CASE REPORT - SUPPLEMENTARY MATERIALS

Primary B-cell lymphoblastic lymphoma of the testis

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DISCUSSION

Primary testicular lymphoma is a rare tumor which generally appears as a unilateral hard painless mass. In 10% of cases patients refer an acute pain because of bleeding inside the tumor.

Ultrasound usually shows either single homogeneously hypoechoic nodule or multifocal lesions of various sizes (mean lesion size 16 mm, range 8-26 mm are reported in literature); parallel hypoechoic lines radiating peripherally from mediastinum testis are often seen representing blood vessels crossing to the lesions and strongly positive at color-Doppler examination (1, 2).

In some cases it is possible to find more nodules with different size which are expression of a multifocal disease. Primary testicular lymphoma is diagnosed by histological assessment following radical orchiectomy, to provide histological tissue for diagnosis and eliminate primary tumor.

Surgical procedure, allows local tumor control and eliminates blood barrier of testis, thus increasing the efficacy of subsequent chemotherapy. Microscopically lymphomas are made of a predominant single-cell pattern. Often cells are small with less cytoplasm and higher nucleo-cytoplasmic ratio.

Gross lymphomas are described as a yellow-to-white mass with possible focal hemorrhage, solid and lobulated. They have a characteristic growth pattern: cells infiltrate diffusely intertubular spaces with recognizable tubular and intratubular precursor germ-cell neoplasia remnants. In some cases lymphomas may invade the seminiferous tubules; microscopically it means a leakage of normal basal regular alignment of intratubular germ cells (3, 4).

Immunohistochemical assay is fundamental to complete the description of the tumor.

The evaluation of markers, expressed on cells, have been used to differentiate diffuse B-cells lymphomas in three groups. First group is characterized by CD10 and/or Bcl6 positive, while MUM 1 is negative. Tumors with these immunohistochemical aspects have a germinal center B-cell-like phenotype and better prognosis. The other two groups are non-germinal center B-cell-like phenotype; they have a worse survival and cells are CD10 and /or Bcl6 negative and MUM1 positive. Lymphatic spread is most frequently to the para-aortic lymphnodes with systemic spread to the CNS (6%-16%), skin (0%-35%), Waldeyer's ring (5%), lung, pleura and soft tissue. For primary testicular disease there isn't a standardized treatment.

After orchifunicolectomy, patients usually undergo adjuvant chemotherapy.

The association of cyclophosphamide, doxorubicin, vincristine and prednisone was considered the most efficacious regimen (CHOP) in the last decades. Recently the CHOP regimen has been modified by the addiction of rituximab, an anti-CD20 monoclonal antibody; and called R-CHOP. The new treatment with rituximab allows a better control of neoplasia and a longer overall survival. Other chemotherapy regiment is called hyper-C-VAD (cyclophosphamide, vincristine, doxorubicin, and dexamethasone given as course A, followed by methotrexate and cytarabine given as course B) and it can be used in those cases with more aggressive aspects and an increased risk of progression of neoplasia as in the case we reported (3, 4).

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