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Presenting a case of a mucinous adenocarcinoma of an exstrophic bladder in an adult patient and a review of literature

Giovanni Di Lauro¹, Fabrizio Iacono², Antonio Ruffo^{2*}, Leo Romis¹, Salvatore Mordente¹, Umberto Pane¹, Ester Illiano², Giuseppe Romeo², Domenico Prezioso², Bruno Amato³

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Abstract

Background: Bladder exstrophy occurs in approximately 1 in 35,000 live births and is associated with an increased incidence of bladder cancer.

The primary mucinous adenocarcinoma of the bladder is an extremely rare urologic entity, which is found in less than 2% of all urinary bladder tumours and is often presented as metastatic. This is the first case in literature of a primary mucinous adenocarcinoma of an unreconstructed exstrophic bladder.

Case presentation: A 55-year old male patient was diagnosed with a primary mucinous adenocarcinoma of an unreconstructed exstrophic bladder. Examination of the entire gastrointestinal tract shown there were not other primary sites. Immunohistochemistry confirmed the nature of the tumour. The patient underwent a radical cystoprostatectomy with en block bilateral pelvic lymphadenectomy, urinary diversion with a cutaneous ureterostomy and epididymis repair.

Conclusion: Adult bladder exstrophy and epispadias correction is a very rare practice in urology due the fact that this congenital disease is diagnosed and corrected in neonates. We advocate the radical surgical management, after exclusion of any primary malignant sites related to the gastrointestinal tract.

Background

Bladder exstrophy is a complex congenital abnormality and its management is normally done in neonates.

Reports on exstrophy patients presenting in adulthood are very rare. Bladder exstrophy occurs in approximately 1 in 35,000 live births and is associated with an increased incidence of bladder cancer. Urinary bladder cancer is the second most frequent tumour of the genitourinary tract [1]. Adenocarcinomas of the bladder is an uncommon malignant neoplasm and account for less than 2% of all bladder cancers [2].

Few series have been reported in literature, but most contain small numbers of cases.

One of the most common forms of adenocarcinoma of the bladder is the metastatic adenocarcinoma. The primary sites for these tumours include the rectum, stomach, endometrium, breast, prostate, and ovaries.

The reported incidence among exstrophy cohorts varies from 3.3% to 7.5%, which is several times greater than in age-matched controls in the general population [3], [4].

In contrast, treatment of bladder exstrophy in patients presenting in adulthood is rarely reported in the literature [5], [6], [7], [8], [9]. Delayed presentation may be due to lack of awareness, ignorance, social embarrassment or even lack of appropriate facilities.

The etiology of this increased incidence remains unclear. The environmental stimuli and chronic infection may cause bladder cells to undergo glandular metaplasia [10]. After excluding patients who underwent

* Correspondence: antonio.ruffo7@gmail.com

²Department of Urology, University "Federico II" of Naples, Via Pansini, 5, 80131, Naples, Italy

Full list of author information is available at the end of the article

reconstruction, only around 113 cases of cancer occurring in exstrophic bladders have been reported thus far. Approximately 90% were adenocarcinomas and 5% were squamous cell carcinomas[3], [4], [10], [11]. These cancers tend to be aggressive, and most patients undergo radical cystectomy as primary therapy[3], [4].

There are a variety of histological types among adenocarcinomas, including papillary, glandular, adenoid cystic, clear cell, mucinous and signet-ring cell carcinomas. The last of these types is characterized by the presence of intracellular vacuoles filled with mucin that displace the hyperchromatic nuclei [12]. This patient was diagnosed with the mucinous and signet-ring cell subtypes that is very rare and correspond to 20% of the bladder adenocarcinoma [12],[13].

Methods

A 55-year-old man with classic bladder exstrophy who had never undergone a reconstruction earlier presented with a lesion of all his bladder [Figure 1]. The patient was otherwise healthy. His renal function tests were normal. The intravenous pyelogram showed bilateral normal excretory kidneys with no cystogram phase. The characteristic skeletal defects of exstrophy complex such as widening of the pubic symphysis caused by malrotation of the innominate bones in relation to the sagittal plane of the body along both sacroiliac joints were present. The metastatic work up was negative as seen on the CT scan. No preoperative bladder biopsy were performed.

We advocate the radical surgical management, after exclusion of any primary malignant sites related to the gastrointestinal tract.

The patient underwent radical cystectomy, closure of the infra-umbilical defect with Cardiff repair with Onlay mesh repair [14] and epispadias repair [Figure 3 - 4]

A wide local lymphadenectomy was performed [Figure 5].

Abdominal wall closure was done with a fasciocutaneous M-plasty [Figure 6].

Osteotomy was not done.

The orientation of the tumor on final clinical and pathologic examination in our patient showed that the bladder of 11x8 cm was completely occupied by a nodular, grey lesion of a maximum diameter of 10 cm producing mucin [Figure 2], thus strongly suggesting the bladder as the primary source.

Results and discussion

Histopathological examination revealed a G3 poorly differentiated (High grade) mucin-producing adenocarcinoma, with intracellular and extracellular production of mucin. The lymph nodes examined were negative showing a *reactive lymphoid hyperplasia*. The skin margins were not involved. The patient was not given any

adjuvant therapy and is doing well at six months of follow-up.

Other primary sites for the tumor had been excluded and, in the absence of digestive tract tumor and for confirmation that it was a primary bladder tumor, an immunohistochemistry study was performed.

Regarding immunohistochemistry, adenocarcinoma of the urinary bladder expresses CEA, CDX-2, MUC-1, MUC-2 and MUC-3, same as colonic adenocarcinoma. Cytokeratins 7 and 20 were positive, in contrast with colonic adenocarcinoma that expresses cytokeratin 20 but not cytokeratin 7 [15].

Conclusions

Bladder exstrophy and epispadia is a very rare congenital disease in urology.

Because the vast majority of patients with bladder exstrophy now undergo reconstruction at a young age, unreconstructed bladder exstrophy in an adult patient is exceedingly rare.

We advocate the radical surgical management, after exclusion of any primary malignant sites related to the gastrointestinal tract.

With proper surgical management of late presenting bladder exstrophy patients, an acceptable functional and cosmetic outcome can be achieved.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

GdL: conception and design, interpretation of data, given final approval of the version to be published. AR: acquisition of data, drafting the manuscript, given final approval of the version to be published. LR: acquisition of data, drafting the manuscript, given final approval of the version to be published. SM: acquisition of data, drafting the manuscript, given final approval of the version to be published. UP: critical revision, interpretation of data, given final approval of the version to be published. FI: critical revision, interpretation of data, given final approval of the version to be published. El: critical revision, interpretation of data, given final approval of the version to be published. DP: critical revision, interpretation of data, given final approval of the version to be published. GR: critical revision, interpretation of data, given final approval of the version to be published. BA: critical revision, interpretation of data, given final approval of the version to be published.

Authors' information

GdL : Consultant in Urology at Hospital Santa Maria delle grazie, Pozzuoli, Naples, Italy. AR : Resident in Urology at Federico II University, Naples, Italy. LR : Resident in Urology at Hospital Santa Maria delle grazie, Pozzuoli, Naples, Italy. SM : Resident in Urology at Hospital Santa Maria delle grazie, Pozzuoli, Naples, Italy. UP : Resident at Hospital Santa Maria delle grazie, Pozzuoli, Naples, Italy. FI : Associate Professor of Urology at Federico II University, Naples, Italy. GR : Resident in Urology at Federico II University, Naples, Italy. El : Resident in Urology at Federico II University, Naples, Italy. DP : Associate Professor of Urology at Federico II University, Naples, Italy. BA : Associate Professor of Surgery at Federico II University, Naples, Italy.

Declarations

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Authors' details

¹Hospital Santa Maria delle grazie, Via Domitiana località La Schiana - 80078 - Pozzuoli, Naples, Italy. ²Department of Urology, University "Federico II" of Naples, Via Pansini, 5, 80131, Naples, Italy. ³Department of General, Geriatric, Oncologic Surgery and Advanced Technologies, University "Federico II" of Naples, Via Pansini, 5, 80131, Naples, Italy.

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