Middle-aged woman with dropped head syndrome due to remote exposure to mantle field radiation

Biplab Saha ¹, Santu Saha,² Woon Hean Chong ³

¹Pulmonary and Critical Care Medicine, Ozarks Medical Center, West Plains, Missouri, USA

²Internal Medicine, Bangladesh Medical College, Dhaka, Dhaka District, Bangladesh

³Pulmonary and Critical Care Medicine, Albany Medical Center, Albany, New York, USA

Correspondence to
Dr Biplab Saha;
spanophiliac@yahoo.com

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DESCRIPTION

A 61-year-old woman presented to the office for evaluation of progressive neck muscle weakness over 10 years. She reported initial difficulty with an eventual inability to lift up her head without manual support when assuming sitting from a supine position. She was also unable to keep her head up for a prolonged period of time. She denied any double vision, drooping of the eyelid (especially at the end of the day), difficulty with swallowing or speech, extremity weakness or fluctuation of her symptoms. Her medical history was significant for Hodgkin's lymphoma as a teenager, and she received mantle field radiation in the 1970s.

Physical examination revealed muscle wasting involving the posterior cervical muscle groups (figure 1A). Anterior cervical muscles were contracted, and the overlying skin was thickened and telangiectatic. Atrophy of bilateral shoulder girdle muscles was also present. There was a significant weakness with flexion, extension and lateral rotations of the neck. No spontaneous fasciculation was noted. Cranial nerve and sensory examinations were normal. She had no motor or sensory deficits in her extremities. The cardiopulmonary examination was normal. Laboratory workup including aldolase and creatine phosphokinase levels was unremarkable. Autoantibody testing against the acetylcholine receptor and muscle-specific tyrosine kinase was negative. CT scan of the neck revealed muscle atrophy in both anterior and posterior muscle groups (figure 1B). Needle electromyography (EMG) revealed combined myopathic and neurogenic abnormalities involving the posterior cervical and paraspinal muscles. The EMG and nerve conduction studies of bilateral upper extremities were normal.

'Dropped head syndrome (DHS)' is a rare long-term complication of extended field radiation (EFR), such as mantle field radiation. It refers to atrophy and weakness of the cervical, paraspinal and shoulder girdle muscles, leading to the inability to keep the head up for a prolonged time. The symptoms typically develop 5–30 years after high-dose radiation therapy.¹ Only a number of cases have been reported in the literature.^{2–4}

Radiation-induced myopathic or neurogenic processes or a combination of both are likely involved in the pathogenesis of DHS.⁵ Although mature myocytes are generally resistant to radiation-induced damage, high-dose radiation may cause altered proliferation of muscle precursor cells and affect spontaneous muscle regeneration. In addition, damage to the capillaries supplying the

muscle fibres may also be affected.^{6,7} Collagenous proliferation and fibrous tissue deposition have also been reported.⁸ Development of muscle contracture has previously been reported. An EMG may show neurogenic or myopathic abnormalities or a combination of both.⁹

Muscle biopsies of the affected muscle groups have shown non-specific findings, such as nemaline myopathy.³ In general, myopathy in radiation-induced muscle atrophy does not reveal any evidence of muscle inflammation. As the spinal cord is exposed to radiation during EFR, damage to the anterior horn cells has been proposed as a pathological mechanism. However, MRI examination of the spinal cord has not revealed any abnormalities.⁴

Although EFR has largely been replaced by focused radiation technique, patients previously treated with high-dose radiation for Hodgkin's disease may experience this rare complication. Awareness of this entity among clinicians may help with prompt diagnosis and avoidance of extensive workup including invasive muscle biopsies.

Figure 1 Photograph of the left side of the neck showing wasting of the posterior cervical muscle groups. The left sternocleidomastoid muscle is contracted and atrophic. The skin overlying the sternocleidomastoid muscle showed thickening and telangiectasia (A). Axial view (B) of the CT of the lower neck showed wasting and fatty infiltration of posterior neck muscles (red arrow). There was also visible wasting asymmetry of sternocleidomastoid muscles (yellow arrow).

Learning points

- ▶ Dropped head syndrome (DHS) is a rare complication of extended field radiation.
- ▶ Cervical muscle atrophy and weakness are the aetiology of radiotherapy-induced DHS.
- ▶ Electromyography can show myopathic or neurogenic pattern, or a combination of both.

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ORCID iDs

Biplab Saha <http://orcid.org/0000-0002-3595-8804>

Woon Hean Chong <http://orcid.org/0000-0002-2070-3585>

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