Tumor Screening, Incidence, and Treatment for Patients with Severe Motor and Intellectual Disabilities

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Background: The increasing age of patients with severe motor and intellectual disabilities (SMID) has become a serious concern. Few studies have investigated tumor treatment in this population. **Methods:** Tumor treatments for 12 SMID patients were examined.

Results: Blood tests and ultrasonography were useful for screening. With regard to treatment, surgery for SMID patients was performed in the same manner as for patients without SMID, and the results were generally satisfactory, without major complications. Typically, cancer was diagnosed at an advanced stage, and many metachronous double cancers were observed.

Conclusions: Treatment yielded satisfactory results for patients with SMID and their families. Future studies should examine the clinical significance of screening and tumor operative method for patients with SMID. (J Nippon Med Sch 2022; 89: 212–214)

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

Introduction

A person 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 lower than 35, according to Oshana's classification criteria¹. In the late 1950s, management of SMID patients became a major social problem. At that time, it was rare for such patients to live to age 15 years, but the life expectancy of SMID patients has increased. In the 1990s, Evenhuis et al.² reported overall morbidity and mortality in an aging group with severe intellectual disabilities. This population growing older, and because of aging, various functional disorders occur. Both their psychomotor function and swallowing function deteriorate, and the risks of bone fracture and infection increase³.

Malignant tumors are a reported cause of death for SMID patients^{4,5}; however, few studies have compared tumor incidence in patients of the same age with and without SMID. Because tumors tend to be diagnosed at an advanced stage in SMID patients, the potential presence of a tumor must be considered in such patients. Treatment of a number of diseases has been described for SMID patients^{1,6,7}, but few reports have described tumor treatment. 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 who were diagnosed as having neoplastic lesions during the period from January 2010 through December 2020 were studied. There were three benign tumors, one of which was resected, and 10 malignant tumors, eight of which were resected. In two patients, the tumors could not be resected because of the advanced stage; these patients received best supportive care (**Fig.**). Cancer surgery was performed for 10 pa-

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tients with SMID, and hospital records were reviewed for the nine patients for which we had complete information. 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 because of 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

Tumors were identified in 12 patients during the study period (**Table**). The mean (±SD) age of the patients was

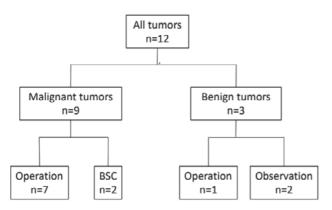


Fig. Flow diagram of patients with severe motor and intellectual disabilities BSC: Best supportive care 49.3±10.7 years (range 27-67 years). There were six men and six women. There were three benign tumors (myoma uteri, uterine polyp, and benign prostate hypertrophy), and surgery was performed for the uterine polyp. The other patients were only observed. There were nine malignant tumors, and surgery was performed for seven of the patients (three men; four women). The age of the patients was 46.0±9.7 years (range 27-57 years). There were three cases of colorectal cancer (stages I, IIa, and IIIb), two cases of breast cancer (IIA and IIB), one case of seminoma (stage I), one case of hepatocellular carcinoma (stage IV), one case of gallbladder cancer (stage IV), and one case of thyroid cancer (stage IV). Eight patients underwent surgery. There were no postoperative complications, and the patients' families were satisfied with the results.

Cancer recurrence was noted in three patients: one with bladder cancer and rectal cancer, one with double breast cancer, and one with double seminoma. Two patients who did not undergo surgery died from cancer, as the disease was already too far advanced for surgery. No recurrence was noted in patients who underwent radical surgery.

Discussion

Screening and accurate diagnosis are critical for patients with SMID⁸. Persons with SMID are usually unable to report abdominal pain or other important symptoms and often cannot communicate sufficiently. The general population undergoes regular screening examinations for cancer, but persons with SMID do not.

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	Ι	Operation		Alive	-
3	57	Μ	Rectal Ca	IIA	Operation	Bladder Ca	Alive	melena
4	27	F	Rt. MMK	IIA	Operation	Lt. MMK	Alive	appearance
5	49	F	Rt. MMK	IIB	Operation		Alive	appearance
6	51	М	HCC	III	Radiofrequency ablation		Alive	AFP
7	34	Μ	Thyroid Ca	IV	Best supportive care		Dead	appearance
8	63	Μ	GB Ca	IV	Best supportive care		Dead	CA19-9
9	53	М	Lt. Seminoma	Ι	Operation	Rt. Seminoma	Alive	appearance
10	67	F	Uterine polyp	-	Operation		Alive	-
11	43	Μ	BPH	-	-		Alive	-
12	51	F	Myoma uterus	-	-		Alive	-

Table Demographic and clinical characteristics of cancer patients with severe motor and intellectual disabilities

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

The diagnostic process for the present patients was examined. At the National Hospital Organization Ehime Medical Center, blood tests (including serum levels of tumor markers CEA, AFP, CA19-9, PSA, CA125, and SCC), fecal occult blood tests, and ultrasound examinations (abdominal, mammary gland, thyroid gland, etc.) were performed twice a year for patients with persons with SMID older than 40 years. CEA is considered 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, serum CEA and levels SCC tend to be slightly elevated. Fecal occult blood testing yielded a positive rate of about 20% (data not shown), and a few patients remained persistently positive. Colon cancer was diagnosed in one of the present patients 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 carefully observe them when they bathe. Among the present patients, those with breast cancer were diagnosed by observation during bathing. Therefore, screening examinations are essential for patients with SMID.

Some studies reported cancer frequency in patients with SMID^{1,2}. The frequencies of prostate cancer, ureteral cancer, and lung cancer are low, and these cancers were not observed in the present patients. However, the frequencies of gallbladder cancer and thyroid cancer were high. In the present series, one case of gallbladder cancer and one case of thyroid cancer were diagnosed. One study reported that these tumors are usually detected at an advanced stage³. In the present study, eight of 10 (80%) tumors were resectable. Cancer recurred in three of the present patients. Further studies, including gene studies, are needed to clarify tumor characteristics in patients with SMID.

The operations for patients with SMID were the same as those for patients without SMID. The results were generally satisfactory, without major complications (except for postoperative intestinal paralysis). Data on cancer incidence and 5-year survival are limited for 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 in SMID patients after surgery is usually weak because they have often intestinal palsy preoperatively. In addition, the antiepileptic drugs they often require are associated with intestinal paralysis. It was necessary to consider the possibility of postoperative intestinal paralysis and to determine the optimal type and timing of food for patients. In such patients, tumors are often diagnosed at an advanced stage. Therefore, the extent of lymph node dissection must be individualized.

In conclusion, tumor screening and treatment for patients with SMID were reviewed. Our approach yielded results that were 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 declare no conflicts of interest.

References

- 1. Shima H, Kitagawa H, Wakisaka M, et al. The usefulness of laryngotracheal separation in the treatment of severe motor and intellectual disabilities. Pediatr Surg Int. 2010; 26:1041-4.
- 2. Evenhuis HM. Medical aspects of ageing in a population with intellectual disability: III. Mobility, internal condition and cancer. J Intellect Disabil Res. 1997;41(Pt 1):8–18.
- Matsuda S, Nakamura M, Hirano E, et al. Characteristics of human metapneumovirus infection prevailing in hospital wards housing patients with severe disabilities. Jpn J Infect Dis. 2013;66:195–200.
- Patja K, Eero P, Iivanainen M. Cancer incidence among people with intellectual disability. J Intellect Disabil Res. 2001;45:300–7.
- Sulivan SG, Hussain R, Threlfall T, et al. The incidence of cancer in people with intellectual disabilities. Cancer Causes Control. 2004;15:1021–5.
- Wong K, Glasson EJ, Jacoby P, et al. Survival of children and adolescents with intellectual disability following gastrostomy insertion. J Intellect Disabil Res. 2020;21:497–511.
- Takano K, Kurose M, Mitsuzawa H, et al. Clinical outcomes of tracheoesophageal diversion and laryngotracheal separation for aspiration in patients with severe motor and intellectual disability. Acta Oto-Laryngologica. 2015;135:1304–10.
- 8. Tretarre B, Bourgarel S, Stoebner-Delbarre A, et al. Breast cancer and screening in persons with an intellectual disability living in institutions in France. J Intellect Disabil Res. 2017;61:266–78.

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