Cloacal anomaly with bladder tumor

Amlesh Seth, Ishwar Ram

Department of Urology, All India Institute of Medical Sciences, New Delhi, 1Department of Urology, Dr. Ram Manohar Lohia Institute of Medical Sciences, Lucknow, India

ABSTRACT

A rare case of squamous cell carcinoma of bladder occurring in a 36-year-old female with persistent cloacal anomaly who presented with frequency, urgency, dysuria, and recurrent urinary tract infection is reported. Contrast Enhanced Computed Tomography with three dimensional reconstruction showed presence of bladder tumor and persistent cloaca. She underwent pelvic exenteration and wet colostomy. Histopathologic findings revealed locally advanced moderately differentiated squamous cell carcinoma.

Key words: Bladder tumor, cloacal anomaly, urinary tract infection

INTRODUCTION

Cloaca is the Latin word for sewer. Persistent cloaca is defined as a defect in which rectum; vagina and urinary tract meet and fuse into a single common channel. This is a rare anomaly seen exclusively in girls with an incidence of 1 in 50,000 live births and accounts for 10% of all anorectal malformations in females. It is a complex malformation with a wide spectrum of severity and remains a difficult reconstructive challenge. Achieving bowel and urinary control, as well as normal sexual function are the main management issues, whereas malignancy is rarely of concern in this group of patients. Although cloacogenic tumors of anorectum, bladder, and vulva are reported, which are mainly adenocarcinomas; development of squamous cell carcinoma of bladder in patients with cloacal malformation is unknown.

CASE REPORT

A 36-year-old married female having two children, both delivered by caesarian section, presented to us with complaints of urinary frequency, urgency, and urge incontinence associated with low-grade fever. She had history of recurrent urinary tract infections (UTI) since childhood and had received multiple courses of antibiotics. She had been diagnosed as having a persistent cloaca and had undergone multiple cutback procedures at birth. On pelvic examination, a single short common channel opening in the perineum was found. All preoperative blood investigations including renal function were normal.

Ultrasound abdomen showed a mass lesion in urinary bladder [Figure 1]. Computed tomography (CT) abdomen and pelvis with three dimensional reconstruction confirmed the presence of persistent cloaca, a large mass in the bladder with infiltration into the pubic bones [Figure 2] with uterus didelphys and rectum crossing between the two uteri and opening into the bladder base [Figure 3]. Cystoscopy revealed a large solid, sessile growth arising from dome and anterior wall of urinary bladder, rectum opening just distal to bladder neck and two cervical openings posteriorly. Cold cup biopsy from growth revealed a moderately differentiated squamous cell carcinoma (SCC).

With a diagnosis of squamous tumor with persistent cloaca, radical cystectomy and urinary diversion were planned. Intraoperatively, we found a 15 cm pouch colon along with a large tumor involving anterior wall of bladder, infiltrating the pubic bones. Two uteri were lying on either side of the bladder [Figure 4]. Pelvic exenteration, standard bilateral pelvic lymph node dissection, pubectomy, and wet colostomy were performed. Operative time was 6 hours with 1200 cc blood loss and the patient received 2 units of packed cell
transfusion. Postoperative course was uneventful except for a small incision disruption which was managed with secondary suturing. Histopathologic findings confirmed the presence of squamous cell carcinoma. Resected margins were free of tumor and 50% of pelvic nodes showed tumor metastases. She was started on adjuvant methotrexate and platin-based combination chemotherapy but finally lost to follow up.

**DISCUSSION**

Development of malignancy in cloacal malformations is rare. Cloacogenic tumors i.e., tumors arising from remnants of cloacal membrane have been reported in the anorectum, vulva, bladder, and vagina. Most of these are adenocarcinomas and less commonly of transitional cell origin. Squamous cell carcinoma arising from cloaca has so far has been reported in reptiles only. To best of our knowledge, this is probably the first case of squamous cell carcinoma of urinary bladder occurring in a patient with persistent cloacal anomaly.

Cloacal anomaly as an isolated risk factor for SCC is unknown. Possibly, tumor induction in this case may be attributed to the recurrent UTI. The reason for recurrent UTI in our patient may be fecal contamination of urinary tract due to persistent cloaca. Studies from John Hopkins Institute have shown that history of UTI is an important risk factor (risk ratio = 1.6, confidence interval (CI) = 1.4-1.8) for development of SCC. Various mechanisms have been proposed. These are: i) Production of excess nitrites by bacteria, ii) conversion of nitrites into nitrosamines, and iii) increased absorption of carcinogens through bladder mucosa because of inflammation.

Management of cloacal anomaly is complex, requiring an expert multidisciplinary team. Development of malignancy in patients with cloacal malformations is exceedingly rare but a possibility should be kept in mind, particularly while treating recurrent and unexplained UTIs in a patient with persistent cloaca. Taking all treatment perspectives together, the most serious problem is the lack of any histological or clinical data allowing a reliable prognosis of future bladder growth and long-term storage and voiding function after birth.

**REFERENCES**

1. Hendren WH. Cloaca, The most severe degree of imperforate anus. Ann Surg 1998;228:331.
2. Pena A. The surgical management of persistent cloaca: Results in
54 patients treated with a posterior sagittal approach. J Pediatr Surg 1989;24:590.
3. Zaidi SN, Conner MG. Primary vulvar adenocarcinoma of cloacogenic origin. South Med J 2001;94:744-6.
4. Sink JD, Kramer SA, Copeland DD, Seigler HF. Cloacogenic carcinoma. Ann Surg 1978;188:53-9.
5. Roe WD, Alley MR, Cooper SM, Hazley L. Squamous cell carcinoma in a tuatara (Sphenodonpunctatus). N Z Vet J 2002;50:207-10.
6. Kantor AF, Hartge P, Hoover RN, Narayana AS, Sullivan JW, Fraumeni JF Jr. Urinary tract infection and risk of bladder cancer. Am J Epidemiol 1984;119:510-5.
7. Anne-Karoline E, Heiko R, Michael L, Wolfgang HR. The extrophy-epispadias complex. Orphanet J Rare Dis 2009;4:23.

How to cite this article: Seth A, Ram I. Cloacal anomaly with bladder tumor. Indian J Urol 2013;29:142-4.
Source of Support: Nil, Conflict of Interest: None declared.