Dropped Head syndrome, Late Complication of Radiotherapy for Nasopharyngeal Carcinoma

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ABSTRACT
The dropped head syndrome (DHS) is a disabling condition caused by severe weakness of the muscles of the neck resulting in progressive reducible kyphosis of the cervical spine with chin-on-chest deformity. A variety of musculoskeletal disorders can cause this syndrome. However, radiotherapy of the neck remains one of the rarest causes. Rowin Bahita The primary disease in the majority of the cases with radiation-induced dropped head syndrome has been Hodgkin lymphoma. There is also nasopharyngeal carcinoma where radiation could cause DHS and neck dystonia, each in one rare occasion.

Herein, we present the second example of DHS after radiotherapy for nasopharyngeal carcinoma in a 50- year-old male developing such a syndrome 3 years after radiotherapy. The patient could hold his head upright in its normal anatomic position for more than 30 seconds and he was unable to perform his daily activities without wearing a collar. However, he was reluctant to undergo surgery and preferred wearing a collar.

KEYWORDS: Cervical Spine, complication, dropped head syndrome, nasopharyngeal carcinoma, radiotherapy

INTRODUCTION
Dropped head syndrome or head ptosis is a reducible flexion deformity of the neck that is caused by a weakness of the extensor muscles of the neck resulting in the chin-on-chest deformity (13,19,21-23). Dropped head syndrome (DHS) might be seen in isolation or in association with a variety of generalized neuromuscular disorders (11, 13, 14, 19, 21-23).

DHS as a late sequel of radiotherapy for malignant pathologies of the neck has been rarely reported (1,3,5,9,10,12,16-18,20). Extensive radiotherapy for Hodgkin lymphoma has been the most common cause of this pathology where radiotherapy for nasopharyngeal carcinoma has been the cause of DHS in one rare occasion (1,3,5,9,10,12,16-18,20). Muscular atrophy and lower motor neuron injury secondary to isolated anterior horn cell injury from radiotherapy emerge as the most likely causes of radiation-induced DHS (1,3,5,9,10,12,16-18,20).

Herein, a new case of DHS developing three years after radiotherapy for nasopharyngeal carcinoma is presented. The patient could cope with this disabling condition by wearing a collar and refused any surgical intervention.

From the medicolegal point of view, the possibility of DHS in patients who will undergo extensive radiotherapy of the neck should be mentioned despite its rarity.

CASE REPORT
A 50-year-old man was referred to our institute because of difficulty in holding his neck for more than 30 seconds, resulting in a chin-to-chest deformity (Figure 1). The head and neck deformity returned to normal position when laying supine. He had history of nasopharyngeal carcinoma...
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where the diagnosis had been made with biopsy. Extensive radiotherapy had been recommended and administered 2 weeks after confirmation of the diagnosis. The patient made an uneventful recovery. About 2 years after radiotherapy he noticed some atrophy in the muscles of his neck. Gradually, holding the head and looking forward became difficult. At that time, he could hold his head with extreme effort for about 4 to 5 minutes, becoming exhausted thereafter. The pathology progressed with time until he could now only look forward for 30 seconds. The neurological exam revealed atrophy of the neck muscles, more marked on the paraspinal muscles. Laboratory studies showed normal serum creatine kinase level, thyroid function tests, and erythrocyte sedimentation rate. MRI was normal except for atrophy of the paraspinal muscles. Electromyography of the paraspinal muscles revealed mixed motor unit action potentials consisting of normal-amplitude, short duration polyphasic potentials with a reduced recruitment myopathic pattern in the paraspinal muscles. Cervicothoracic stabilization and fusion was proposed but the patient preferred to wear a collar and refused surgery.

DISCUSSION

The dropped head syndrome (DHS) is a disabling condition caused by severe weakness of the neck extensor muscles causing progressive reducible kyphosis of the cervical spine and the inability to hold the head up (13, 19, 21-23). At its extreme, the condition may lead to a "chin-on-chest deformity" where the chin rests on the chest wall and the patient is unable to look straight ahead (13, 19, 21-23). Heffner et al were the first to define the dropped head syndrome in 1977 (13, 19, 21-23). The weakness may occur in isolation or in association with a generalized neuromuscular disorder. Three causes of isolated weakness of the extensors have been described: isolated neck extensors myopathy (INEM) of unknown etiology (idiopathic), isolated myositis of extensor muscles of the neck, and isolated extensor myopathy as a sequel of neck radiotherapy for a variety of malignant pathologies (1,3,5,9,10-13,14,16-23). Idiopathic isolated neck extensors myopathy is a disease of the elderly, but the age of the patient is variable in radiation-induced myopathy as well as in isolated myositis.

Radiotherapy of the neck might damage the spinal cord and corresponding roots, causing five different syndromes (8). The first syndrome is that of a transient myelopathy, mostly manifesting with Lhermitte's symptom and other sensory disturbances. The second syndrome is acute myelopathy being manifested with rapidly evolving quadriplegia as a result of spinal cord infarction (7,8) the third syndrome presents as a chronic progressive myelopathy (7,8). In all types of myelopathy, the main pathological features are asymmetrical demyelination of the lateral columns and to a lesser degree the posterior and anterior columns (7). Surprisingly, the cervical region is the most common part of the cord to be affected, and most reports of the pathology of radiation myelopathy have been of lesions at this site (7,8). In the fourth syndrome, radiation can cause a single root or several roots paresis, unilaterally or bilaterally (2,6,25,26). The case report by Brander and colleagues describing diaphragm weakness due to one of the many of severe diaphragm weakness due to phrenic nerve damage from radiotherapy is an example (6). Delayed lumbar roots affection mostly presented with foot drop and even cauda equina syndrome in radiotherapy of the lumbar lesion can clarify radiation induced preganglionic root

Figure 1: Photograph of the patient showing a chin-on-chest deformity.
affection (25,26). In the fifth form, radiation cause selective damage of the anterior horn cells, resulting in limited lower motor neuron disease (1,3,5,9,10,12,16-18,20). Actually, the pathogenesis of late-onset radiation-induced DHS can be explained with the 5th syndrome where radiotherapy might compromise the microcirculation of anterior horn cells innervating the extensor muscles of the neck. Furthermore, there is another hypothesis for this pathology that is less probable. Accordingly, radiation-induced DHS is attributed to primary damage of the paraspinal muscles or myopathy. However, it seems that both these hypotheses might be involved in occurrence of radiation-induced DHS (1, 3, 5, 9, 10, 12, 16-18, 20).

Furthermore, radiation-induced cervical anterocollis dystonia has been reported in a 57-year-old man suffering from well-differentiated squamous cell carcinoma of the larynx where post-radiation segmental spinal cord damage has been implicated in the pathogenesis of this disabling condition (4).

Tsang et al. in 1999 reported a localized neuromyotonia of neck muscles after radiotherapy for nasopharyngeal carcinoma characterized by continuous and spontaneous muscle fiber contractions. Radiation-induced hyperexcitability of motor nerves has been suggested as a cause of this rare pathology (24).

Spasmodic torticollis has also been described in a 52-year-old man who had received radiotherapy for a Pancoast tumor due to lung adenocarcinoma (15).

Other late side effects of radiotherapy are hypothyroidism, dryness of the mouth, weakness of the diaphragm due to phrenic nerve affection and plexopathy (20).

Radiation-induced DHS has been mostly seen in Hodgkin’s Lymphoma (1,3,5,9,12,16-18,20). Standard radiotherapy for Hodgkin’s lymphoma is the mantle field which encompasses from the submandibular region to hilar lymph nodes (1, 3, 5, 9, 12, 16-18, 20). This large field might be responsible for late-onset DHS. The field and the dose of radiotherapy for nasopharyngeal carcinoma could have been the cause of DHS in two reported cases including the current case (10).

Nonetheless, dropped head syndrome regardless of etiology may severely compromise the patient’s quality of life and result in significant disability necessitating appropriate therapy. In dropped head syndrome secondary to isolated neck extensor myopathy, conservative care is limited to strengthening exercises and wearing collars (13,19,21-23). Cervical collars, despite their ability to maintain the head in an upright position, are frequently not tolerated well by the patient and may lead to a pressure ulcer under the chin. Recently, baseball cap orthosis was developed to overcome this problem (19).

Actually, surgery is the treatment of choice in idiopathic INEM for achieving a good outcome and long life expectancy (13, 19, 21-23). However, there is no clear consensus on the optimal treatment or timing in post-radiotherapy INEM owing to the paucity of information regarding surgical intervention. Nonetheless, the type of treatment should be individualized to the patient. Development of scar tissues in the neck muscle and obliteration of the small vessels might hamper surgery where the possibility of the surgical wound dehiscence and post instrumentation infection need to be considered. Therefore, conservative treatment is the preferred mode of management in those with short life expectancy or mal-nutrition whereas surgery is recommended in patients with long life expectancy.

In surgical intervention, correction of kyphosis is the main stay of treatment in dropped head syndrome. Nowadays, cervicothoracic osteosynthesis from C2 to T2 or T3 followed by arthrodesis is the most accepted mode of surgery (13, 19, 21-23).

In summary, the possibility of dropped head syndrome in radiotherapy of neck pathologies should be borne in mind. In order to prevent medicolegal consequences, it should be added to the informed consent of radiotherapy departments. The present case is consistent with DHS that strongly seems to be related to radiotherapy.

In contrast to idiopathic isolated neck extensors myopathy where surgical stabilization seems the best option of treatment, the treatment should be individualized in post radiotherapy isolated neck extensors myopathy and surgery can be recommended only in those with long-life expectancy.

REFERENCES


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