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Anorectal Adenocarcinoma after Pull-through Procedure for Imperforate Anus

Jing-Jim Ou[1], Shu-Wen Jao[1], Jung-Cheng Kang[1,2], Huang-Jen Lai[1], and Cheng-Wen Hsiao[1*]

[1]Division of Colon and Rectal Surgery, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan, Republic of China
[2]Division of Colon and Rectal Surgery, Department of Surgery, Buddhist Tzu Chi General Hospital, Taipei Branch, Taiwan, Republic of China

The abdominoperineal pull-through procedure was the standard management of imperforate anus before the introduction of posterior sagittal anorectoplasty. Adenocarcinoma arising from the pulled-through anorectum is extremely rare and has been documented in only three reports. We describe a 21-year-old man with history of imperforate anus that was treated by pull-through procedure at the age of one year, who later developed adenocarcinoma over the formed anorectum. Despite radical surgery and adjuvant chemoradiation therapy, the patient died of his disease six months after surgery. Review of his family pedigree showed no familial history of colorectal cancer or associated genetic disease. Chronic inflammation of the mucocutaneous junction seems to be the predisposing factor for malignant generation. We suggest that patients with an imperforate anus undergoing the pull-through operation should receive annual follow-up with careful digital rectal examination. If a suspicious lesion appears, incisional biopsy should be made to detect the inflammatory process and early malignant changes in the pulled-through anorectum to allow for appropriate treatment in the early stage.

Key words: adenocarcinoma; imperforate anus; pull-through procedure

INTRODUCTION

Imperforate anus occurs in one of every 4000-5000 newborns[1]. Before posterior sagittal anorectoplasty was introduced by deVries and Peña[2] in 1982, the abdominoperineal pull-through procedure was the standard treatment of anorectal malformation[3,4]. Complications associated with the pull-through procedure are fecal incontinence, soiling, constipation, and rectal stricture[5,6]. Malignancy of the pulled-through anorectum is rare, but lethal. To our knowledge, only three reports have documented this malignancy[6,7,8]. We describe a rare case of anorectal adenocarcinoma after the pull-through procedure to treat an imperforate anus. We review the literature and discuss the possible cause of the carcinoma of the pulled-through anorectum.

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*Corresponding author: Cheng-Wen Hsiao, Division of Colon and Rectal Surgery, Department of Surgery, Tri-Service General Hospital, No. 325, Sec. 2, Cheng-Gong Road, Neihu 114, Taipei, Taiwan, Republic of China. Tel:+886-2-8792-7223; Fax: +886-2-8792-7411; E-mail: hamnet.tw@yahoo.com.tw

CASE REPORT

A 21-year-old man was diagnosed with imperforate anus with rectoperineal fistula at birth in 1981. He underwent a transverse loop colostomy for diversion of feces immediately after birth. The abdominoperineal pull-through procedure was performed in 1982 when he was one year old, and a closure of transverse loop colostomy was undertaken six months later. After the successful pull-through procedure, the patient grew up without apparent anorectal symptoms during his childhood. The follow-up of the patient was lost after the operation. Although the anal fistula was not diagnosed at local clinics until the age of 16 years, no evaluation or treatment was performed in the following five years.

In 2002, at the age of 21, the patient visited our hospital and presented with painful defecation and an anal mass. Digital rectal examination revealed an induration with tenderness measuring about 3×4 cm over the left anterior aspect of the formed anorectum. Computed tomography of the pelvis demonstrated a lobulated heterogeneous soft tissue mass 5 cm in diameter over the left anterior wall of the anorectum and a few small soft-tissue nodules over the adjacent mesorectum (Fig. 1). An incisional biopsy for anorectal mass was performed and the pathological result

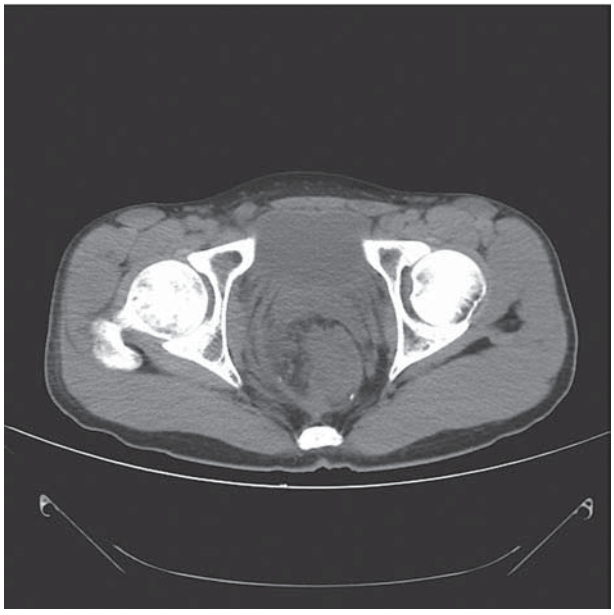

Fig. 1 Computed tomography of the pelvis demonstrates a lobulated heterogeneous soft-tissue mass 5 cm in diameter over the left anterior wall of the anorectum with a few small soft tissue nodules over the adjacent mesorectum.

proved to be adenocarcinoma. The carcinoembryonic antigen concentration was 14.46 ng/ml (reference range <3.0 ng/ml), and results of other laboratory studies were in the normal range. Review of his family pedigree showed no familial history of colorectal cancer or associated genetic disease. We performed abdominoperineal resection with an adequate cutting end and repaired the damage to the urethra caused by tumor invasion to the bulbous urethra. Pathologic examination demonstrated a fungating tumor measuring 5×4×3 cm in size, arising from the mucocutaneous junction of the formed anorectum (Fig. 2). The adenocarcinoma was identified microscopically as poorly differentiated and invading to the bulbous urethra with metastatic adenocarcinoma in 15 of 22 dissected lymph nodes. The postoperative recovery was uneventful and the patient received adjuvant radiochemotherapy. Unfortunately, distant metastasis developed and the patient expired six months after surgery.

DISCUSSION

Malignancy of a pulled-through anorectum is rare, but lethal, and has been documented in only three reports. In 1982, Polk *et al*.[7] reported a 63-year-old man with a history of anorectal malformation who underwent the pull-through

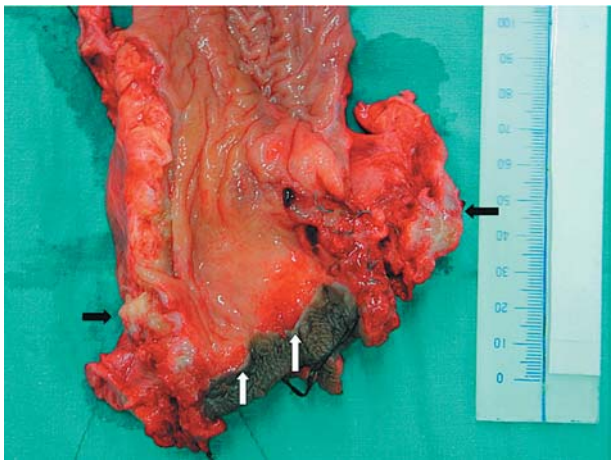

Fig. 2 Gross photograph of the specimen demonstrates a mucocutaneous anastomosis (white arrow) instead of a dentate line. The black arrow indicates the fungating tumor arising from the formed anorectum.

procedure in childhood and later developed adenocarcinoma of the rectosigmoid colon. The pathologic examination showed adenocarcinoma arising from villous adenoma 12 cm above the anastomosis. In 1988, Mukawa *et al*.[6] reported a 35-year-old man with long-standing fecal incontinence as a complication of a pull-through procedure for an imperforate anus at birth. They speculated that the carcinoma developed because of repeated trauma to the mucocutaneous junction by consecutive fecal streams. In 2000, Posey *et al*.[8] reported a patient with signet-ring cell carcinoma arising from the pulled-through sigmoid colon. This patient had concomitant genitourinary and anorectal malformation and recurrent urinary tract infection caused by the mix of urinary and fecal streams, which may also cause carcinogenesis. Table 1 summarizes the data on these three patients and our patient. The previous patients had either rectourethral fistula or concomitant genitourinary anomaly, which may have generated carcinogens by allowing a mix of urinary and fecal streams[9]. Our patient did not experience concomitant genitourinary anomaly or rectourethral fistula, and it appears that carcinogenesis was caused by factors different than those causing carcinoma in the previous patients.

Adenocarcinoma of the rectum occurs rarely in patients younger than 30 years old, and accounts for only 1% of the incidence of colorectal cancer[10,11]. Most colorectal cancers in young people are related to familial history of colorectal cancers or associated genetic diseases. In contrast, our patient had neither precipitating factor. He had a preexisting anal fistula, which had bothered the patient for several years, although he received no treatment for it. The occur-

Table 1 Summary of Reported Cases of Carcinoma after Pull-through Procedure for Imperforate Anus

Case	Age/Sex	Type of anorectal malformation	Concomitant GU anomaly	Location of carcinoma	Type of carcinoma	Probable cause of carcinogenesis	Reference
1	63/M	Rectourethral fistula (bulbar)	Hypospadia Chordee	Rectosigmoid colon 12 cm from anal verge	Adenocarcinoma	Arising from villous adenoma	【7】
2	35/M	Rectourethral fistula (prostatic)	None	Anorectum	Adenocarcinoma	Repeated trauma due to fecal incontinence	【6】
3	25/M	Not mentioned	Hypospadia Neurogenic bladder	Rectosigmoid colon 2 cm from anal verge	Signet-ring cell carcinoma	Mix of urinary and fecal streams	【8】
Author's	21/M	Rectoperineal fistula	None	Anorectum	Adenocarcinoma	Long-standing chronic inflammation of mucocutaneous junction	-

rence of neoplasm in a long-lasting fistula has been reported[12,13], although it is difficult to determine whether the tumor is a complication of a longstanding anal fistula or whether the fistula is merely a manifestation of the malignancy itself[12]. However, the anal fistula implied an ongoing inflammatory process over the mucocutaneous junction of the pulled-through anorectum. Getz *et al*.[13] demonstrated the relationship between chronic inflammation and carcinogenesis, and thought that the origin of carcinoma in an area of chronic inflammation arises when the area in question becomes an "immunologically privileged site" with disrupted lymphatics and impaired immunologic surveillance against developing metaplastic or frankly neoplastic cells. This suggests that malignancy resulted from chronic inflammation of the mucocutaneous junction in our patient. We suggest that patients with imperforate anus undergoing pull-through operation, or even posterior sagittal anorectoplasty, should receive an annual follow-up with careful digital rectal examination and questioning about any symptoms of itching, pain, or induration of the mucocutaneous junction. For suspicious malignant lesions, an incisional biopsy should be made to detect the inflammatory process or an early malignant change of the anorectum to allow for treatment during the early stage.

In our patient, urethral injury occurred during abdominoperineal resection for an anorectal mass. Urethral injury may be caused by tumor invasion, although anterior mislocation of the rectum in patients with an anorectal malformation may also contribute[3]. Preoperative genitourinary evaluation is advised to identify the potential urethral involvement and to prevent urinary tract injury during surgery. A thorough understanding of anatomy and the corrective operation is mandatory when anorectal surgery is planned in patients with a history of imperforate anus. Preoperative chemoradiation for locally advanced rectal cancer is a promising regimen that provides excellent local control and favorable overall survival[14]. Patients with carcinoma arising from a pulled-through anorectum to treat an imperforate anus may benefit from preoperative chemoradiation, which may provide the advantage of a downstaging effect, and this should improve overall survival.

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