

CASE REPORT

A difficult case of inflammatory myositis

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Abstract

A 79-year-old lady presented with subclinical autoimmune hypothyroidism and proximal muscle weakness, which failed to respond to thyroxine. Subsequent electromyography and muscle biopsy confirmed polymyositis, which responded well to immunosuppressive therapy. She presented 4 months later with a thyroid carcinoma, most likely anaplastic, and died shortly afterwards. Although the association between malignancy and polymyositis remains controversial, to our knowledge this is the first report of thyroid carcinoma occurring in association with polymyositis and autoimmune hypothyroidism.

Keywords: *hypothyroidism, polymyositis, thyroid neoplasms*

Case report

A 79-year-old lady presented with a 5-week history of proximal muscle weakness. She had a history of essential hypertension, treated with atenolol and prazosin, and was an ex-smoker for the last 30 years. Systemic review was unremarkable. Clinically she was euthyroid with no goitre. There was non-tender proximal weakness in all four limbs, with atrophy in the thigh muscles. Examination was otherwise unremarkable. Initial investigations revealed thyroid stimulating hormone (TSH) 22.98 mU/l, thyroxine 127 nmol/l, positive thyroid microsomal antibody (1/1600), and creatine kinase (CK) 7839 U/l. Chest x-ray revealed mild cardiac enlargement. A provisional diagnosis of subclinical autoimmune hypothyroidism with hypothyroid myopathy was made, and thyroxine was commenced at 50 mcg daily.

After 2 months treatment with thyroxine she felt weaker. TSH was 15.39 mU/l, CK 5536 U/l, C-reactive protein (CRP) 29 mg/l, positive anti-Ro/SSA antibody, negative anti-Jo-1 antibody. Electromyography (EMG) revealed occasional small fasciculations and recruitment of small polyphasic motor units, suggestive of polymyositis (PM). Muscle biopsy demonstrated an inflammatory myositis, compatible with PM, with focal lymphohistiocytic inflammatory infiltrate and active fibre phagocytosis. There was variation in fibre size with degenerate and regenerating fibres visible, but no

evidence of fibre type atrophy, ragged red fibres or rimmed vacuoles. After 10 weeks treatment with thyroxine, she was commenced on azathioprine and prednisolone. Three weeks after commencing azathioprine and prednisolone, there was improvement in the proximal weakness with CK 55 U/l.

Four months after the original presentation, she presented acutely with a 2-week history of dysphagia, hoarseness and worsening stridor. Clinically there was a firm, immobile mass over the anterior neck, which the patient had noticed a few days previously. Direct laryngoscopy and computed tomography of the neck demonstrated a large infiltrating thyroid mass, with tracheal displacement and oesophageal compression. Although no tissue diagnosis was made, fine-needle aspiration revealed malignant looking cells and the mass was considered most likely to be an anaplastic thyroid carcinoma. She was referred for palliative radiotherapy, but died shortly afterwards.

Discussion

Neuromuscular disorders and elevation of serum CK are common in hypothyroidism [1]. Proximal myopathy occurs in approximately 38% of cases [2], can mimic PM [1, 3], and may rarely present as the sole manifestation of overt or subclinical hypothyroidism [4]. Appropriate

therapy with thyroxine to normalise thyroid biochemistry usually produces an improvement in the clinical symptoms and normalises serum CK, in an average of 3 months [5, 6]. However, clinical symptoms and elevated serum CK may rarely persist at 1 year [2, 6]. After 8 weeks of treatment with thyroxine, our patient was still biochemically hypothyroid, and felt weaker. CK and CRP remained persistently elevated. Despite a relatively short period of observation on thyroxine treatment, these features prompted us to review the diagnosis.

In the light of the EMG and muscle biopsy, the criteria for diagnosis of PM were met [7]. Although anti-Jo-1 is a myositis specific antibody, it is only detected in around 20–30% of patients with PM or dermatomyositis (DM), whereas anti-Ro/SSA, a myositis associated antibody, is found in 10–20% [7, 8]. PM/DM are associated with various other autoimmune disorders, but the occurrence of autoimmune hypothyroidism in patients with PM/DM is not well documented [9–12].

Any association between PM and malignancy remains controversial, although DM is generally accepted as having a higher risk of malignancy, compared to an age matched population, usually preceding, or within the first year after diagnosis [13, 14]. In female patients with PM, the relative risk of malignancy has been reported as 1.7 (95% CI, 1.0–2.5), compared to 3.4 (95% CI, 2.4–4.7) in female patients with DM [15]. Ovarian and pulmonary carcinomas are the most frequently associated malignancies, and to our knowledge, this is the first report of a thyroid carcinoma associated with PM and autoimmune hypothyroidism.

Key points

- Neuromuscular disorders are common in hypothyroidism.
- Proximal muscle weakness in hypothyroidism may mimic polymyositis, and warrants further evaluation with electromyography and muscle biopsy, particularly if there is a poor response to thyroxine therapy.
- Any association between polymyositis and malignancy remains controversial, although dermatomyositis is generally accepted as having a higher risk of malignancy, compared to an age matched population.

Acknowledgements

We are very grateful to Dr Richard Bonshek, lecturer and consultant in pathology, Academic Department of Ophthalmology, Central Manchester University Hospitals

NHS Trust, for his assistance regarding the muscle biopsy histopathology.

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Received 21 May 2002; accepted in revised form 29 October 2002