Dysphagia during glucocorticoid treatment of dermatomyositis: a differential diagnostic challenge

Key message

- Laryngopharyngeal glucocorticoid-induced myopathy cannot be overlooked when dysphagia presents during CS treatment of dermatomyositis.

Sr., a 66-year-old man had recently been diagnosed at the Rheumatology department of the University Hospitals Leuven with transcriptional intermediary factor 1-gamma (TIF-1γ) autoantibody-positive DM, evidenced by the pathognomonic tetrad of a heliotrope eruption, shawl sign, Gottron’s papules and proximal muscle weakness [1]. Screening for systemic complications revealed no cardiopulmonary or ocular involvement; however, an underlying superficial transitional cell carcinoma of the bladder was found [2]. Pulses of 500 mg methylprednisolone i.v. were administered for 3 days, followed by a maintenance dose of 64 mg methylprednisolone daily. There was a favourable effect on muscle and skin abnormalities, allowing for further outpatient management (Fig. 1).

Seven days later, however, the patient was re-admitted because of progressive new-onset dysphagia. Associated coughing, dysphonia and nasal fluid egress after swallowing suggested oropharyngeal dysfunction [3]. Initially, we considered muscle exhaustion as a consequence of a superimposed rhinopharyngitis. A flare-up of DM or glucocorticoid-induced myopathy were deemed less likely, because peripheral muscle strength had remained stable [2, 4]. Even so, dysphagia in DM can develop independently of peripheral muscle weakness [5]. As creatine kinase (CK) concentrations increased significantly in the following days and considering the association between TIF-1γ antibodies and dysphagia, the tentative diagnosis of a DM exacerbation was made [6]. Further laboratory testing, lumbar puncture (including paraneoplastic antibody assay) and brain imaging ruled out other neuromyogenic causes. Structural lesions were excluded, and severity was assessed by functional endoscopic evaluation of swallowing and a videofluoroscopic swallowing study, showing nearly absent swallowing response, massive pharyngeal residue and aspiration without cough reflex. Nasogastric feeding was initiated and repeated high-dose i.v. pulses of methylprednisolone were administered. Lack of clinical response prompted a trial of normal human immunoglobulins (30 g/day for 5 days) in association with a maintenance dose of 60 mg methylprednisolone daily; a treatment shown to be effective for corticoid-resistant DM by Wang et al. [7]. CK concentrations normalized, but dysphagia and dysphonia remained unaltered.

Despite its rarity, the possibility of isolated laryngopharyngeal steroid myopathy had to be reconsidered, and electromyography of the larynx was carried out. Vocals and cricothyroid muscles displayed short-duration, low-amplitude, polyturn motor unit action potentials with multiple serrations, resulting in reduced maximal contractility without spontaneous activity; findings most consistent with glucocorticoid-induced myopathy, although not excluding active myositis. The dose of methylprednisolone was reduced, after which the symptoms improved rapidly. Given the fact that normal human immunoglobulins were administered shortly before dose reduction of CSs, a flare-up of DM responding to immunoglobulin therapy cannot be excluded completely. Nevertheless, the clinical course along with electromyographic findings are strongly suggestive of a final diagnosis of steroid myopathy.

Statistically, dysphagia occurs in 12–54% of DM cases, whereas it is a very rare presentation of glucocorticoid-induced myopathy [8]. We found only two case reports by Izumi et al. [9] of pharyngeal steroid myopathy complicating treatment of PM. Moreover, progressive peripheral muscle weakness accompanied dysphagia in these cases, emphasizing the uniqueness of isolated laryngopharyngeal steroid myopathy.

When differentiating a flare-up of inflammatory muscle disease from glucocorticoid-induced myopathy, clinical clues to the latter are concomitant Cushingoind features and progressive symptoms despite declining CK levels [4, 9]. It occurs typically after 1 month of prednisone or equivalent drug use in doses >10 mg/day, although high-dose regimens can produce symptoms within 2 weeks. Laboratory tests are usually unremarkable, except in the acute phase when muscle enzyme levels may be fairly high [4]. Electromyography yields normal or myopathic signals without spontaneous activity in glucocorticoid-induced myopathy, whereas a myopathic pattern with spontaneous activity is almost diagnostic for active myositis [4, 10]. However, these typical findings are absent in ~20% of DM cases; a number rising to 40% in patients using CSs [10]. Muscle biopsy can be undertaken if doubt remains. Steroid myopathy is marked by atrophy of type IIb muscle fibres without an inflammatory infiltrate. Discontinuation or reduction of the dose of glucocorticoids is the mainstay of
The degree of muscle impairment is based on clinical estimation. CK: creatine kinase; IVIG: intravenous immunoglobulin.

Funding: There is no funding to be declared.

Disclosure statement: This manuscript has not been submitted or published elsewhere. The authors declare no conflicts of interest.

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Accepted 17 March 2018

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