CLINICAL CASE

Bilateral testicular tumor: extranasal type NK/T-cell non-Hodgkin’s lymphoma


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Abstract  Lymphoma is a malignant neoplasia characterized by the proliferation of cells native to the lymphatic system. It makes up 1% to 9% of testicular neoplasia. Bilateral testicular tumor presentation was first described by Aberhouse in 1955. Extranodal nasal-type natural killer (NK) lymphoma is the most frequent variant of non-Hodgkin’s tumors and presents with extensive vascular destruction and necrosis; the most common extranodal site is the upper respiratory system. It is characterized by poor outcome and systemic dissemination and does not respond well to current treatments.

A 45-year-old man presented with bilateral increased testicular volume of 6-month progression, having received multiple treatments. Testicular ultrasound and abdominal computed tomography scan revealed increased testicular volume, retroperitoneal metastasis, and metastasis to the subcutaneous cell tissue. Tumor markers were negative. Right radical orchiectomy and biopsy of the left testis were performed. The definitive histopathologic diagnosis was extranasal-type NK/T-cell non-Hodgkin’s lymphoma. The patient was referred to the Hematology Service and chemotherapy and hormone replacement treatment were begun. This tumor represents 1% of all non-Hodgkin’s lymphomas and 5% of testicular masses. Mean presentation age is 50 years and there is a higher incidence in Asia and South America. Twenty percent of the cases affect areas outside of the nasal cavity. There is no staging classification for this disease. Five-year survival is 60% and 17% for disseminated disease. None of the current treatments (orchiectomy, chemotherapy and/or chemo-radiotherapy) are considered the criterion standard.
Introduction

Testicular cancer is the most common neoplasia in male adolescents and adults between the ages of 15 and 35 years; they have a 500 to 1,000-fold higher probability of developing a contralateral tumor. Bilateral tumors have been observed in 0.5% to 7% of patients, according to different case series studied.1

Lymphoma is a malignant neoplasia characterized by the proliferation of cells native to the lymphatic system and makes up 1% to 9% of testicular neoplasias. Bilateral testicular lymphoma presentation was first reported by Aberhouse in 1955. The majority of patients are above 50 years of age and there is a higher incidence between the ages of 60 and 70 years. Bilateral testicular lymphoma incidence is 19%, compared with the presentation of testicular germ cell tumors at 3%. Systemic symptoms present in 25% to 40% of the cases.2 The disease is prone to extragonadal metastases, especially to the central nervous system, skin, Waldeyer’s ring, bone, and lung. 3,4

Nasal natural killer (NK)/T-cell lymphoma is an uncommon form of non-Hodgkin’s lymphoma. And despite being rare, the extranodal NK/T-cell lymphoma nasal type is the most frequent variant.1,5-7 These tumors are characterized by extensive vascular destruction and prominent tissue necrosis. The most common extranodal site is the upper respiratory tract, especially the nasal cavity.

The most common confusion with other pathologies (i.e. chronic orchiopididymitis) can produce a delay in the diagnosis and treatment. NK/T-cell lymphoma nasal type is characterized by poor prognosis and systemic dissemination in a short time and it has a poor response to current treatments.

Case presentation

A 45-year-old man came to the outpatient service of the Urology Division of the Hospital Dr. Manuel Gea González in May 2013 for the clinical symptoms of increased bilateral testicular volume of 6-month progression that was painless. There was no improvement with previous antibiotic and antituberculosis treatment and he had a weight loss of 5 Kg in 6 months. Physical examination revealed a bilateral increase in testicular volume, primarily of the left testis; the right testis was indurated, as were both spermatic cords. Testicular ultrasound showed increased testicular volume with heterogeneous echo-architecture and hyperechogenic nodular areas with rounded hypoechoic lesions. The right testis measured 76 x 39 x 35 mm and the left testis 72 x 47 x 39 mm (fig. 1). Chest x-ray did not reveal lesions or widening of the mediastinum. Abdominal tomography scan showed a para-aortic lymph node conglomerate of 32 mm at its greatest diameter, 20 HU in the non-contrast phase enhanced to 34 HU in the contrasted phase, with inguinal adenopathies and thickening of the spermatic cord structures in bilateral form, as well as a bilateral increase in testicular volume with heterogeneous densities (fig. 2). Lactate dehydrogenase (LDH) was 148 U/L, human chorionic gonadotropin (hCG) was 0.14 mIU/mL, and alpha-fetoprotein (AFP) was 2.12 ng/mL (normal ranges). Right radical orchietomy + left testicular biopsy was performed with an intraoperative report of the spermatic cords and stroma, ending in left orchietomy.

The definitive histopathologic diagnosis was: right testis weighing 126 g, measuring 7.5 x 5.5 x 3.5 cm, slice showing multiple confluent, nodular lesions measuring 6.5 cm; left
testis weighing 186 g, multinodular, measuring 7 x 4.9 x 5 cm; extranasal NK/T cell non-Hodgkin lymphoma affecting 40% of the right testicular parenchyma and 50% of the left testicular parenchyma; neoplasia of angiocentric pattern with positive immunohistochemical development for the CD3+, CD56+, and CD45+ markers (figs. 3 and 4); stage IV Ann Arbor Classification (disseminated or diffuse infiltration of one or more extralymphatic sites with or without associated lymph node involvement or isolated extralymphatic organ involvement with distant node involvement. The patient was referred to the Hematology Service for treatment with chemotherapy and hormone replacement with testosterone.

**Discussion**

Primary lymphoma of the testis is an uncommon and little-reported entity that corresponds to 1% of all non-Hodgkin lymphomas and approximately 5% of all testicular masses. Cases of bilateral presentation are even rarer. The mean age for presentation is 50 years, with greater incidence in Asia, South America, Central America, and the indigenous Mexican populations. A low frequency of the HLA-A*0201 allele has been reported in these patients. The primary tumor is located outside the nasal cavity in 20% of the cases (the upper aerodigestive tract, skin, gastrointestinal tract, spleen, lung, brain, soft tissues, and testis). There is no specific classification for staging this type of lymphoma. They can be staged using the Ann Arbor Classification or through the International Prognostic Index (IPI): age, stage, LDH level, extra nodal sites, and general status.

Ultrasound reveals a hyperechoic pattern with an increase in intrallesional blood flow, regardless of tumor size, and striated hypoechoic bands with parallel hyperechoic lines radiating in the periphery.

Histologically, it has an angiocentric pattern, necrosis, and the expression of cells with the NK/T cell phenotype. Immunohistochemical study is essential for making the definitive diagnosis, the positive expression of CD56, CD3-
lymphoid system, in the brain, and the nerves and epithelial cells of the rete testis. At the histologic level, supported by immunohistochemistry, the differential diagnosis must be made, primarily with histiocytic sarcoma, myeloid sarcoma, and extranodal manifestations of myelocytic leukemia.7

Primary tumor of the testis has a 5-year survival of 60% and 17% for disseminated disease. There is no standard criterion for treatment, but the best options are orchiectomy, with chemotherapy (CHOP) and/or chemo-radiotherapy.8-9 In general, the clinical course is of poor outcome in the cases of systemic disease, with very little response to chemotherapy management. The present case of bilateral testicular tumor is added to the case series reported at our division of the Hospital General Dr. Manuel Gea Gonzalez within the time frame of 1989 and 2000, with 4 cases reported (4.6%, seminoma, 3 synchronous, and 1 metachronous).

Conclusions

The low incidence of extranasal NK/T cell non-Hodgkin’s lymphoma presenting in the testes makes it impossible to develop the diagnostic, classification, and treatment regimen criteria that would have an impact on the outcome of these patients. Follow-up of the few cases reported on in the literature will be extremely useful for establishing management protocols.

Conflict of interest

The authors declare that there is no conflict of interest.

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References