CYSTOADENOCARCINOMA OF THE URACHUS

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Abstract

The cystoadenoma of the urachus is an uncommon disease which should evoked in case of a upper vesical (supra vesical) cystic mass. Many authors recommended total cystectomy but recent reports considered that conservative surgical treatment improved the quality of life without modifying the survival rate.

Key words: cystoadenocarcinoma - urchus - blader cancer - conservative surgical treatment.

Resumen

El cistoadenocarcinoma de uraco es una enfermedad poco frecuente que debe ser tenida en cuenta en presencia de una masa quística supravesical. Muchos autores recomiendan la cistectomía total, pero recientes reportes consideran que el tratamiento quirúrgico conservador, mejora la calidad de vida sin modificar la tasa de sobrevida.

Palabras Claves: Cistoadenocarcinoma - Uraco - Cáncer de vejiga - Tratamiento quirúrgico conservador.

Introduction

Adenocarcinoma of the urachus is a rare form of bladder cancer. We report a case urachal cancer and we review the diagnostic and therapeutic details of this non-so well-known disease.

Case report

A 56 year-old caucasian man, with no medical history was hospitalized for an endoscopic removal of a left ureteral stone. On this occasion, CT scan revealed a 6 cm upper vesical cystc mass (PHOTO N° 1). Its contents was of a liquid density with several areas of solid structure. The CA 19-9 was at 46.3 U/ml (normal range <37 U/ml). Laparoscopic surgical approach, showed a preperitoneal mass developed from the urachus. Cautious inspection of the peritoneal cavity was normal. Disecction of the antero-inferior part of the mass difficult and complicated by a bladder perforation. The intermediary of a protective bag carried out the extraction of both tumour and part of external wall of the bladder, by a 3 cm Phannestiel incision.

Pathologic examination showed a unilocular cyst filled with thick mucoid viscous fluid. The internal surface was smooth without any papillary structure. Histologically, the wall consisted of a fibrous layer surrounded by smooth muscle fascicles develop from the external surface. The internal surface consisted of a mucinous epithelium exhibiting varyng degree of differentiation (PHOTO N° 2). In some areas, the epithelium appeared more as mucinous cystoadenoma (PHOTO N° 3) and in others, it was more a mucinous cystoadenocarcinoma with nuclear atypia and lossed polarity (PHOTO N° 4). The wall was not infiltrated by the carcinoma.

The patient had an uneventfull postoperative course and chemotherapy with 5-fluorouracil 300 mg and cysplatyl was begun at the 30th postoperative day. Because of the small focus of adenocarcinomatous trasformation on histopathological examination, we decided not to complete the surgery.

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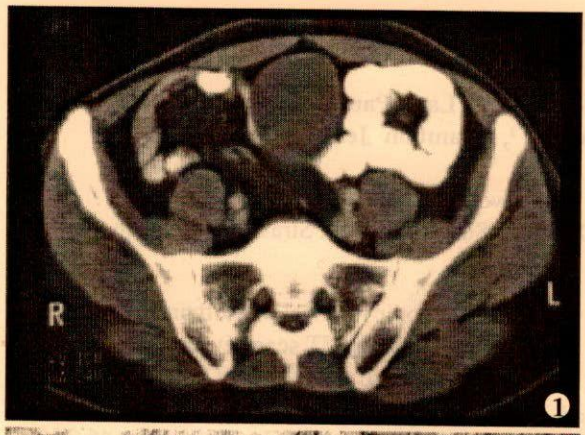

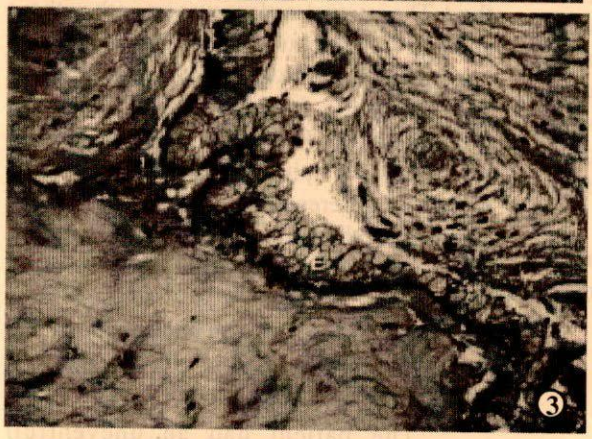

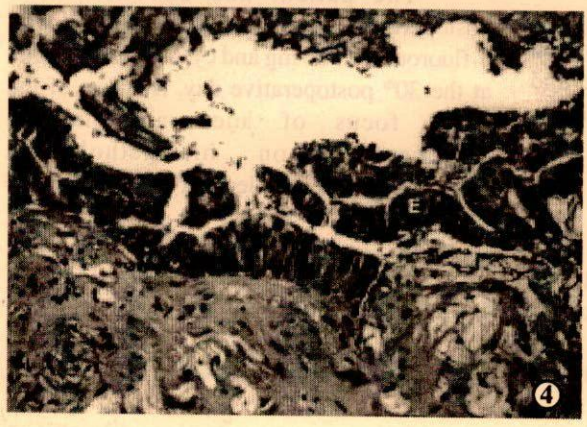

PHOTO N° 1: Abdominal C.T. shows a supra vesical cystic mass.

PHOTO N° 2: Histological examination: The wall made with a fibrous layer connectiv tissue (F) with some linear calcification (C). The internal surface was linead by an internal type epithelium (E) without atypia (HE x 6).

PHOTO N° 3: Part of the tumor with an appearance of mucinous cystoadenoma (E). The epithelial cells structure: basal nuclei and supranuclear mucin vesicle (HE x 50).

PHOTO N° 4: Typical aspects of cystoadenocarcinoma. The epithelial cells nuclear atypia (large and hyperchromatic nuclei) with loss of polarity (E).

Discussion

Mucinous cystoadenoma of the urachus was described by Begg as a particular anatomo-clinical entity (1). However, most authors consider cancer of the urachus like an uncommon type of vesical cancer. Mucinous adenocarcinomas represent 85 % of urachal cancers (2). Two theories explain the development of mucinous adenocarcinomas. The first based on the adenocarcinomatous transformation from glandular metaplasy of urothelial cells (3,4). The second theory suggests that the common embryological origin of the bladder and the rectum leads sometimes to the inclusion of intestinal cell in the urachus (7,9). The differentiation of the cells leads to the formation of a mucosecretive epithelium.

The tumour is highly invasive and diagnosis is often revealed by urological signs (5): hematuria is usual (75 % of the cases). Patients may also present mictional troubles by compresion or irritation of the bladder (50 % of the cases). Mucusuria, which is considered as pathognomonic, is common. Pelvic heaviness or palpation of a hypogastric mass sometimes leads to diagnosis. Calcifications on the X ray are frequent.

The principal locations are in the bladder dome and the anterior surface of the bladder. These tumours are exceptionally confined to the urachus but some case have been described (7). Concerning postoperative recurrences, most authors underline the frequency of local recurrence, in 40-50 % of case (7,8). By decreased order, lymph nodes locations (30%), lung and peritoneal metastases are found (8).

Treatment always surgical. There is controversy between the supporters of radical treatment and those who advocate conservative surgical treatment. Some authors, propose a total cystectomy associated with tumorectomy for urachal cancers (8,10). They underline the frecuency of local recurrence and the frequent and unexpectedly wide and depp vesical infiltration (8). Others authors prefer partial cystectomy with umbilicalectomy associated with tumorectomy. This conservative treatment did not modify the 5-year survival rate but has proved to decrease postoperative morbility and improved quality of life (7). The 5-year survival rate of urachal cancer is 40 %. Chemotherapy and radiotherapy do not seem to be efficient even if long-term survival after irradiation has been described (6).

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