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► **To cite this version:**

Stefano Licci, Luca Morelli, Renato Covello. Primary mantle cell lymphoma of the testis. *Annals of Hematology*, 2010, 90 (4), pp.483-484. 10.1007/s00277-010-1049-3 . hal-00583547

HAL Id: hal-00583547

<https://hal.science/hal-00583547>

Submitted on 6 Apr 2011

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Primary mantle cell lymphoma of the testis

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Dear Editor,

A 45-year-old man presented with a 3-week history of right lower quadrant pain radiating to the right inguinal area and testis. Physical examination revealed an enlarged right testis. Ultrasound examination of the right testicle showed a hypoechoic mass involving almost the entire organ and consistent with a neoplastic lesion. Biochemical assays for alpha fetoprotein and beta human chorionic gonadotrophin were within normal limits. A right orchifuniculectomy was performed and histological examination revealed the neoplastic mass was represented by a diffuse proliferation of predominantly medium-sized monotonous lymphoid cells resembling lymphoblasts, with dispersed chromatin, scant cytoplasm and a high mitotic rate (25 per 10 high power field) (Fig. 1A-B). Semeniferous tubules appeared entrapped in the lymphomatous tissue, were immature and lacking spermatogenesis (Fig 1A-B). On immunohistochemistry, the neoplastic cells expressed CD45, CD20 (Fig. 1C), CD5, cyclin D1 (Fig. 1D), bcl-2, and, focally, CD43, with no expression of CD3, CD10, bcl-6, CD23, TdT, CD34, MUM1/IRF4 and CD138. The proliferation index measured by immunostaining with monoclonal antibody MIB1 (anti-Ki-67 nuclear antigen) was high with more than 60% of the cells staining positive. Morphological and immunohistochemical features were consistent with a mantle cell lymphoma (MCL), blastoid variant. A fluorescence in situ hybridization (FISH) revealed that more than 75% of the cells contained t(11;14). Biochemical and hematological tests were normal, and computed tomography scans of the head and neck, thorax, abdomen, and pelvis were unremarkable. A bilateral bone marrow biopsy revealed no infiltration by lymphoma. On the basis of all these findings, a diagnosis of primary MCL of the testis, blastoid variant, was made. The patient underwent localized radiotherapy and six cycles of CHOP chemotherapy (doxorubicin, cyclophosphamide, vincristine, prednisone). Currently, 10 months after completion of therapy, he continues to be healthy with no apparent manifestation of the disease by physical examination and total body computed tomography.

MCL is a B cell non Hodgkin lymphoma (NHL) of older adults, with a male preponderance. Most patients present with disseminated disease, with lymphadenopathy, often with splenomegaly, and frequent involvement of the bone marrow. A minority of patients present with extranodal disease, most often involving the gastrointestinal tract¹ and occasionally Waldeyer ring², but rarely involving other sites, such as the skin³ or the ocular adnexa⁴. Nearly all patients with extranodal presentation of MCL will be found to have lymphadenopathy or more widespread disease on staging.

However, cases of primary extranodal mantle cell lymphoma have been described. The most frequent site is represented by the gastrointestinal tract⁵, with the characteristic clinico-pathological presentation of a multiple lymphomatous polyposis. Other primary MCLs have been reported in the skin^{3,6}, the oral cavity⁷ and the conjunctiva⁸, but they can be considered extremely rare.

Primary testicular lymphoma represents 1-2% of all NHLs⁹. This entity is chiefly a disease of the elderly, with 85% of the cases occurring in patients older than 60 among whom it is the most common testicular tumour⁹. Almost all cases are diffuse large B cell lymphoma (DLBCL). Few cases of primary testis follicular lymphoma (FL) have been reported, mainly in children¹⁰, even though adults also rarely develop testicular FL¹⁰. A case of MCL has been previously described in the testis¹¹, but it represented a rare involvement of this organ in a patient with disseminated disease.

As far as we are aware, our case represents the first report in the English literature of primary MCL of the testis.

In cases of primary testis lymphoma which are not morphologically consistent with DLBCL, it is recommended to make a correct differential diagnosis among small to medium-sized B cell NHL. The distinction is important for therapeutic and prognostic implications, since MCL has a significantly worse outcome than other lymphomas, in particular primary testis FL, whose spread beyond the primary site is very unusual, with a consequent very good prognosis¹⁰.

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Figure legend

Fig. 1 The testicular neoplastic lesion was composed of a diffuse proliferation of medium-sized lymphoid cells resembling lymphoblasts, with dispersed chromatin and scant cytoplasm. A seminiferous tubule is entrapped in the lymphomatous tissue, and shows only immature intraluminal germinal cells (A, hematoxylin-eosin, original magnification 200x; B, hematoxylin-eosin, original magnification 630x). Neoplastic cells show a diffuse cytoplasmic positivity for CD20 (C, original magnification 200x) and a strong nuclear positivity for cyclin D1 (D, original magnification 200x).

