

Oncologic Concerns in An Exstrophied Urinary Bladder - An Indian Scenario

S SHWETHA¹, HEMANTH S GHALIGE², LOVE GOYAL³, PREETY JAIN⁴, FAKHRUDDIN⁵

ABSTRACT

Exstrophy of the urinary bladder is a rare congenital anomaly which if untreated causes bladder carcinoma and intestinal tumours noted if urinary diversion is performed. It is seen that 50% of all persons afflicted with exstrophy are dead by their tenth year and 66-67% are dead by their twentieth year. It is thus a great rarity to see a case of ectopia vesicae in adulthood. Still more uncommon is to see a case of exstrophy complicated by carcinoma. Here, we report a case of papillary adenocarcinoma of ectopic urinary bladder in a 42-year-old male patient. In view of locally advanced disease, patient was given neoadjuvant chemotherapy. The case is being reported on account of its rarity to sensitise clinicians about rising incidence of carcinoma if mismanaged due to lack of protocol in oncological screening.

Keywords: Adenocarcinoma, Bladder cancer, Ectopia vesicae

CASE REPORT

A 42-year-old male with congenital genito-urinary defect presented to Department of Radiotherapy, Government Cancer Hospital with complaints of growth in supra pubic region since last 3 months [Table/Fig-1]. Patient had a history of dribbling of urine at two separate sites on the anterior abdominal wall since birth. Patient was poorly built with findings of exstrophied bladder in supra pubic area with slight urinary spillage. A solitary growth measuring 10x9 cm, hard in consistency and bleeding on touch filled whole of ectopia vesicae. Also, scrotum examination revealed a left sided hydrocele. Per rectum examination did not reveal any metastasis. There were no palpable lymph nodes.

Contrast enhanced computerized tomography of abdomen and pelvis showed widening of pubic symphysis [Table/Fig-2] and revealed heterogeneously enhancing growth in mid line suggestive of malignancy of urinary bladder [Table/Fig-3]. Working diagnosis



[Table/Fig-1]: Clinical picture showing a 10x9 cm growth in the exstrophied urinary bladder and a left sided hydrocele



[Table/Fig-2]: Coronal section of CT scan showing widening of pubic symphysis and a growth in the midline



[Table/Fig-3]: Axial section of CT abdomen showing absence of abdominal muscles and growth over anterior abdominal wall

of a carcinoma was made on clinical and imaging studies and confirmed with a biopsy from the growth showing papillary adenocarcinoma. Patient's nutrition was built and started on chemotherapy with gemcitabine (1.4g) and cisplatin (70mg). Since there was no response after four cycles, the regimen was changed to monotherapy with docetaxel. The patient still had no response after two cycles of docetaxel (120mg) and his general condition deteriorated. And hence the patient was retrieved of chemotherapy and put on palliative care. Patient has been on follow up since then. During the last follow up in February 2015, the patient doesn't show any evidence of metastasis but there is gross increase in the size of the growth with local bleeding and necrosis.

DISCUSSION

The taboos and myths of a child born with congenital anomalies haunts the humans even in 21st century, more so in the Indian sub-continent. Quiet often these babies are abandoned or not taken care of leading to medical neglect. Present patient is also such a case with a late presentation of a congenital anomaly with complication. Bladder exstrophy (EB) is a condition often encountered by Paediatricians and Paediatric Urologists, however atypical presentation in adulthood of an untreated EB often complicates the clinical scenario. Though adenocarcinoma in a patient with bladder exstrophy has an average latent period ranging from 20-26 years, it has been reported within 10 years as well [1].

Inguinal hernia is noted often in infancy due to the fact of separated

pubic bone and patent processus vaginalis and more often testes are repositioned. A Spanish study group have awakened the medical community about the need for long term follow up with Ultrasonography to unearth exact aetiology and true incidence of testicular tumours in exstrophy-epispadias complex [2]. Present case had a hydrocele of the left testes with no evidence of malignancy.

Though incidence of bladder carcinoma is less if intervened early in life, uncommonly bladder cancer is noted even in patients who had undergone cystectomy before 5 years of age [3]. Present case presented for the first time in the fifth decade without any intervention in the past.

Tumour development following urinary diversion is an accepted late complication and such individuals carry a significant risk for intestinal tumour development compared to the general population. Austen M et al., [4], has proposed mandatory regular endoscopic control beginning from 3rd postoperative year for early detection of secondary malignancies. Some authors have opined that screening is not required for atleast 15 years after augmentation or replacement cystoplasty [5]. Considering the variable occurrences of malignancy in these patients, authors feel the need for early annual screening to provide a better quality of life.

The incidence of bladder exstrophy has been estimated at between 1 in 10,000 and 1 in 50,000 live births [6]. With literature reviews, it is seen that 50% of all persons afflicted with exstrophy are dead by their tenth year, and 66-67 percent are dead by their twentieth year. It is estimated that some 10 percent reach maturity and of this number, a few develop carcinoma of the exposed bladder. It is thus a great rarity to see a case of untreated ectopia vesicae presenting in adulthood with malignancy. Adenocarcinoma of urinary bladder is infrequent constituting <2% of all bladder cancers. In contrast to this, more than 50% of cancers occurring in ectopic bladder are adenocarcinoma in nature.

There are three possible theories regarding the origin of adenocarcinoma in exstrophic bladder: Chronic irritation and infection leading to metaplasia of urothelium; Malignant degeneration of embryonic rests of gastrointestinal tissue; or from the colonic epithelium covering the mucosa of the organ.

Though systemic chemotherapy regimens used to treat transitional cell carcinoma are generally ineffective for tumours with other histology overall experience with chemotherapy in nonurothelial carcinomas is limited. Hence the treatment of adenocarcinoma of urinary bladder remains similar to transitional cell carcinoma. Studies have shown, GC (gemcitabine plus cisplatin) has better safety profile and was considered not inferior to the traditional MVAC (MVAC: methotrexate, vinblastine, doxorubicin, and cisplatin) in metastatic disease with a higher overall recurrence rate (57%) and cure rate (15 to 21%) [7]. Though surgical options were discussed, considering patient profile and choice we initiated GC regimen in the patient. Since there was no response to GC, after two cycles the patient was put on docetaxel. There was no response after two cycles of docetaxel and the patient was put on palliative care.

CONCLUSION

The awareness about malignant potential of exstrophied bladder needs to be told to patients and patient care givers. Timely correction of the deformity and vigilant monitoring perhaps improves the quality of life. The oncological principles and screening programmes need to be standardised especially in a country like ours with scarce medical resources.

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PARTICULARS OF CONTRIBUTORS:

1. Junior Resident, Department of Radiotherapy, MGMMC, Indore, India.
2. Senior Resident, Department of Surgery, ESIC Model Hospital, Indore, India.
3. Junior Resident, Department of Radiotherapy, MGMMC, Indore, India.
4. Assistant Professor, Department of Radiotherapy, MGMMC, Indore, India.
5. Professor and HOD, Department of Radiotherapy, MGMMC, Indore, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. S. Shwetha,
301, 9th Main, Canara Bank Colony, Nagarabhavi Road, Bangalore-560072, India.
Email: dr.ninu71.sn@gmail.com

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