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Case Report

Adult rectal duplication and canceration: a case report and literature review

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Abstract: Intestinal duplications especially rectal duplications are uncommon congenital developmental anomalies. Rectal duplication complicated with adenocarcinoma is even scarcer. Here we present a rare case of a 38 year-old Chinese male who was diagnosed with rectal duplication with canceration. After reviewing lots of related literatures from domestic and abroad, we'd like to discuss the pathogenesis, diagnostic criteria, the clinical characteristics, the treatment modalities, the prognosis of this disease and provide treatments for similar cases. Although 12 cases of rectal duplication complicated with adenocarcinoma has been described, we failed to discover any reports about the precise curative effect of postoperative radio-chemotherapy on the disease. The patient received abdominal perineal resection and six times chemotherapy without evidence of tumor recurrence in 18 months' follow-up.

Keywords: Rectal duplication, adult, cancerization

Introduction

Rectal duplication (RD) is a very rare disease in malformation of digestive tracts. Clinically, duplication of small intestines is most commonly seen, ileocecal junction takes second place, and RD is relatively rare. Studies showed that the ratio of RD is below 5% in the malformation of digestive tracts [1], and the cases could only be found in few of the adult patients. It was reported that the age group of incidence mostly focused on the period of 30-40 years in the patients, with the male female ratio of 1:3 [2]. RD with canceration was first described by Ballantyne EN in 1932 [3]. Eleven additional cases of RD with adenocarcinoma have been reported [4-6]. Here we present a rare case of a 38 year-old Chinese male who was diagnosed with rectal duplication and canceration.

Case report

A 38-year old male visited our hospital (Gansu province people hospital, China) with "left buttock lump plus progressive enlargement for 30 years" on May, 2014. His mother had no particular problems during pregnancy and he did not have a family history of tumors. The patient was diagnosed with complex anal fistula in the local hospital when he was 8. He had undergone four operations on the disease between the ages of 8 to 38 years old, but without postoperative pathologic examination. However, the disease progressed after surgery at the site of incision, the sacrococcygeal region with frequent pyorrhea, pain and bleeding gradually. In recent 10 years the patient was aware of an obvious enlargement of the tumor.

The abdomen and both inguinal lymph nodes examination were unremarkable. In the 11 o'clock to 1 o'clock direction of the knee-chest posture colorectal examination, we found a hard lump measuring about 4 × 5 cm protruding from the body surface with uneven surface and several old vertical operation scar situated about 5 cm from the anal verge. We identified thin purulent fluid which was being discharged from the fistulas with gradually increasing local tenderness. In the digital rectal examination (DRE), the mucous membrane of rectum was

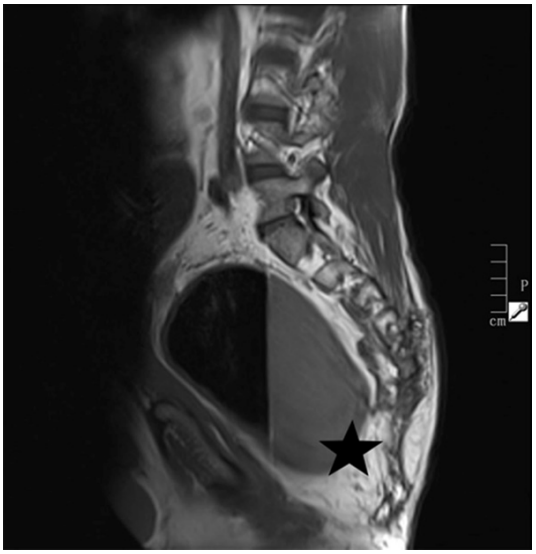

Figure 1. Pelvic MRI showed at sacrococcygeal region and in pelvic cavity, there was a giant tumor presenting miscibility, about 16 × 18.35 cm in size, considered preliminarily as teratoma.

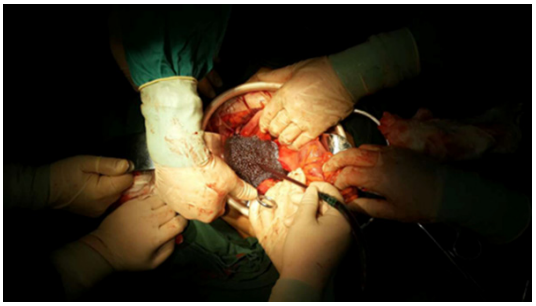

Figure 2. During surgery, the upper bound of tumor was cystoid. As it is broken, there was about 1000 ml of pitchy stinking fluids in the tumor.

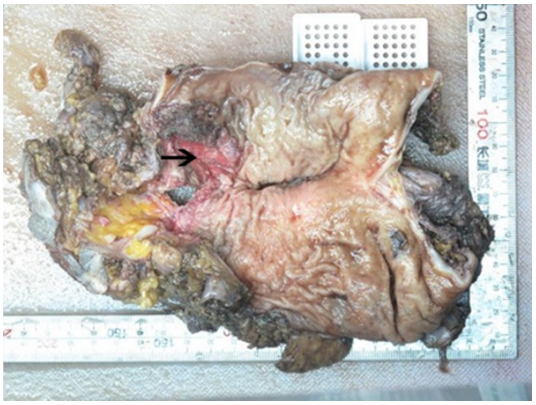

Figure 3. There was digestive tract mucosa on the inner wall of excised sample. The tumor located at the distal end, ulcerative-type, and perforating the skin by the anal side.

smooth. We found a giant lump located the left-rear side of the rectum 4 cm from the anal verge. The levels of tumor markers, such as carcinoembryonic antigen (CEA), carbohydrate antigen 19-9 (CA 199) and alpha fetoprotein (AFP), were all within normal limits. The enhancement MRI scan of pelvic region indicated that there was a giant tumor in the sacrococcygeal region and pelvic cavity of about 16 cm × 18.35 cm, with a primary confirmation of a teratoma (**Figure 1**). Ultrasonography of sacrococcygeal region showed the tumor in the sacrococcygeal region by subcutaneous mixed echo diagnosis. The orthotopic X-ray film of the pelvis demonstrated that in the presacral region, there was an air fluid level with a diameter of about 7.67 cm. So it was initially diagnosed as a presacral lesion. The enhancement CT of the pelvic cavity demonstrated that there was a gigantic space-occupying lesion at the left-of-center pelvis, which can usually be diagnosed as the concurrent infection of teratoma (wide extent of disease, more subcutaneous fatty components, and cystic lesion ranking first in pelvis). Intra-operative exploration demonstrated that the tumor was located in the front of sacrum and at the left side of the rear of the rectum. The upper boundary of tumor reached the surface of the sacropromontory, covered with peritoneum, and its lower boundary could not be determined. It was thought that it was being integrated with the lump at the left-side buttock. The lump had nearly taken up the whole pelvic cavity, and the rectum could not function well. The boundary between the tumor and the mesorectum was unclear. During the separation process, the capsule wall of tumor was broken, approximate 1000 ml fuscous smelly fluids discharged (**Figure 2**). Since it was clear in the next separation that the tumor had been integrated with the buttock lump, a joint resection of the tumors at abdomen and perineum was conducted.

On visual examination of the resected specimens, there was a capsular space on the lateral side of the anal specimen, about 6 × 8 cm in size. The inner wall of capsular space was smooth, the wall thickness was about 1 cm, and the tissue elements such as hairs, teeth and cartilages were not seen. The duplicated intestinal canal at the anal side specimen was about 8 cm in length, and a solitary hard lump was growing on the surface of mucous mem-

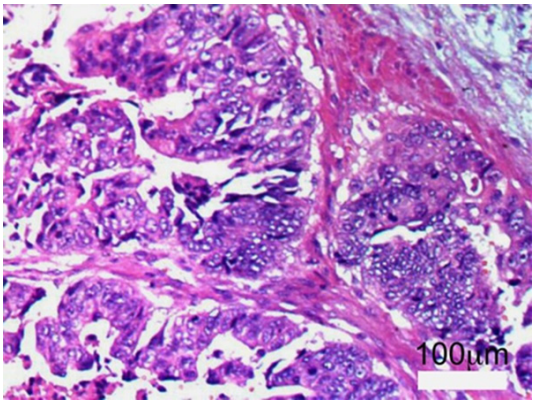

Figure 4. By microscopy, the cancer cells presented a nidulant array, part of cancer cells secreted mucosa, sabotage the cell matrix, smooth muscle can be seen around the cancer cells (HE, 20 ×).

brane, approximately 4 × 4 cm in size, presenting the ulcerative growth. The full-thickness was infiltrated on the basilar part, involving the adventitia, which had a close adhesion with the normal mesorectum, breaking through the anal side until the skin of perianal area (**Figure 3**). In routine H & E staining, it was found by 20 × 10 microscopy that the cancer cells present a nest bulk array, and the form of the glandular organ was aberrant. Part of the cancer cells secreted mucus, forming the mucin pool, with sabotage of the normal cell matrix. Smooth muscle was also seen around the cancer cells (**Figure 4**). The results of pathological diagnosis showed that it was RD, and a tubiform papillary adenocarcinoma was seen on the lower end of the duplicated rectum, and part of the tissues was mucinous adenocarcinoma. The cancer broke through the perianal skin. A remedial radical operation of rectal cancer (a proctectomy with total mesorectal excision, Miles) was conducted one week after the surgery. The postoperative rectal lumen was normal, and there was no evidence of lymph node metastasis. The TNM classification was defined as T4N0M0; stage II B. Following the discussion meeting of a multidisciplinary team, postoperative adjuvant chemotherapy (tegafur, leucovorin and oxaliplatine) was conducted based on the principle of management of rectal cancer and free of evidence of recurrence in the follow-up after one and a half years.

Discussion

RD is an infrequent congenital developmental malformation in the digestive tracts, also known as enterogenous cyst or enteral cyst, which is more frequently seen in childhood, and extremely rare in adults. Also, RD complicated with adenocarcinoma is even more uncommon. To date, only 12 cases have been described (Ten women, two men, aged 32-65) [4-6]. The majority of scholars comply with Ladd and Gross's criteria to diagnose RD [7], including the following: (1) the cyst should be close to or in continuity with the rectum; (2) the cyst should have a smooth muscle cover; (3) the cyst should be covered by mucosa found in the gastrointestinal tract. Therefore, the case accords with Ladd and Gross criteria. Although several theories about the etiology of RD have been proposed, none of these theories can explain all the known RD alone. The majority of scholars accepted the theory that duplication occurs when a diverticulum forms in the 8-9-week-old embryo [8]. It was reported that RD's overall ratio of malignant transformation was about 12% [4], however, the probability of the canceration was more significantly frequent in the adults (30 years old and older) [6]. In this case, the patient has undergone a repeating infection for more than thirty years, and 1000 ml of pitchy stinking fluids in the cyst cavity was found in the operation. It suggests that along with the age grow, the repeating and chronic inflammation stimulation may be an important risk factor for malignant transformation. Although some of cases of RD with canceration have been reported now, the mechanism of malignant transformation is still unclear. Further research should focus on the mechanism of malignant transformation in RD.

The clinical manifestation of the disease is diverse, lacking specific symptoms and signs. Preoperative diagnosis was less clear, and discovery of the complicated cancerization was also difficult by routine means. At present, the inspection method of specificity is not yet unavailable, and the misdiagnosis rate is high. The preoperative final diagnosis rate is reported to be below 20% [9]. The differential diagnosis of RD includes anterior meningocele, sacrococcygeal teratoma, dermoid cyst, epidermal cyst, tailgut cyst, anus gland cyst, cystic neuroblastoma, chordoma, rectal leiomyosarcoma, cloacogenic carcinoma and complex anal fistula. This case had previously been misdiagnosed as complicated anal fistula for a long time. This disease usually had no incipient symptom, and the patient went to see the doctors only

because of a complication. So, the diagnosis could just rely on the clinical manifestations. The DRE may be involved in the rectal lumen extruded by the smooth muscle and hard lump, resulting in the enteric cavity deformation and narrowing. Therefore, clinically, routine DRE should be carried in the patient with prolonged unhealed anal fistula and difficult defecation. Although this method had produced a low positive diagnosis rate, it still had important role of leading to a diagnosis. The preoperative aspiration biopsy had a relatively higher positive diagnosis rate, nevertheless, it might have the risk of tumor cell dissemination and metastasis. So, concerning the extraintestinal and presacral tumorous diseases, biopsy is not recommended presently [10]. Llauger [11] thought that barium enema, B-ultrasonic, CT, and MRI examinations have their value, however, the pathological diagnosis after surgery should be the most accurate diagnosis.

Presently, surgical ablation is the most effective way of treating RD cancerization. The choice of the operation method depends on the site and scope of the deformity, and whether it is communicated with the enteric cavity. The surgery can be performed in a combination mode, i.e. via abdomen, per anus, and via coccygeal incision or transabdominal perineum. The operation can also be completed by laparoscopy-assisted surgery and transanal endoscopic microsurgery (TEM) [1, 12]. It was reported by scholars [6] that total resection of the rectum (total mesorectal excision) could be performed where there is cancer. In principle, a standard radical proctectomy for rectal cancer should be carried in the case, with joint resection of the duplicated and cancerated rectum. In this case, since the duplicated rectal cancer had totally infiltrated the full-thickness, there were ulcerations in the deep fascia of the rectum during the excision, and the distant end of the cancer perforated the perianal skin, the transabdominal perineum joint resection of the duplicated rectum and cancer should be conducted. During this surgery, however, the doctors failed to make final diagnosis of RD cancerization because of insufficient experience, and because the duplicated rectum and cancer were excised first. After obtaining the clear pathological results which indicated the adenocarcinoma, where part of the tissues was mucinous adenocarcinoma, the remedial transabdominal perineum radical resection of rectal cancer was performed. Currently there are no statistical reports about the prognosis of a large number of cases of RD patients with canceration, but the previous reports [3, 13-15] suggest that a high local recurrence rate despite radical surgery. We didn't discover any reports about the precise curative effect of postoperative radio-chemotherapy on the disease [4-6, 8]. In our case, postoperative six times systemic intravenous chemotherapy was conducted, and without evidence of tumor recurrence in 18 months' follow-up.

Conclusion

RD is a very rare disease in malformation of digestive tracts. The mechanism of malignant transformation is still unclear, further research should focus on the mechanism of malignant transformation in RD. The experience of this case and other reports indicate that the disease was very easily to be misdiagnosed, once the canceration occurred in RD, the rate of relapse and metastasis is high. Reasonable diagnosis should be made, and radical surgical treatment should be carried out as early as possible. Adjuvant treatment (radio/chemotherapy) should also be considered in order to achieve a favorable recovery. In addition, regular follow-up after operation should be performed.

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Disclosure of conflict of interest

None.

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