CASE REPORT

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Primary sebaceous carcinoma of the lacrimal gland treated by carbon ion radiotherapy

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Abstract  Sebaceous carcinoma is a rare primary neoplasm of the lacrimal gland and, to the best of our knowledge, only six cases have been reported previously. Sebaceous carcinoma of the orbit more commonly occurs as secondary invasion from the eyelid, but may occur by way of metastatic spread from elsewhere in the body. We describe a patient who presented with a rapidly growing neoplasm of the lacrimal gland which, histologically, was a sebaceous carcinoma. The eyelid was entirely normal on examination. In this patient we found a single tumor cell line within the normal lacrimal gland. In this article we describe the diagnosis and treatment with carbon ion radiotherapy of primary sebaceous carcinoma of the lacrimal gland, a condition not previously reported in Japan.

Key words  Carbon ion radiotherapy · Heavy charged particle (therapy) · Sebaceous carcinoma · Lacrimal gland

Introduction

Clinical trials of carbon ion radiotherapy for cancers were initiated in June 1994 at the Hospital, Research Center for Charged Particle Therapy, National Institute of Radiological Sciences (NIRS), using the world’s first heavy ion medical accelerator, in Chiba (HIMAC) dedicated to medical purposes. Heavy charged particles such as carbon and neon ions excel in their physical dose distribution and high biological effectiveness. Preliminary results of phase II clinical trials have shown extremely favorable therapeutic results in the treatment of tumors otherwise intractable with conventional photon radiation.

Sebaceous carcinoma arising from the lacrimal gland is, histologically, a very rare malignant tumor. So rare is the condition that only six cases have been reported worldwide. Here, we report on a patient with sebaceous carcinoma of the lacrimal gland treated with carbon ion radiotherapy alone at the NIRS hospital.

Case report

A 73-year-old Japanese man visited an ophthalmologist in April 2001, complaining of some vague discomfort in his left eye. He was diagnosed as having keratitis filamentosa, but no other physical signs were in evidence. On follow-up examinations, however, the symptoms showed no decrecence and the patient was therefore referred to the Department of Ophthalmology of the National Okayama Medical Center, in August 2001. His condition was diagnosed as lacrimal gland tumor, and the tumor was surgically resected. Pathological examination revealed the presence of sebaceous carcinoma (T4N0M0). Further surgery after the first resection was planned to remove the remaining tumor, but this would have necessitated enucleation of the residual eyeball. The patient was informed of the difficulty of complete surgical resection and was advised to undergo radiotherapy. He then gave his informed consent to a course of carbon ion radiotherapy and was referred to the NIRS hospital in September 2001.

On examination, nonaxial proptosis was apparent, with a mass palpable in the left upper quadrant beneath the orbital rim. No preauricular or cervical lymph nodes were palpable. The horizontal and vertical movement of the left eye was severely restricted. Magnetic resonance imaging (MRI) demonstrated the presence of a tumor in the left lacrimal gland (Fig. 1), this being the primary tumor that had caused an invasive spread to the left eyeball, left optic canal, and left cavernous sinus. The size of the tumor was about 65 mm × 25 mm × 25 mm. On hematoxylin and eosin staining, the
Lacrimal gland tissue was found to be infiltrated by a carcinoma with large neoplastic cells and prominent nucleoli, composed of nets and sheets of large epithelial cells with abundant vacuolated cytoplasm (Fig. 2). Many cells were vacuolated and contained lipid matter, as confirmed by fat stains on unprocessed, fixed material. Thus, these cells were considered to contain extensive fat vesicles. These histological findings were consistent with the accepted diagnostic criteria for sebaceous carcinoma. Positron emission tomography (PET) using $^{11}$C-methionine was performed and revealed increased accumulation in the left orbital tumor. $^{11}$C-methionine PET also pointed to the possibility of invasion into a part of the optic canal behind the orbit (Fig. 3). The above findings were taken as evidence that the patient qualified for phase II clinical trials using carbon ion radiotherapy for the treatment of cancers of the head and neck.

An eye test was performed prior to the initiation of carbon ion radiotherapy and showed that the patient's visual acuity was 1.5 for the right eye (OD) and 0.2 for the left eye (OS). The patient also had a poor light reflex in the left eye, as he was unable to open the eye due to pressure in the ocular fundus caused by the left eye tumor. He also had pain in the left eye.

Heavy charged particle (carbon ion) therapy was performed, using a total dose of 57.6 GyE in 16 fractions over 4 weeks. Figure 4 shows the dose distribution associated with 290 MeV/n carbon ion irradiation. A side effect was noted during the treatment course, in terms of a mild skin reaction detectable on the left upper eyelid.

Immediately after commencing the therapy, however, the patient was able to open the eye and the ocular pain disappeared completely. Seven months after completion of the treatment, his light reflex was also restored to normal, and his vision had improved to 1.2 (OD) and to 1.0 (OS).

MRI performed 6 months after first treatment showed evidence of marginal tumor invasion to the left zygomatic bone concentrated around the upper left maxillary sinus. The marginal tumor had thus recurred and was again treated by carbon ion radiotherapy. The outcome was favorable, with no decline in vision and visual function.

At present, 16 months after completion of the second course of carbon ion radiotherapy, the primary tumor that had been found by MRI has completely disappeared (Fig. 1b). The patient is still alive without signs of lymph node metastasis or distant metastasis.