J Ped Surg Case Reports 3 (2015) 385–388

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Undifferentiated sarcoma developing 14 years after colocystoplasty: Our experience and literature review

Yutaka Hayashi [a,*], Satoko Shiyanagi [a], Manabu Okawada [b], Hiroyuki Koga [b], Junya Fujimura [c], Itsuro Nagae [a], Akihiko Tsuchida [a], Atsuyuki Yamataka [b]

[a] Department of Gastrointestinal and Pediatric Surgery, Tokyo Medical University, Tokyo, Japan
[b] Department of Pediatric General and Urogenital Surgery, Juntendo University School of Medicine, Tokyo, Japan
[c] Department of Pediatrics and Adolescent Medicine, Juntendo University School of Medicine, Tokyo, Japan

ARTICLE INFO

Article history:
Received 24 July 2015
Accepted 3 August 2015

Key words:
Bladder augmentation
Malignancy
Sarcoma
Neurogenic bladder

ABSTRACT

A boy with myelomeningocele who had sigmoidocolocystoplasty and ureteric reimplantation when 2-years old and normal annual cystoscopies, developed hematuria and abdominal pain with liver dysfunction 14 years postoperatively. Computed tomography showed a tumor on the left side of the augmented bladder, a large lymph node, and large multiple probable metastases in the liver. Cystoscopy 3 months earlier had been normal, but when repeated showed a tumor originating from the augmented sigmoid colon. Biopsy showed undifferentiated sarcoma. Despite chemotherapy, he died 3 months later. The diagnosis at autopsy was undifferentiated sarcoma originating from the sigmoid colon. We report the first case of undifferentiated sarcoma developing 14 years after sigmoidocolocystoplasty for meningomyelocele, and also review the 55 cases of post-bladder augmentation malignancy in the literature.

Since Mikuliz's [1] report published in 1899, the indications for enterocystoplasty (ECP) using colon, ileum, or stomach, have expanded dramatically, and ECP has become an accepted reconstructive option for intractable incontinence and poor bladder compliance in many neurogenic and non-neurogenic disorders [2].

However, recently, there have been an increasing number of reports of benign and malignant tumors developing in the neo-bladder in post-ECP patients on long-term follow-up, especially around the line of anastomosis between the colon cap and the native bladder remnant [2]. On histopathology, the malignant tumors identified were adenocarcinoma, transitional cell carcinoma, and malignant sarcoma.

We have performed 120 sigmoidocolocystoplasties over the past 30 years in our departments. Of these, one case developed sarcoma at the patch site of the sigmoidocolocystoplasty. We report this case and review the literature for post bladder augmentation malignancies.

1. Case report

A 16-year-old boy was hospitalized after becoming febrile with hematuria, dysuria, general fatigue, and right upper quadrant pain. He was born with myelomeningocele and neurogenic bladder with bilateral ureterovesical junction stenosis, bilateral hydroureter and hydronephrosis, bilateral renal dysfunction, and low bladder compliance. He underwent sigmoidocolocystoplasty and bilateral ureteric reimplantation when he was 2-years old.

On admission, white cell count (7800/μL), C-reactive protein (8.3 mg/dL), and transaminase (AST: 83 IU/L, ALT: 57 IU/L) were elevated among the laboratory data. However, tumor markers such as α-fetoprotein, carcinoembryonic antigen, and carbohydrate antigen 19-9 were not elevated. Computed tomography showed a tumor on the left side of the augmented bladder, swelling of the lymph nodes in the pelvic cavity, and large multiple probable metastases in the liver (Fig. 1a and b). Cystoscopy 3 months earlier had been normal, but when repeated showed a tumor originating from the augmented sigmoid colon (Fig. 2). Histopathology revealed that the tumor was composed of small

* Corresponding author. Department of Gastrointestinal and Pediatric Surgery, Tokyo Medical University, 6-7-1 Nishi-shinjuku, Shinjuku-ku, Tokyo 160-0023, Japan. Tel.: +81 3 3342 6111; fax: +81 3 3340 4575.
E-mail address: rimpoo@tokyo-med.ac.jp (Y. Hayashi).

2213-5766/
http://dx.doi.org/10.1016/j.epsc.2015.08.001

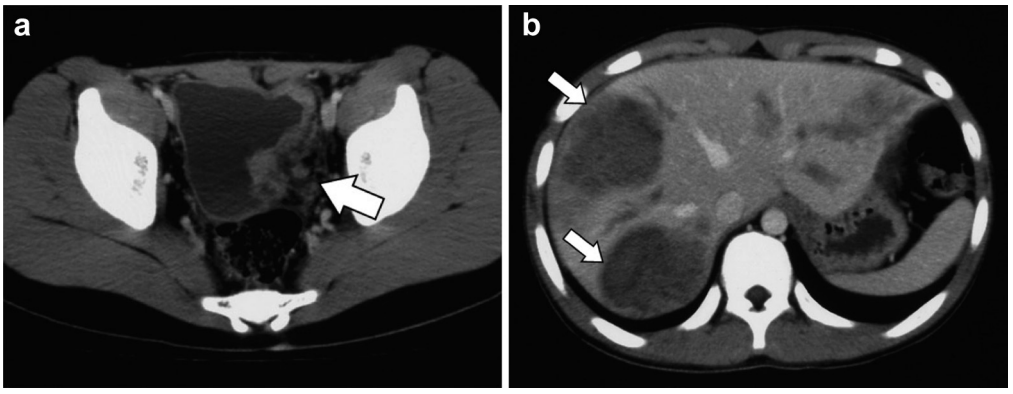

Fig. 1. Computed tomography images of the patient. a: Image of the patient's bladder, displaying thickening of the bladder wall (arrow). b: Image of the patient's liver, displaying large multiple masses (arrows).

round cells (Fig. 3a and b) that stained positively for vimentin, but negatively for cluster of differentiation 99 (also known as MIC-2), desmin, myoglobin, smooth muscle actin, S-100, chromogranin, cluster of differentiation 56, neuron specific enolase, synaptophysin, cluster of differentiation 34, and podoplanin (a 38-kDa O-linked transmembrane sialoglycoprotein recognized by the D2-40 antibody). From these findings, a diagnosis of undifferentiated sarcoma was made.

The tumor grew rapidly, and his general condition worsened. The multiple liver metastases ruptured repeatedly and his condition was thought to be beyond surgical cure. Chemotherapy was therefore commenced.

We initially selected vincristine, actinomycin D, and cyclophosphamide (VAC) therapy, because the tumor was histopathologically similar to rhabdomyosarcoma. However, when we evaluated the efficacy of the chemotherapy by computed tomography, 9 days after the start of chemotherapy, we found that the size of the liver metastasis had increased. Therefore, we concluded that VAC therapy was not effective and changed to ifosfamide and etoposide (IE) therapy, which is used for the treatment of Ewing sarcoma. However, intratumoral hemorrhage occurred during the course of chemotherapy, and his general condition worsened. From that time, he developed jaundice, breathing failure due to the abdominal distension, and systemic edema. IE therapy was ineffective and we changed his treatment to cisplatin and Adriamycin. However, this was also ineffective and he died 3 months later from multiple organ failure (Fig. 4). The diagnosis at autopsy was undifferentiated sarcoma originating from the sigmoid colon (Fig. 5).

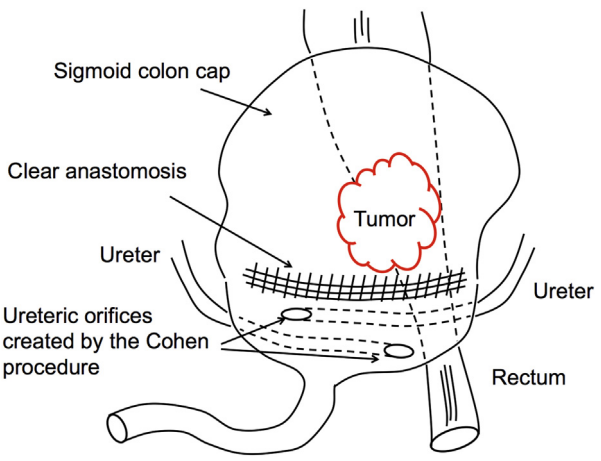

Fig. 2. Diagram of cystoscopic findings prior to commencing treatment for the malignancy. A tumor was identified in the left side of the augmented bladder. The tumor existed away from the anastomosis, between the sigmoid colon cap and the native bladder. There were no malignant changes in the native bladder, including in the ureteric orifices.

2. Literature review

To the best of our knowledge, there are 55 cases of malignancy occurring after bladder augmentation reported in the literature (Table 1). The sex distribution was 24 male patients (44%), 16 female patients (29%), and 15 undescribed patients (27%). Age at ECP ranged from 5 to 59 years (mean: 31 years), with 7 cases receiving ECP when less than 15 years old (13%); 7 between 16 and 20 years old (13%), 10 between 21 and 30 years old (18%), 3 between 31 and 40 years old (5%), 7 between 41 and 50 years old (13%), 8 more than 50 years old (15%), and 13 not described (24%). Accordingly, most patients developed malignancy as adults. Duration of follow-up ranged from 0.25 to 38 years (mean: 19.3), with follow-up for 0–5 years in 5 (10%), 6–10 years in 4 (7%), 11–15 years in 12 (22%), 16–20 years in 10 (18%), and 21–30 years in 13 (24%), 31 or more in 9 (16%), and not described in 2 (4%). Underlying pathology was urinary tuberculosis in 17 cases (31%), neurogenic bladder in 15 cases (27%), schistosomiasis in 2 cases (4%), post bladder surgery in 1 case (2%), and unknown in 20 cases (36%). Bladder augmentation involved the small bowel in 30 cases (55%), colon in 13 cases (24%), stomach in 8 cases (15%), and ileocecum in 4 cases (7%). Histopathology confirmed adenocarcinoma in 36 cases (65%), transitional cell carcinoma in 11 cases (20%), signet ring cell carcinoma in 4 cases (7%), squamous cell carcinoma in 2 cases (4%), oat cell carcinoma in 1 case (2%), and sarcoma after ileocystoplasty in 1 case (2%). Malignancies arose from the anastomosis in 27 cases (49%), intestinal cap in 10 cases (18%), native bladder in 9 cases (16%), and unknown in 9 cases (16%). Treatment was cystectomy in 38 cases (69%), ileal conduit in 15 cases (27%), ureterostomy in 8 cases (15%), sigmoid conduit in 4 cases (7%), ileocystoplasty in 3 cases (5%), pelvic exenteration in 2 cases (4%), transurethral resection in 2 cases (4%), and cecocystoplasty in 1 case (2%).

3. Discussion

There have recently been reports of an increased incidence of malignancy after ECP [2–5], most developing in patients with tuberculous cystitis. It is well known that malignant epithelial