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Neuroendocrine Carcinoma Associated to Mucinous Urachal Adenocarcinoma - Case Report

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Abstract

Malignant urachal transformation is extremely rare, representing less than 1% of bladder tumors. Adenocarcinomas are the most frequent subtype. Mucinous urachal tumors are far less common. The most usual symptom of urachal tumors is hematuria due to erosion of the vesical dome. Standard treatment is surgery. This is a case report about a 63-year-old patient diagnosed with a urachal tumor and the conduct after diagnosis with follow-up until the patient's death.

Keywords: Malignantneoplasia; Urachus; Surgical treatment

Introduction

The urachus is a fibrous remnant ofthe urogenital sinus and the allantois, which connects the vesical dome to the umbilicus. It is found in the middle line between the posterior rectus sheath, anteriorly, and the peritoneum posteriorly. Clinically, it remains undetectable, except if it becomes symptomatic or is identified through imaging [1-4].

Involution of the urachus occurs in the antenatal period, between the 6th and 12th week. However, complete obliteration may not occur, forming cysts in the urachus– more frequent - or patenturachus. Remaining tracescan also suffer metaplasia and the formation of tumors [1,5].

Malignant urachal transformation is quite rare, representing less than 1% of bladder tumors, and is more commonly found in males. Adenocarcinomas are the most frequentamong the subtypes. Urachal mucinous tumors are much less common and can be the cause of peritoneal pseudomyxomas [1,3,6]. The most common symptom of urachal tumors is hematuria due to the erosion of the vesical dome. Standard treatment is surgery, preferably partial cystectomy, associated to pelvic bilateral lymphadenectomy [3,7].

The present study aims at reporting the case of a patient with a urachal lesion, from diagnosis to surgical treatment and conduct after the anatomopathological findings.

Case Presentation

A male patient, 63 years of age, smoked, began with about 4 months of urinary storage symptoms, associated to macroscopic hematuria, when he sought medical attention. The kidney and urinary tract ultrasound showed: focal parietal thickening of the bladder, parietal nodular formation on the right lateral wall measuring 1.6 x 0.8 cm, formation of a cystic aspect and thickened walls adjacent to the vesical dome extending towards the umbilicus, measuring around 10 x 5.5 x 3.8 cm. Due to these findings he was submitted to a contrast-enhanced abdominal and pelvic CT-scan, which showed an elongated cystic image with parietal calcifications based on the vesicle dome (Figure1).

A bulge was observed in the vesical dome during the cystoscopy that appeared to be extrinsic compression without signs of lesions in the vesical mucosa. A resection of this area was carried out to obtain a sample for an anatomopathological study and results were conclusive only after the immunohistochemical study: malignant neoplasia of small cells disposed in blocks extending below the typically scanty urothelium.

A block resection of the urachal tumor was performedwith partial cystectomy and a bilateral pelvic lymphadenectomy, limited by the bifurcation of the iliac vessels (Figure 2-5). There was satisfactory and uneventful progress in the postoperative phase. The anatomopathological exam showed a solid undifferentiated predominant carcinoma (90%) with an associated mucinous adenocarcinoma (10%), involving the perivesical adipose tissue, perineural invasion with margins affected in extent and depth; 10 lymphnodes free of lesions; staging pT3N0. The immune-histochemical study revealed

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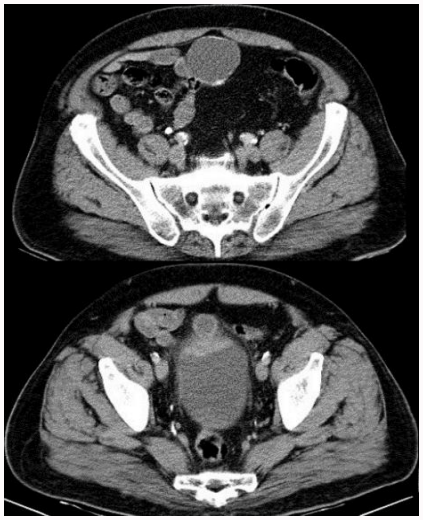

Figure 1: Pre-operative CT scan.

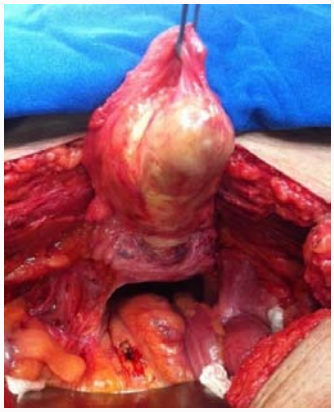

Figure 2: Lesion Identification.

a neuroendocrine invasive bladder carcinoma (negative CDX 2 and p63; positive CK7, CK20, cromogranine, Ki67) with a high rate of cellular proliferation. The systemic staging did not exhibit lesions, and adjuvant radiotherapy and chemotherapy (with cisplatin and etoposide) was indicated.

The patient had an irregular follow-up, only finishing radiotherapy and not the full chemotherapy regimen. Around 18 months after the surgical procedure, he was admitted to the emergency room with an acute intestinal obstruction. An exploratory laparotomy was performed and the findings suggested a peritoneal carcinomatosis, confirmed anatomopathologically. A loop ileostomy was performed with palliative care procedures defined thereafter. The patient subsequently died about one month after the second approach.

Discussion

Malignant urachal neoplasias are rare, with very few cases reported, involving mainly men between 50 and 70 years of age [3,4]. In most cases – up to80% of the time the lesion is found in the junction with the vesical dome [3,4,7]. The most common symptom is hematuria (90% of the cases), followed by suprapubic pain, irritative symptoms and mucosuria [4,8].

Initial imaging is usually ultrasound, which can suggest the diagnosis with a complex lesion found in the middle line in contact

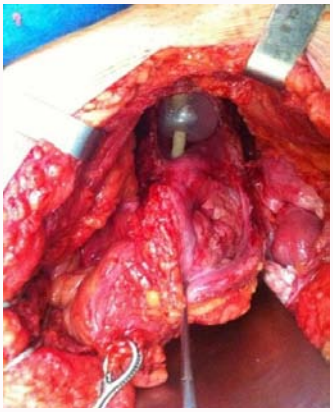

Figure 3: Beginning of lesion resection.

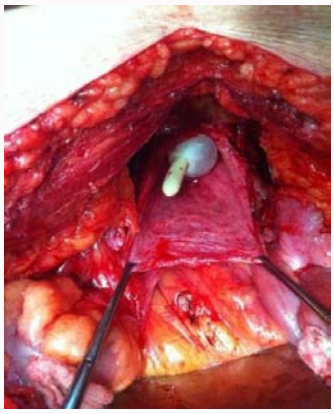

Figure 4: Bladder after resection.

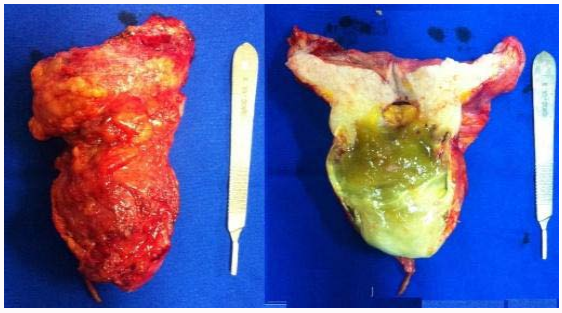

Figure 5: Surgical specimen after removal and its opening.

with the bladder. However, CT-scans, and mainly, MRIs, provide better information [1,4]. Cystoscopy is usually indicated to evaluate the bladder, where lesions or bulging of the dome are commonly found and a resection is performed to obtain material for a pathological study and more accurate diagnosis [4,7].

The most frequent histological type is the adenocarcinoma, mostlymucinous, but transitional cell as well as squamous cell neoplasias and sarcomas can also be found [3,4]. There is an important structural similarity between urachal and intestinal adenocarcinomas, including the production of the carcinoembryonic antigen, and the origin of the lesion is frequently defined (primary or secondary) after an immunohistochemical study [3,4,8].

The gold standard treatment for malignant urachal tumors, whenever possible, is partial surgical block resection of the

bladder with the lesion up to the navel associated to bilateral pelvic lymphadenectomy [3,4,7,8]. The open approach is more frequent, although the procedure can be successfully performed laparoscopically or also robotically, in spite of less long-term study outcomes [8-10]. The prognosis for patients with adenocarcinomas depends directly on resections with free margins and absence of positive lymph nodes [4,8,11].

Currently, no standard protocols are available for adjuvant treatment of locally advanced or metastatic urachal tumors. In general, supplementary treatment considers the experience of the assisting medical team that many times uses its knowledge of other types of tumors with similar behavior-especially those in intestinal neoplasia [4,8].

Neuroendocrine tumor of the bladder is an extremely rare neoplasm and does not yet have standard treatment, being extrapolated treatments of neoplasms with similar histopathological findings for its treatment [12]. The neuroendocrine tumor of the urachus is even rarer and, like tumors of similar histology of bladder origin, its treatment is done with extrapolation of tumors from other sites [13].

Chemotherapy similar to that indicated for gastrointestinal tumors is frequently used-based on cisplatin and the FOLFOX regimen [14-16]. In cases like ours, with a predominantly neuroendocrine variant, some defend the use of chemotherapeutic agents like those used in small cell lung tumors [14].

An adjuvant regimen resembling what was done with patients who had endocrine intestinal neoplasia as per the service protocol was used in the case reported.

A urachal neuroendocrine tumor is an extremely aggressive variant and progression of the disease occurs even with adjuvant therapy. Ebara et al recently showed the use of gemcitabine, cisplatin and paclitaxel afterprogression of local and systemic disease following radical cystectomy and obtained a good response [14]. Ismaili observed that the best kind of therapy is multimodal, beginning with a resection. He also showed that even the use of combination therapies and a close follow-up the prognosis is still poor [17].

Conclusion

We report a rare case of neuroendocrine tumor of the urachus and how it was conducted to try to some add informationto the poor literature in these cases. This rare neoplasm does not have adjuvant therapy protocols and should be individualized according to each patient and experience of the assistant team. In addition, we suggest that in similar cases the patient be strictly monitored due to aggressive behavior of this tumor.

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