Case report

A case of sigmoidectomy for sigmoid colon cancer with severe pulmonary arterial hypertension associated with mixed tissue connected disease: A case report

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ABSTRACT

Introduction: Patients with mixed connective tissue disease (MCTD) have higher rates of pulmonary arterial hypertension (PAH) than the general population. PAH is a risk for perioperative respiratory and heart failure, and marked edema of colonic stoma after sigmoidectomy. We report a case of sigmoidectomy for sigmoid colon cancer in a patient with PAH associated with MCTD for whom perioperative treatment was planned to control pulmonary arterial pressure (PAP), and a surgical strategy to avoid complications attributable to PAH and MCTD was employed.

Case presentation: A 52-year-old woman with sigmoid cancer and severe PAH associated with MCTD underwent surgery. We controlled PAH by using intravenous epoprostenol. We selected open surgery without laparoscopy and Hartmann’s operation. After surgery, severe perioperative complications were not detected, and the patient discharged from hospital 17 days after the operation.

Discussion: During surgery under general anesthesia, the mortality rate of PAH is high because of heart and respiratory failure. We planned to switch the PAH treatment from an oral agent to intravenous epoprostenol only in the preoperative period, and selected open surgery. We ligated the inferior mesenteric artery (IMA) and inferior mesenteric vein (IMV) below the branch of LCA to avoid marked edema of stoma. Consequently, we could avoid severe intraoperative and postoperative complications.

Conclusions: Controlling PAP using epoprostenol, open surgery, stoma and the ligation level for the IMA and IMV were important to avoid perioperative complications of sigmoid colon cancer complicated by severe PAH.

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1. Introduction

Patients with mixed connective tissue disease (MCTD) have higher rates of pulmonary arterial hypertension (PAH) than the general population. Patients with MCTD accompanied by PAH have an especially poor prognosis [1,2]. The rate of PAH is as high as 7.0% in patients with MCTD [3].

Unlike other autoimmune diseases, MCTD has no relevance to malignancies [2]. PAH is associated with portal hypertension [4], and there are case reports of marked edema of the colonic stoma after colectomy in patients with PAH [5]. Moreover, patients with PAH carry risks of heart and respiratory failure [6]. We report a case of sigmoidectomy in a patient with PAH accompanied by MCTD for whom perioperative treatment was planned to control PAP, and a surgical strategy was devised to avoid complications attributable to PAH and MCTD. This case report has been reported according to the SCARE checklist in 2020 [7].

2. Case presentation

A 52-year-old woman with PAH associated with MCTD was admitted to our institution because of anemia. Colonoscopy was performed to investigate the cause of anemia, and invasive sigmoid colon cancer was revealed. After a type 1 tumor was identified, pathological examination revealed adenocarcinoma (tub1). There was no finding of lymph node and distant metastasis on computed tomography (CT), magnetic resonance imaging, and positron emission tomography–CT.

In this patient, PAH was classified as WHO-PH/NYHA III/III, and oxygen therapy was essential. Cardiac testing revealed that the mean pulmonary arterial pressure (mPAP) was as high as 40 mmHg. Because the oral intake of medicines would be restricted for several days after abdominal
Surgery, we planned to switch from oral therapy to intravenous epoprostenol. Twenty days before surgery, treatment with a continuous drip infusion of 0.5 ng/kg/min epoprostenol, in addition to the use of tadalafil, macitentan, and selexipag, was initiated. The dose of epoprostenol was increased daily by 0.5 ng/kg/min at day 4, 1.0 ng/kg/min at day 19 and that of the oral drug was concomitantly decreased. Five days before surgery, we used epoprostenol alone, and increased to 17 ng/kg/min. The mPAP had decreased to 26 mmHg at day 17 (Fig. 1).

The patient underwent Hartmann’s operation at day 19. The level of ligation of the inferior mesenteric artery (IMA) was below the branch of the left colic artery (LCA), and the inferior mesenteric vein (IMV) was dissected at the same level. The first branch of the sigmoid artery and sigmoid vein was dissected as presented in Fig. 2a and c.

Moderate edema of the colonic stoma was observed after the operation, although this complication gradually improved as presented in Fig. 3. However, mPAP increased as high as 40 mmHg after surgery, we decided to manage the patient in the ICU and adjusted the dose of epoprostenol. On the second day after surgery, mPAP decreased to 20–30 mmHg, and the patient was moved to the general ward. Subsequently, mPAP continued to improve, and we gradually decreased the dose of epoprostenol on the eighth day after surgery. The treatment was ultimately switched from epoprostenol to oral therapy. The patient was discharged from hospital 17 days after surgery without marked complications. She has undergone no remarkable complications and no recurrence 12 months after surgery.

3. Discussion

MCTD was first described by Sharp et al. in 1972. This disease causes Raynaud’s phenomenon, arthritis, PAH, and other complications [2]. Several other autoimmune diseases have been reported to be associated

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**Fig. 1.** Graphs of perioperative therapies for pulmonary hypertension switching oral agent and epoprostenol. The line graph shows the dose of epoprostenol, and the bar graph indicates the date of oral administration of Tadalafil, Macitentan, and Selexipag.

**Fig. 2.** a, c. The ligation level of the inferior mesenteric artery (IMA) was below the branch of the left colic artery (LCA). The inferior mesenteric vein (IMV) was dissected at the same level. The first branches of the sigmoid artery (SA) and sigmoid vein (SV) were dissected at the origins of these vessels. b. Colonoscopy revealed a type 1 cancer of the sigmoid colon.

**Fig. 3.** However, mPAP increased as high as 40 mmHg after surgery, we decided to manage the patient in the ICU and adjusted the dose of epoprostenol. On the second day after surgery, mPAP decreased to 20–30 mmHg, and the patient was moved to the general ward. Subsequently, mPAP continued to improve, and we gradually decreased the dose of epoprostenol on the eighth day after surgery. The treatment was ultimately switched from epoprostenol to oral therapy. The patient was discharged from hospital 17 days after surgery without marked complications. She has undergone no remarkable complications and no recurrence 12 months after surgery.

3. Discussion

MCTD was first described by Sharp et al. in 1972. This disease causes Raynaud’s phenomenon, arthritis, PAH, and other complications [2]. Several other autoimmune diseases have been reported to be associated
with malignancies. However, there are few reported cases of coexisting malignant tumors in patients with MCTD [2]. The overall survival rate of patients with MCTD is 91.8% at 10 years [2], and thus, it can be said that the prognosis of MCTD is good.

However, the prognosis of patients with MCTD accompanied by PAH is poor [1,2]. PAH is defined as mPAP at rest exceeding 25 mmHg as determined using the right cardiac catheter test [7,8]. Patients with MCTD have a higher risk of PAH given that 7%–10% of such patients develop PAH [2]. Moreover, the leading cause of death in patients with MCTD is PAH [2,3]. MCTD with PAH is treated by the combination of immunosuppressant agents and pulmonary vasodilators. Regarding pulmonary vasodilators, epoprostenol is useful, especially in the preoperative period [9]. During surgery under general anesthesia, the mortality rate of PAH is 3.5% because of heart and respiratory failure [6]. The current case is rare in coexisting sigmoid colon cancer in a patient with MCTD and PAH. Therefore, we believed that the risk of surgical complications was high in this patient. However, because the patient was relatively young and her tumor was in an early stage, we decided to perform surgery. We planned to switch the PAH treatment from an oral agent to intravenous epoprostenol only in the preoperative period. We selected open surgery and avoided laparoscopic surgery. Laparoscopic colorectal surgery requires a head-down position and pneumoperitoneum, which can elevate the systematic vascular resistance index and compression of the lungs. These complications can increase the risk of heart and respiratory failure in patients with PAH. Because we selected open surgery, severe intraoperative and postoperative complications did not occur.

PAH is associated with portal hypertension [4]. According to Hojo et al., the level of ligation of the IMA and IMV is essential for preventing stoma edema [6]. Furthermore, there is no difference in oncological outcomes between the ligation level, origin of the IMA, and the LCA [10]. We ligated the IMA and IMV at below the branch of LCA. Consequently, marked stoma edema and other severe complications were not observed. The patient had a long history of steroid use for MCTD, which carries a high risk of anastomotic leakage [11]. Thus, we selected Hartmann’s operation and avoided severe complications.

4. Conclusions

We experienced a case of MCTD complicated by severe PAH. It is crucial to control mPAP via treatment with epoprostenol. Open surgery, stoma and the ligation level of the IMA and IMV are essential to avoid stoma complications in such patients.

Ethical approval

This surgical technique was not performed in the context of a study that need ethical approval.
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