Dropped Head Syndrome Induced by Chemoradiotherapy for Nasopharyngeal Carcinoma: A Case Report

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‘Dropped head syndrome’ (DHS) is characterized by severe weakness of the muscles of the back of the neck, resulting in chin-on-chest deformity. Dropped head syndrome induced by radiotherapy is very rare. We report a case of DHS following chemoradiotherapy with a total of 64.8 Gy in 36 fractions for nasopharyngeal carcinoma.

Key words: dropped head syndrome – chemoradiotherapy – nasopharyngeal carcinoma

INTRODUCTION

Dropped head syndrome (DHS) is characterized by severe weakness of the muscles of the back of the neck, resulting in chin-on-chest deformity. In 1992, Suarez and Kelly (1) reported four cases of non-inflammatory myopathy causing a downward sagging of the neck showing a relatively confirmed muscle weakness of neck extension, and this syndrome was named DHS. In most cases, DHS is caused by a specific generalized disorder, including neuromuscular diseases, e.g. myasthenia gravis, motor neuron disease, congenital myopathy, chronic inflammatory demyelinating polyneuropathy and mitochondriopathy (2,3). When the cause of DHS is unknown, it is collectively called isolated neck extensor myopathy (INEM). DHS appears to be a very rare complication of radiotherapy. Radiation-induced DHS was first reported in 1998 by Johansson et al. (4). Thus, there are only several reports that mantle irradiation for Hodgkin’s disease (HD) induced DHS (5–11). To our knowledge, there have been no previous reports on radiation-induced DHS in patients with head and neck carcinoma but not HD, including reports in Japan.

We report a case of DHS in a 59-year-old man treated for nasopharyngeal carcinoma by chemoradiotherapy with a total dose of 64.8 Gy in 36 fractions.

CASE REPORT

A 59-year-old man visited our hospital complaining of nasal congestion and left ear fullness in March 2006. Computed tomography (CT) and magnetic resonance imaging showed a large tumor of the left nasopharynx extending to the oropharynx with bilateral cervical lymph nodes’ swelling. Other examinations including chest X-ray and FDG-PET were performed to investigate distant metastases. No distant metastasis was observed. A biopsy from the nasopharynx revealed a moderately differentiated squamous cell carcinoma. The patient was given a diagnosis of Stage III nasopharyngeal carcinoma (T2bN2M0) under the TNM classification system (UICC Sixth Edition, 2002). He had no past history or family history of neuromuscular diseases.

The patient received radiotherapy with alternating chemotherapy. Radiation therapy was delivered by a high-energy photon with energy of 10 MV X-ray. The patient was treated using two lateral parallel opposing fields encompassing the primary tumor and upper and middle cervical lymph nodes with a daily dose of 1.8 Gy, five times a week. After a total dose of 39.6 Gy in 22 fractions, boost therapy was given for the primary lesion with localized field using the same radiation technique and involved right accessory lymph nodes with a 9 MeV electron beam. A total dose of 25.2 Gy was
boosted with the same fraction size for the primary lesion and involved cervical lymph nodes. Lower neck-supraclavicular and infraclavicular lymph nodes were irradiated by a 6 MV photon beam prophylactically with a single anterior field for a total dose of 39.6 Gy in 22 fractions (Fig. 1).

A regimen of alternating chemotherapy consisted of 5-FU 700 mg/m² on days 1–5 and cisplatin 50 mg/m² on days 6 and 7. Chemotherapy was repeated twice, prior to and during radiotherapy. Grade 3 mucositis was observed and morphine and steroids were administered to reduce these symptoms. Primary tumor and cervical lymph node metastases completely disappeared at the end of treatment. He was followed up monthly for 1 year after treatment and then at every 3 months. There was no evidence of tumor recurrence and/or distant metastases.

The patient, however, became aware of the difficulty of looking at the front when riding a motorcycle 2 years after treatment. This symptom progressed gradually and he showed abnormal head posture in flexion, resulting in chin-on-chest deformity (Fig. 2). He was readmitted to our hospital for a DHS diagnosis. Laboratory findings including thyroid function and immunology tests showed no abnormality. Serum creatine kinase level and creatine kinase isozymes levels were also within normal limits. On examination, no sensory disturbance or deteriorated muscular strength was observed, and the deep tendon reflex was normal.

Electromyographic (EMG) findings showed myogenic changes. CT of the cervical spine showed a marked atrophy of bilateral paraspinal muscles (Fig. 3). No evidence of spinal cord pathology was observed. Muscle biopsy was not performed. Therefore, DHS (INEM) was diagnosed and he has treated with induction chemoradiotherapy.

**DISCUSSION**

DHS as a result of radiotherapy is a rare but striking clinical entity. There have been some case reports in the literature (4–11), but no studies have been reported in Japan. It has been rarely described as a late effect of external beam radiotherapy for HD (3). Patients with cervical or mediastinal HD classically underwent extended-field (mantle-field) radiotherapy. The symptoms developed many years after mantle field radiotherapy, ranging from 5 to 30 years (most cases occurring by 10 to more than 20 years) (12,13). The weakness
may progress over the years but does not extend beyond the initially involved muscle groups. In the current case, DHS developed 2 years after chemoradiotherapy. Astudillo et al. (14) reported cervical dystonia mimicking DHS 3 months after radiotherapy for laryngeal carcinoma. They reported that DHS was ruled out because spontaneous rapid remission was not consistent with DHS.

The mechanism of late-onset radiation-induced DHS remains unclear although pathophysiologically, it is thought to result from primary muscle damage or anterior horn or root lesions at the upper cervical level within the radiation field. The current case demonstrates myogenic changes on electromyogram. EMG findings were, however, heterogeneous, and both myopathic and neurogenic electromyographic changes have been reported (8,11,13). The absence of an unequivocally neurogenic EMG pattern advocates for a more complex mechanism, probably implying some myogenic participation.

Irradiation has not been considered to cause relatively little or no direct damage to mature muscle fibers. High radiation doses are known to proliferate the fibrous tissue into muscle secondarily by damaging endothelial cells and vascular smooth muscle cells resulting in proliferation of fibrous tissue. Except for the current case, all of radiation-induced DHS appeared after mantle radiation therapy for HD (4–11). The reason why radiation-induced DHS developed in patients with HD has not been clear. It has, however, been considered that the radiation dose was relatively little for damaging mature muscle fibers because DHS was induced by radiation therapy with a total dose of <40 Gy. In fact, radiation myositis is a rare adverse effect of radiation therapy for HD. These are the two reasons considered as to why radiation-induced DHS appears most commonly on HD, although total radiation doses are smaller compared with those employed to head and neck cancer. The mantle field encompasses the submandibular, cervical, supraclavicular, infraclavicular, axillary, mediastinal subcranial and hilar lymph nodes. Whole neck irradiation may cause DHS. In the current case, the radiation field almost encompassed the whole neck including the posterior neck (up to 39.6 Gy as shown in Fig. 2), although the neck irradiation field for head and neck cancer does not encompass the posterior part of the neck ordinarily. Another reason may be enhancement effects of chemotherapy. Chemotherapy with MOPP (CPA/VCR/PCZ/PSL) or ABVD (DXR/BLM/VLB/DTIC) is frequently combined with radiation therapy for the treatment of HD. The agents combined with mantle irradiation might enhance radiation-induced chronic adverse effects (12,13,15). Some anticancer drugs are known to induce neuromuscular complications. Peripheral neuropathy is an important dose-limiting toxic side effect of two commonly utilized agents, vinca alkaloids and platinum compounds which were used in the current case (12). Patients developing DHS demonstrated muscle atrophy within the radiation portal, selectively. A synergistic effect of both treatment modalities in the pathogenesis of this condition cannot be excluded. Physicians using chemotherapy combined with radiation for head and neck cancer should be aware of its potential to induce a delayed form of radiosensitization as the use of anticancer agents in combination of radiation therapy has increased.

Treatment of DHS is considered specific to the underlining disease. Treatment of DHS is less effective but more supportive, including employing a cervical collar to maintain the head in an upright position, because it is considered benign and it does not spread or become worse.

We have reported here a rare case of DHS following chemoradiotherapy for nasopharyngeal cancer. To our knowledge, there has been no previous description of radiation-induced DHS in patients with head and neck cancer but not HD.

Conflict of interest statement
None declared.

References