Clinical note

Morphological-metabolic evaluation of the urachal mucinous adenocarcinoma by positron emission tomography-computed tomography (PET-CT)∗


Servicio de PET-CT, Hospital Ángeles del Pedregal, México D.F., Mexico

abstract

The urachal mucinous adenocarcinoma is a rare malignant neoplasm located between the bladder and the umbilicus. It is usually found in an advanced stage at the moment of diagnosis. We have analyzed a clinical case in which the PET-TC study provided valuable morphological and metabolic information for diagnosis and staging.

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Introduction

The urachus is a midline embryonal remnant extending from the superior, anterior edge of the bladder to the umbilicus.1,2 Urachal tumors are very rare, representing less than 0.5% of bladder tumors and less than 0.01% of all adult neoplasms.2,3 PET-CT is a useful diagnostic imaging tool which allows adequate morphological characterization of the primary lesion, identification of distant lesions and provides metabolic information. All of this confers high diagnostic sensitivity and specificity and the ability to perform whole body staging. We present a case of urachal mucinous adenocarcinoma confirmed by histopathology and evaluated by PET-CT with 18FDG and multiphase contrast tomographic study with the aim of demonstrating the usefulness of this imaging method for the diagnosis and staging of this neoplastic disease.

Clinical case

A 50-year-old woman with a history of hysterectomy due to myomatosis 3 years previously and without other relevant pathological data was referred to our department for the presence of a mass in the lower hemiabdomen of 18 months of evolution. The mass had grown progressively until the development of cutaneous ulceration, with a fetid, greenish mucosal exudate associated with urinary frequency and the presence of mucous in the urine. PET-CT study with 18FDG demonstrated a pelvic abdominal mass with a maximum dimension of 16.3 cm with mixed metabolism, localized in the midline with extension and invasion of the abdominal wall and the urinary bladder associated with inguinal and hypermetabolic bilateral hip iliac adenopathies. It was determined to be grade IV-A according to the classification by Sheldon et al.4 Palliative chemotherapy was performed (Figs. 1–3).

Discussion

Incomplete regression of the urachus leads to multiple anomalies, with urachal cyst being the most frequent (30%). These anomalous structures are sheathed by transitional epithelium and it has been suggested that the etiopathogenesis of urachal carcinoma is related to metaplasia in the epithelial column following malignant transformation.2,3 Carcinoma of the urachus is very rare. Around 90% of the cases are well differentiated, albeit infiltrating, mucinous adenocarcinomas, and the remaining are transitional cells and squamous cell carcinomas.1 As in any other mucin producing tumor, there is a trend to contain psammomatous calcifications.3 The clinical manifestations include hematuria, dysuria, suprapubic mass with exudation of pus, blood or mucous. However, some symptoms may
be late with an elevated risk of metastasis because of the advanced stage of the tumor at the time of diagnosis. The prognosis is very poor, as in the present case. Metastases are generally produced in pelvic and inguinal lymph nodes as well as the lungs, brain, liver and bone in this order of frequency. In 1984, Sheldon et al. published a staging system according to the prognosis (Table 1).

Surgery is a potentially curative treatment in early stages. Radiotherapy is not considered since most of these tumors are radioresistant. Adjuvant and/or neoadjuvant chemotherapy provide minimum benefits, achieving objective response, although without improving the survival. With 50 years of experience Ashley et al. published a global 5-year survival of 49%, which varied according to the stage. They concluded that complete, early, extensive surgical resection with umbilicectomy is critical for patient survival. The primarily affected anatomical localization is the perivesical space anteriorly between the transversalis fascia and dorsally between the umbilicovesical fasia, also known as the Retzius space. From an imaging point of view it is important to consider the differential diagnoses of midline lesions of the abdominal wall among which the desmoid tumor, carcinomas of the urinary bladder, metastatic umbilical nodule and pathological neoplasms of the Meckel's diverticulum may be included.

Simple X-ray of the abdomen may identify single or multiple, lineal or curve-shaped calcifications localized in the median-infraumbilical region. Excretory urography rarely shows a repletion defect in the vesical dome in early stages. CT and MRI are diagnostic methods which allow characterization of the primary lesion and to thereby suggest the diagnosis and estimation of the stage. Thali-Schwab et al. described the morphological characteristics of this tumor in one of the largest series published including 25 patients. The average maximum tumor dimension was of 6 cm; 84% of the tumors demonstrated a mixed component (solid and liquid) and 16% were solid. Predominantly peripheral calcifications were observed in 72%. Invasion of the vesical wall was found in 92%, but the greatest tumoral volume was extravesical in 88%. Metastasis was present in 32%. PET-CT allowed morphometabolic characterization of the primary lesion and identified distant hypermetabolic

<table>
<thead>
<tr>
<th>Stage</th>
<th>Characteristics</th>
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<tbody>
<tr>
<td>I</td>
<td>Tumor limited to vesico-urachal mucosa</td>
</tr>
<tr>
<td>II</td>
<td>Tumor invaded the submucous or muscularis but confined to the urachus</td>
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<tr>
<td>III</td>
<td>Invasion beyond the urachus</td>
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<td></td>
<td>A. Extension to bladder</td>
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<td></td>
<td>B. Extension to abdominal wall</td>
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<td></td>
<td>C. Extension to peritoneum</td>
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<td></td>
<td>D. Extension to local viscera</td>
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<tr>
<td>IV</td>
<td>Metástasis</td>
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<td></td>
<td>A. Metastasis to regional lymph nodes</td>
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<td>B. Distant metastasis</td>
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Fig. 1. Picture of the infraumbilical abdominal mass. Cutaneous ulcerations may be seen with exudation of hemorrhagic and mucosal material.

Fig. 2. Axial CT images in the venous phase (a1, b1), PET (a2, b2) and fused PET-CT (a3, b3). A lobulated, vascularized mass with hypodense and hypometabolic zones is observed suggestive of necrosis and/or mucinous content and zones of reinforcement and hypermetabolism related to viable malignant tumoral tissue (arrows in a1, a2 and a3). Hypermetabolic inguinal adenopathies are shown (arrows in b1, b2 and b3). The discontinuous arrows in a3 indicate ureters.
lesions which translated into greater diagnostic precision and reliable staging, in addition to being promising for other subsequent indications such as evaluation of response to treatment and follow-up. In the present case, multiplanar tomographic evaluation allowed determination of the mixed component of the lesion (zones with attenuation of liquid), defined the regional extension and demonstrated that the greater tumoral volume was extravesical, which is useful for differentiation from a vesical carcinoma in which the mass is predominantly intravesical. Likewise, absence of dilatation of the intestinal loops and perforation of the hollow viscus ruled out Meckel’s diverticulum. On the other hand, the desmoid tumor is a benign fibromatous process in solid and homogeneous scar sites which compresses rather than invades structures. Metabolic characterization confirmed the malignant nature of the lesion and the presence of hypermetabolic metastasis to regional lymph nodes. It is well known that urinary excretion of $^{18}$FDG conditions high activity in the bladder, interfering with correct evaluation of the neoplastic disease in the pelvis, mainly that confined to the bladder or predominantly intravesical.

Multiple procedures have been developed to palliate this interference, with the technique of retrograde vesical filling showing good results and improvement in diagnostic precision.\textsuperscript{10} Since there are no reports on the use of PET-CT in this type of neoplasm, we considered it important to present this clinical case to favor the evidence and scientific knowledge of future reports.

References