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BRIEF REPORT

Histiocytic sarcoma with bladder involvement: Case report and literature review

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KEYWORDS
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Abstract We report an unusual case of histiocytic sarcoma with bladder involvement. An 80 year-old man with a previous history of diffuse large B-cell malignant lymphoma presented with hematuria and back pain. Serial urine cytologies revealed no urothelial malignant cells, but cystoscopy showed a large intravesical mass. The patient underwent transurethral resection (TUR) of the tumor. The bladder TUR specimen showed a widely infiltrating epithelioid neoplasm, with intense immunohistochemical positivity for CD45 and histiocytic markers (CD68, lysozime and fascin). Histopathological diagnosis was histiocytic sarcoma. As the patient's condition was progressively deteriorating, only palliative care was indicated and he died one month after TUR. Although histiocytic sarcoma can often be widespread at the time of diagnosis, to our knowledge, this is the first report of a case presenting with urinary symptoms. Histiocytic sarcoma can mimic many other malignant lesions, and only immunohistochemistry can define the tumor cells, allowing correct therapy. We discuss the differential diagnosis and possible associations.

PALABRAS CLAVE
Linfoma maligno;
Neoplasia histiocítica;
Sarcoma;
Tumores vesicales

Sarcoma histiocítico con afectación vesical: notificación de un caso y revisión de la bibliografía

Resumen Presentamos un caso infrecuente de sarcoma histiocítico con afectación vesical. Varón de 80 años con historia previa de linfoma maligno difuso de células B grandes que presenta hematuria y dolor de espalda. Citologías seriadas de orina no mostraron células uroteliales malignas, pero una citoscopia reveló una gran masa intravesical. Se practicó una resección transuretral del tumor. La resección transuretral de la vejiga mostró una neoplasia tipo epitelioide con fuerte expresión de CD45 y de marcadores histiocíticos tales como CD68 lisozima y fascina con diagnóstico de sarcoma histiocítico. Tras el estudio, el paciente se deterioró

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progresivamente indicándose solo cuidados paliativos, con exitus al mes de la cirugía. Aunque el sarcoma histiocítico suele estar en estadio avanzado en el momento del diagnóstico, creemos que este caso es el primero en comenzar con síntomas urinarios. El sarcoma histiocítico puede simular diversas lesiones neoplásicas, siendo la inmunohistoquímica necesaria para un diagnóstico correcto. Discutimos diagnósticos diferenciales y posibles asociaciones.

Introduction

Neoplasms originating from histiocytic and dendritic cells are very uncommon, representing less than 1% of all hematolymphoid neoplasms.[1] The most recent 2008 classification of hematolymphoid tumors has established clear-cut criteria for the diagnosis of these neoplasms.[2] Within this category we find Langerhans cell histiocytosis (LCH), histiocytic sarcoma (HC), follicular dendritic cell sarcoma (FDCS), interdigitating cell sarcoma (ICS), indeterminate dendritic cell sarcoma (IDCS) and fibroblastic reticular cell tumor (FRCT), with presumed different embryological origins. Although the old term histiocytic tumors persists, in fact most of them are not truly macrophagic, but originate rather from stem cell CD34+ precursors. Diagnosis of these tumors relies on immunohistochemistry (IHC), for they share morphological features with many other malignant tumors. Due to their infrequency, therapeutical experience is limited; in the literature there are isolated case reports or short case series. Interestingly, our literature review revealed many veterinary cases; this kind of tumor would appear to be much more frequent in animals, especially in dogs and cats.[3,4] We report a case of histiocytic sarcoma that presented with urinary symptoms and discuss the differential diagnoses and possible associations.

Case report

An 80-year-old Caucasian man presented with hematuria and back pain. He had a past history of diffuse large B cell lymphoma in 2006 with node and spleen involvement, for which he had been splenectomized and treated with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) and rituximab with a favorable response. Follow-ups at the Hematology Unit of Hospital Clínico San Carlos showed no recurrences. In February 2016 he presented with acute hematuria and back pain. Serial urine cytological samples showed no malignant urothelial cells and no further action was taken. Eight months later during a routine follow-up consultation he complained of weight loss and malaise. Computed tomography (CT) showed enlarged retroperitoneal lymph nodes. Positron emission tomography–computed tomography (PET-TC) did not show any increased glucose uptake in nodes, but revealed a suspicious lesion in the bladder. Cystoscopy revealed a large infiltrating ulcerative mass involving the bladder wall. Transurethral resection (TUR) of the mass obtained a fairly large amount of tumor tissue.

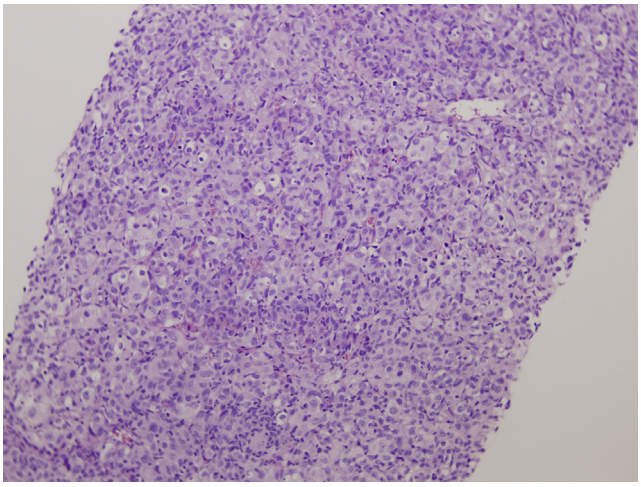

Figure 1 Low power view of the tumor cells showing clear cytoplasms and slightly pleomorphic nuclei (H-E 100×).

Histopathology showed a diffuse neoplasm, composed of medium sized cells with abundant clear cytoplasm and irregular and slightly pleomorphic nuclei (Fig. 1). Most cells had prominent nucleoli, but no melanin pigment was found. The overall picture was concordant with a clear cell carcinoma. The tumor cells widely infiltrated the muscular layer with vascular invasion. Immunohistochemistry for cytokeratin (wide spectrum AE1-AE3) was negative, as it was for GATA3 and epithelial membrane antigen (EMA). P63 was positive in 30% of the tumor cells. Subsequently we performed a much wider immunohistochemical panel, including melanocytic, lymphoid and muscular specific markers (namely, HMB45, Melan A, S100 protein, CD20, CD79a, BCL2, BCL6, CD10, CD3, desmin and actin) with negative results. The lack of specific IHC staining led us to review the H&E stained slides and we noted that some tumor cell nuclei showed a clear indentation (Fig. 2). For this reason we added immunohistochemical markers of hystiocytic differentiation, namely CD68 (EBM clone 11, Dako Denmark), which was intensely positive (Fig. 3). We then performed CD45, fascin, CD23, CD21, CD1a and lysozime to discard dendritic follicular cells or Langerhans cells origin. Only lysozime and fascin were positive and we could establish a definite diagnosis of histiocytic sarcoma. PCR was performed and confirmed rearrangement of IgH in the tumor.

Due to the advanced age of the patient and the clear and fast decline in his overall condition, no adjuvant therapy was given; he received palliative care only. He died one month after diagnosis.

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