Dropped-head Syndrome Due to High-dose Irradiation

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Dropped-head syndrome is a rare disorder characterized by weakness of neck extensor muscles causing an inability to extend the neck and resulting in a chin-on-chest deformity1. We observed 2 such cases.

Patient 1, a 55-year-old woman, had a history of Hodgkin’s lymphoma and high-dose mantle field irradiation 24 years prior to presentation. Over a period of 10 years she developed a severe weakness and atrophy of the neck extensor muscles (Figure 1).

Patient 2, a 58-year-old man, had a history of Hodgkin’s lymphoma and high-dose mantle field irradiation of his neck and abdomen 39 years prior to presentation. For 8 years he had an ongoing atrophy of his neck muscles and of his proximal and distal right leg. Examination showed kyphosis, chin-on-chest deformity (Figure 1), and severe atrophy of his trapezius and neck extensor muscles and of the extensor muscles of his leg. Aside from elevated creatine kinase (638 U/l) laboratory results were normal. Magnetic resonance imaging and biopsy of his neck region showed atrophy with no signs of myositis.

Dropped-head syndrome may be present in a variety of neurological disorders such as neuromuscular and motor neuron disease as well as (non-)inflammatory, dystrophic, or metabolic myopathies. It has been described after high-dose irradiation for Hodgkin’s lymphoma2. Pathophysiology is not completely understood. The lower motor neurons or the peripheral nerves have been suggested to be the primary location of injury.

Therapy is very difficult and physiotherapy and surgery have not been very successful in the past.

REFERENCES

Figure 1. Patients 1 and 2; Patient 2 attempting to extend the neck and showing a chin-on-chest deformity.