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Case Report

Pelvic Clear Cell Adenocarcinoma in a Young Man: a Case Report

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Abstract

Clear cell adenocarcinoma is a relatively common neoplasm in the female genital tract, which can also occur in the bladder and urethra. In men, it is a rare neoplasm . We describe the case of a young man with large pelvic mass and diagnosed as clear cell adenocarcinoma. The patient had increased serum levels of CA 125 and carcinoembryonic antigen (CEA) . The tumor was immunohistochemically positive for CA 125 and CK7 and negative for PSA and CK20. Due to the size of the neoplasm at diagnosis and the morphological, immunohistochemical and clinical characteristics, the probable histogenetic origins of the tumor are prostate, prostatic urethra or seminal vesicle.

Keywords: Prostate, Prostate Neoplasms, urethra, Seminal vesicles

Introduction

Clear cell adenocarcinoma (CCA) of the urinary tract is a rare neoplasm occurring predominantly in the urethra or bladder of adult women,[1-3] having identical morphologic aspect to the tumor of same name originating in the female genital tract.[1] A still rarer tumor is CCA of the prostate.[4] Prostatic malignant neoplasms in children and young adults are uncommon and a great majority of them are of mesenchymal origin.[5]

We describe a case of CCA located in the pelvis of a young man that could have origin in the prostate, prostatic urethra or seminal vesicles.

Case Report

Clinical Characteristics

A 20-year-old youth, presented with a history of pain in the right flank for three months, accompanied with an increased volume of the bilateral inguinal lymph nodes. Patient sought medical service in the home town where a biopsy of the left inguinal lymph node was performed and the , diagnosis of metastatic adenocarcinoma of unknown primary site was done. Directed to our service for treatment, a histologic review of the case was carried out, confirming the same diagnosis after immunohistochemical study.

A computerized tomography of the pelvis showed a retrovesical mass (located at prostate and seminal vesicles) measuring 11.0 x 9.0 x 9.0cm with an extensive area of necrosis that displaced the bladder anteriorly and the rectum posteriorly, with determined urethral dilation to the right. The bladder presented

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smooth walls and homogeneous content. An upper abdominal computerized tomography reveale hepatic nodules compatible with metastases, retroperitoneal lymphadenomegaly, with left renal agenesis and right kidney moderate hydronephrotic (Figure 1).

The serum level for CA 125 was increased (916U/ml; normal <35Uml), as well as the serum

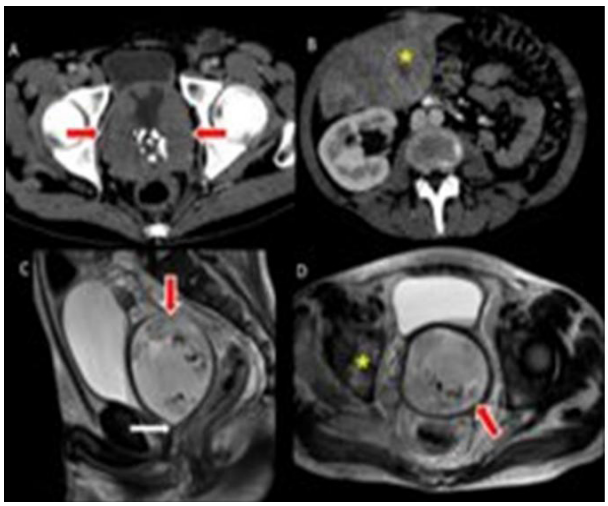

Figura 1- (A-D) – A) - Computerized tomography, axial plan, demonstrates heterogeneous retrovesical mass with central necrosis and rude calcifications (arrows). B) - Higher cuts identify left renal agenesis and hepatic metastases (*). C) - Magnetic resonance in the sagittal T2 plane, three months after the first exam and after discreet reduction of the lesion by chemotherapy effect, it demonstrates the eccentric location of the wide lesion (arrow) in relation to the prostate, in the interface among the fine utricle (arrow) and the seminal vesicles. D) - In the axial plan, the lesion (arrow) incurving the subsequent vesicle wall and right acetabular metastasis (*).

level for CEA (89.39ng/ml; normal <3.0ng/ml). The serum levels of PSA, CA 19.9 and alpha-phetoprotein were in normal levels.

Due to the necessity of primary site of neoplasm confirmation, as well as determination of histologic type, a biopsy of the pelvic mass guided by ultrasound was done; and diagnose as clear cell adenocarcinoma, with prostate, seminal vesicles or prostatic urethra as probable origin. In parallel, the patient received chemotherapy with carboplatine and taxol. Despite of the treatment, the patient evolved quickly with pyelonephritis and postrenal insufficiency due to right urethral obstruction by the tumor . Urinary derivation was not performed due to absence of clinical conditions and poor prognosis . However, he developed sepsis and died 7 months after the onset of symptoms.

Pathologic characteristics

The tumor was constituted by tubule-cystic glandular formations, covered by nonciliated cuboidal cells, with clear cytoplasm, In some cells the cytoplasm was eosinophilic. Hobnail type cells were a frequent ly found. Necrosis was also present. The nuclei exhibited moderate pleomorphism and presented prominent nucleoli. The mitotic index was 4/10 CGA. Psammous bodies and dystrophic calcifications were not found. Histologic characteristics were similar to those encountered in the inguinal lymph node biopsy (Figure 2).

Immunohistochemical study showed similar results in both biopsies tumor cells were strongly positive low molecular weight keratin 35BH11 (Dako, clone 35B, 1:800 dilution), cytokeratin 7 (Dako, CK-7, clone OV-TL 12/30, 1:900 dilution) and CA 125 (Dako, clone OC125 - mouse, 1:300 dilution). The neoplasm was weakly positive for keratin 34BE12 (Dako, clone 34BE12, 1:400 dilution) and negative for cytokeratin 20 (Dako, CK-20, clone Ks 20.8, 1:400 dilution), vimentin

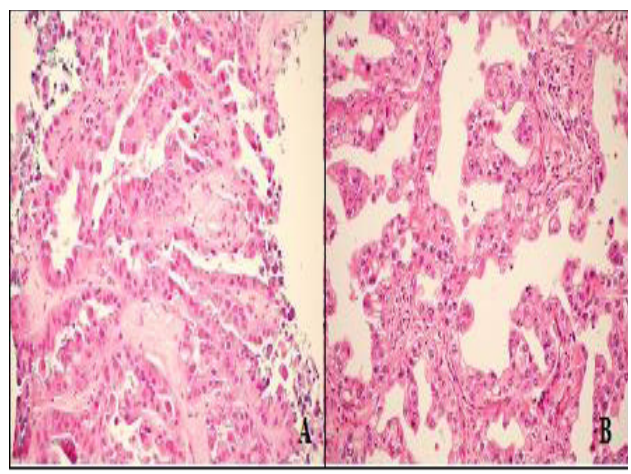

Figura 2 - (A-B) - Clear cell adenocarcinoma A) - Prostatic biopsy (HE, x 100). B) - Inguinal lymph node biopsy (HE, x 200).

(Dako, clone V9, 1:2000 dilution) and PSA (Biogenex, clone ErPr-8, 1:500 dilution) (Figure 3).

Discussion

Primary malignant neoplasm of the prostate in young adults (less than 40 years) when occurs, have mesenchymal origin been rhabdomyosarcomas and leiomyosarcomas[5] the most frequent histological

subtypes.Primary malignant neoplasm of the prostate gland in young adults, has been described exceptionally

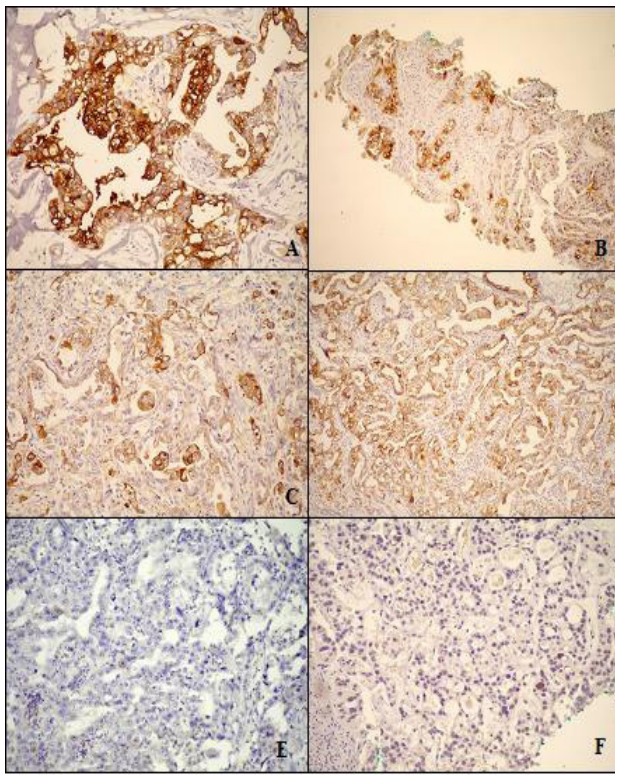

Figura 3- Clear cell adenocarcinoma: immunohistochemical staining A) - Inguinal lymph node biopsy (CA 125, x 400). B) - Prostatic biopsy (CA 125, x 40). C) - Inguinal lymph node biopsy (CK7, x 100). D) - Inguinal lymph node biopsy (34BE12, x 100). E) - Inguinal lymph node biopsy (PSA, x 100). F) - Prostatic biopsy (PSA, x 100).

in the literature,[6] as well as carcinoma of the seminal vesicle.[7-8] CCA is a relatively common epithelial neoplasm in the female genital tract, which can occur in the lower urinary tract. In males , this tumor can also affect the bladder and prostatic urethra and are extremely rare in the prostate,[4] mainly in young adults.[5] There is no report in the literature of CCA with origin in the seminal vesicle.

In our case, the patient presented a retrovesical mass with disseminated metastases. Due to the size of the neoplasm at diagnosis and the anatomical location in contact with several structures, it is not possible to assume that the neoplasm had primarily origin in the prostatic urethra, prostatic parenchyma or in the seminal vesicles. CCA originating in the lower urinary tract with prostatic origin presents a controversial histogenesis.There are some hypothesis of mesonephric origin, [9] Müllerian,[2,10-11] urothelial[12-13] or a malignant transformation of a nephrogenic adenoma in the case of CCA of the urinary tract.[14-15] CCA was initially assumed to arise in mesonephric duct remnants or intermediate mesodermic remnants near the vagina and was designated as "mesonephric adenocarcinoma". However, this hypothesis is little supported by the literature.[11] As for nephrogenic adenoma, there is a morphological similarity between this and CCA. However, nephrogenic adenoma is much more common in men and presents itself in different locations.[1]

The hypothesis of Müllerian origin for this neoplasm is supported by morphological similarities with ACC of the female genital tract coexisting or originating in , endometriosis and the strong positivity for CA 125, a known marker for Müllerian differentiation .[1-2,11,13] However, there are few reports showing coexistence of CCA and endometriosis and assuming that the CA125 expression is not specific for Müllerian differentiation.[1] Our case presents a very elevated serum level for CA 125, as well as immunohistochemical staining strongly positive for this marker.

The divergent differentiation is a well-known characteristic of urothelial neoplasms; a reason for which some authors sustain a urothelial origin for CCA.[1,13] Some studies evidenced the coexistence found in some CCAs with urothelium or urothelial carcinomas. This hypothesis is supported by the positivity for CK-7 and the negativity for CK-20 found in CCA, a common immunohistochemical expression also in the urothelial neoplasms.[1,13] A recent study using fluorescent *in situ* hybridization (FISH) techniques and analysis of X-chromosome inactivation of CCA in comparison with urothelial neoplasms and urothelium support this origin.[1]

Due to the location of the tumor, there is also a possibility neoplastic origin in the seminal vesicle. although there is no description of ACC originating in the seminal vesicle. Carcinomas originating in this site are also positive for CA 125 and CK7 and negative for CK20 and PSA.[7-8] This fact supports the hypothesis of acc originate from that location.Additionally, the elevated serum levels for CA 125 in carcinomas of this origin and the elevation of this marker has been correlated with disease progression.[7]

Another curiosity of this case is the fact that the patient presented renal agenesis. Gualco and colleagues [5] described a case of CCA of the prostatic utricle in a 16-year-old adolescent who also presented renal agenesis. The prostatic utricle has a mixed origin: the cranial portion derived from the Müllerian ducts, the caudal segment is derived from the Mullerian ducts, Wolffian and urogenital sinus.[5,16] Neoplasms of this

location can be related with urogenital malformations, such as hypospadia and renal agenesis.[5] Due to the size of the neoplasm , we cannot exclude the hypothesis of it having originated in the prostatic utricle and this has relation with its malformation. Nevertheless, this matter remains obscure and there is only speculation of a possible common pathological origin between CCA and renal agenesis.

In conclusion, we described a rare case of CCA in the pelvic region in a young man with left renal agenesis possibly originating in the prostate, prostatic urethra or seminal vesicle.

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