the clinical presentation in these cases is "malignant" priapism.[5]

As far as the lymphomas of the low urinary tract and genitalia are concerned, the testis is the most common site of lymphomatous involvement of the male genitalia.[4] Diffuse large B-cell lymphomas are the most frequent lymphomas of male genitalia. Classical treatment of extranodal large B-cell lymphomas is rituximab-combined chemotherapy R-CHOP,[3] and 4000–4400 rads radiation is added in selected cases. In extranodal primary affection, surgical excision should be performed in small lesions when these are reachable,[2] but radical surgery and amputations should be avoided in low urinary tract lymphomas.

In the case presented here, surgical excision was not possible. The location and extension of the tumor conditioned nonsurgical treatment. Six cycles of rituximab-combined chemotherapy R-CHOP were prescribed. Radiation therapy was envisaged, but the length of the mass invited us to reserve radiation for the residual tumor after chemotherapy. The clinical response was successful with chemotherapy alone. Perineal swelling decreased and the urinary flow increased. However, heart toxicity of chemotherapy led to the patient's death 8 months after the diagnosis.

The origin primary diffuse large B-cell lymphoma of the corpora cavernosa is uncertain. Because there is no lymphoid tissue in the corpora cavernosa, it may be initially considered as a primary lymphoma. However, it could also be a clinical manifestation of hidden nodal disease. The CT excluded thoracic or abdominal primary origin. Magnetic resonance is the best radiological exploration in those cases, because it provides optimum information about tumor stage and infiltration of neighbor organs. However, magnetic resonance cannot be performed in patients bearing a cardiac pacemaker.

The absence of priapism in this case might be due to a severe peripheral vasculopathy and the Leriche syndrome. Other primary origin of this tumor could be considered, such as the prostate, the urethra, or the rectum. Compression of bulbar urethra showed up as low urinary tract syndrome, simulating a prostatic obstruction. Prostatic and rectal diseases were excluded by digital rectal examination, ultrasonography, and PSA. Voiding cistourethrography also excluded the urethra as the origin of the lymphoma.

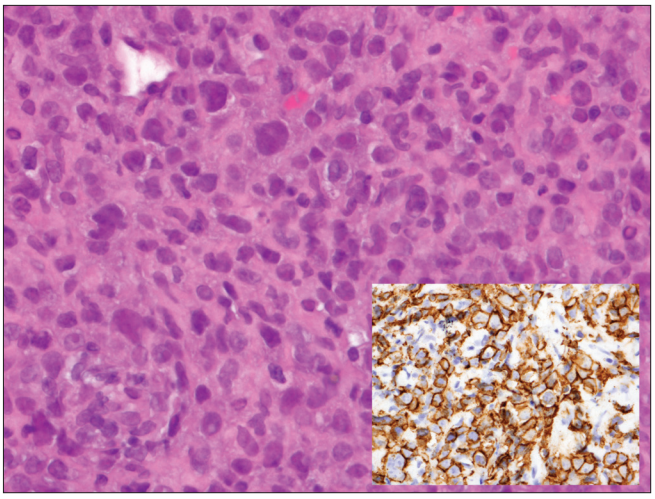

Figure 1: Diffuse infiltration of large, atypical lymphoid cells (H and E stain, ×400), strongly positive for CD20 (inset, ×400)

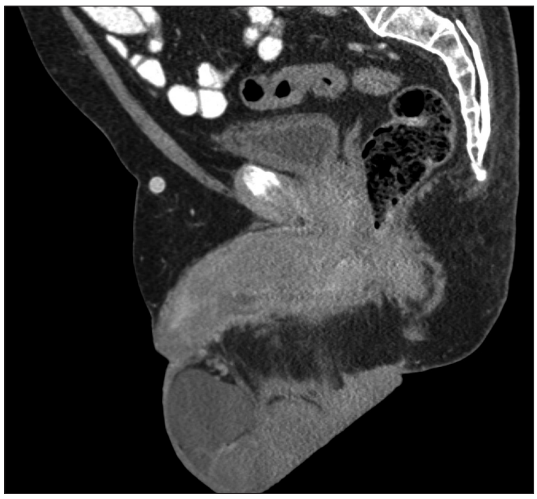

Figure 2: Sagital TC view: The anatomy of corpora cavernosa is replaced by a heterogeneous 6 × 4.5 cm mass

To our knowledge, this is the first case of lymphoma of the corpora cavernosa without penile involvement described in the medical literature. The relevance of this case resides in its exceptionality, its clinical presentation simulating a urethral stricture, the need of biopsy to provide an accurate the diagnosis, and the potential toxicity of the aggressive chemotherapy treatment used in these cases of lymphomas in aged population.

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