The screening program, incidence, and treatment for tumors in patients with severe motor and intellectual disabilities

Running title: Tumor surgery in SMID patients

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Abstract

Background: Recently, aging in patients with severe motor and intellectual disabilities (SMID) has been causing serious problems. There have been few reports about treatment for tumors in patients with SMID.

Methods: The treatments performed for tumors of 12 SMID patients were examined.

Results: Blood tests and ultrasonography were useful in screening. With regard to treatment, the surgery for SMID patients was performed in the same manner as in healthy cases, and the results were generally satisfactory, without major complications. Typically, the patients were diagnosed in advanced stages; many metachronous double cancers were observed. Conclusions: We reported a tumor therapy to the patients with SMID. Our therapy provides satisfactory results for patients with SMID and their families. Further studies are required to clarify the clinical significance of the screening test and the tumor operative method for the patients with SMID.

Key words: severe motor and intellectual disabilities, tumor, surgery, intestinal paralysis, bowel motility
Introduction

A patient with severe motor and intellectual disabilities (SMID) is defined as one who is bedridden or able to sit, crawl, or walk with support, and has an intelligence quotient (IQ) less than 35, according to Oshana’s classification criteria.¹

In the late 1950s, the management of SMID patients became a major social problem. At that time, it was extremely rare that such patients would live up to the age of 15 years, but the life expectancy of SMID patients has now become longer. In the 1990s, Evenhuis et al.² reported the overall morbidity and mortality in the ageing group with severe intellectual disability. The patients are now getting older, and due to aging, various functional disorders occur. Not only psychomotor function, but also their swallowing function deteriorates. In addition, the risks of bone fractures and infections also increase.³ Malignant tumors are reported as the cause of death of SMID patients.⁴ ⁵ There are few reports comparing the incidence of tumors in patients of the same age with and without SMID. Tumors in SMID patients tend to be diagnosed in advanced stages. Therefore, we always have to keep in mind the potential existence of a tumor in patients with SMID. Treatments for various diseases have been carried out for SMID patients,¹ ⁶ ⁷ but there are few reports of treatments for tumors. The present study examined surgical treatments for tumors in patients with SMID at National Hospital Organization Ehime Medical Center.
and Ehime University Hospital.

Materials and methods

Patient data

Twelve patients were diagnosed with neoplastic lesions from January 2010 to December 2020. There were three benign tumors, and one of them was resected. There were ten malignant tumors, and eight of them were resected. In two patients, the tumors could not be resected because of their advanced stages; therefore, these patients were given best supportive care (Fig. 1). Cancer surgery was performed for ten patients with SMID. Of these cases, hospital records were available for nine, and they were reviewed. This study was approved by the Institutional Review Boards of National Hospital Organization Ehime Medical Center and Ehime University Graduate School of Medicine. The need for informed consent was waived due to the retrospective design of the study.

Operative procedure

The surgery was performed at two hospitals, National Hospital Organization Ehime Medical Center and Ehime University Graduate School of Medicine. All operations were performed under general anesthesia.
Results

Twelve patients were diagnosed with tumors during the study period (Table 1). The age of the patients was 49.3±10.7 (mean±SD) years, with an age range of 27-67 years. There were six male and six female patients. There were three benign tumors (myoma uteri, uterine polyp, and benign prostate hypertrophy). Surgery was performed for the uterine polyp. The other patients were only observed. There were nine malignant tumors, and surgery was performed in seven (three male and four female) of the patients. The age of the patients was 46.0±9.7 (mean±SD) years, with an age range of 27-57 years. There were three cases of colorectal cancer (stages I, IIa, and IIIb), two of breast cancer (IIA and IIB), one of seminoma (stage I), one of hepatocellular carcinoma (stage IV), one of gallbladder cancer (stage IV), and one of thyroid cancer (stage IV).

Eight cases underwent surgery. There were no postoperative complications. The patients’ families were satisfied with the results.

Three patients had repeated cancers. One patient was diagnosed with bladder cancer and rectal cancer, one had double breast cancers, and one had double seminomas. Two patients who did not undergo surgery died from their cancers. When they were diagnosed, their cancers were already too far advanced for surgery. Patients who underwent radical surgery had no recurrences.
Discussion

Screening is very important for patients with SMID. In patients with SMID, the diagnostic process is very important. Usually, patients with SMID cannot complain of abdominal pain and other chief complaints, and they often cannot communicate sufficiently. The general population has regular screening examinations for cancer, but patients with SMID cannot undergo them. The diagnostic processes in the present cases were examined. At the National Hospital Organization Ehime Medical Center, blood tests (including serum levels of tumor markers CEA, AFP, CA19-9, PSA, CA125, SCC), fecal occult blood tests, and ultrasound examinations (abdominal, mammary gland, thyroid gland, etc.) were performed twice a year for patients with SMID over the age of 40 years. CEA is considered to be a tumor marker for lung cancer, breast cancer, gastric cancer, biliary tract cancer, pancreatic cancer, and colon cancer. AFP, CA19-9, PSA, CA125, and SCC are recognized as tumor markers for hepatocellular carcinoma, biliary tract cancer, pancreatic cancer, prostate cancer, breast cancer, pancreatic or ovarian cancer, and esophageal cancer or uterine cancer. In patients with SMID, the serum levels of CEA and SCC tend to remain mildly high. The fecal occult blood test had a positive rate of about 20% (data not shown). A few cases remained positive for a long time. In the present cases, one colon cancer was diagnosed because of a high CEA level. Measuring CEA levels
appears effective as tumor screening for patients with SMID. To screen patients with SMID for breast cancer, we instruct the caregiver to observe carefully when the patient takes a bath. In the present cases, all patients with breast cancer were diagnosed by observation during bath time. Therefore, screening examinations are very important for patients with SMID.

Let us consider the features of cancer in patients with SMID. There are some reports on the frequency of cancer in patients with SMID.\(^1,2\) It has been reported that the frequencies of prostate cancer, ureteral cancer, and lung cancer are low. In the present cases, none of these types of cancers was found. In addition, it was reported that the frequencies of gallbladder cancer and thyroid cancer were high. In the present cases, one gallbladder cancer and one thyroid cancer were detected. It has also been reported that these tumors are detected in advanced stages.\(^3\) In the present study, eight of 10 (80%) tumors were resectable, with some unresectable cases. Three cases of repeated cancers are included in this report. Further studies, including genetic research, are needed to clarify them in patients with SMID.

The operations for patients with SMID were the same as for patients without SMID. The results were generally satisfactory, without major complications (except for postoperative intestinal paralysis). There is some limited information about cancer
incidence and 5-year survival in SMID patients. Colostomy is an appropriate operative method for rectal cancer in SMID patients, because they often require assisted excretion, and the risk of suture failure can be avoided. Although, only one patient developed ileus, bowel motility of SMID patients after surgery is weak on the whole because they have often intestinal palsy even before surgery. In addition, the antiepileptic drugs they often take may also be associated with intestinal paralysis. It was necessary to pay attention to postoperative intestinal paralysis and to determine what kind of food the patients should eat and when they could start eating. In such patients, tumors are often diagnosed in advanced stages. Therefore, it is necessary to determine the extent of lymph node dissection for each case.

In conclusion, screening and treatments for tumors of patients with SMID were reviewed. Our approach provides results that are satisfactory for patients with SMID and their families. Further studies are needed to clarify the clinical significance of screening tests and the operative methods for tumors in patients with SMID.

**Conflict of interest**

The authors have no conflict of interest to disclose.
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Figure legend

Fig 1. Flow diagram of patients with severe motor and intellectual disabilities

Footnote of Fig 1

BSC : Best supportive care

Footnote of Table 1

MMK : Breast cancer, HCC : Hepatocellular carcinoma, GB Ca : Gallbladder cancer, BPH : Benign Prostate Hypertrophy, CEA : Carcinoembryonic antigen, AFP : Alpha-fetoprotein, CA19-9 : carbohydrate antigen 19-9
Fig. 1

All tumors
n=12

Malignant tumors
n=9

Operation
n=7

BSC
n=2

Bengine tumors
n=3

Operation
n=1

Observation
n=2
| Case | Age | Sex | Diagnosis       | Stage | Therapy          | second primary | outcome | Opportunity of close examination |
|------|-----|-----|-----------------|-------|------------------|----------------|---------|---------------------------------|
| 1    | 52  | F   | Rectal Ca       | IIIB  | Operation        | Alive          | CEA     |                                 |
| 2    | 45  | F   | Colon Ca        | I     | Operation        | Alive          | -       |                                 |
| 3    | 57  | M   | Rectal Ca       | IIA   | Operation        | Alive          | melanin |                                 |
| 4    | 27  | F   | Rt. MMK         | IIA   | Operation        | Alive          | appearance |                                 |
| 5    | 49  | F   | Rt. MMK         | IIB   | Operation        | Alive          | appearance |                                 |
| 6    | 51  | M   | HCC             | III   | Radiofrequency ablation | Alive      | AFP     |                                 |
| 7    | 34  | M   | Thyroid Ca      | IV    | Best supportive care | Dead         | appearance |                                 |
| 8    | 63  | M   | GB Ca           | IV    | Best supportive care | Dead         | CA19-9  |                                 |
| 9    | 53  | M   | Lt. Seminoma    | I     | Operation        | Alive          | appearance |                                 |
| 10   | 67  | F   | Uterine polyp   | -     | Operation        | Alive          | -       |                                 |
| 11   | 43  | M   | BPH             | -     | -                | Alive          | -       |                                 |
| 12   | 51  | F   | Myoma uterus    | -     | -                | Alive          | -       |                                 |