Oropharyngeal dysphagia as dominant and life-threatening symptom in dermatomyositis

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Abstract

Background. Dysphagia can be a serious problem in patients with inflammatory myopathies. It may be associated with nutritional deficit, aspiration pneumonia, and poor prognosis. Case report. We presented a 60-year-old male, suffering from difficulty in swallowing, pain and weakness in the proximal parts of his extremities, and skin manifestation. Laboratory findings showed increased creatine kinase and aldolase. Antinuclear antibodies to HEP-2 substrate revealed titer of 1:40. Electromyoneurography demonstrated evidence of a proximal myopathy. A muscle biopsy revealed myositis. The barium swallow test was remarkable for regurgitation, and nasal emerging of barium. Nuclear magnetic resonance images of cranium was normal. Tumor markers CEA, and Ca 19-9 were increased. A dose of 1 mg daily prednisolone was administered and percutaneous enteral feeding was performed. Two months later, the patient developed febrile state, aspiration pneumonia, and died due to respiratory failure. Conclusion. In cases of dermatomyositis with the serious dysphagia, percutaneous endoscopic gastrostomy should be performed as soon as possible. Overall survival rate is low, even with an adequate therapy administration. Inflammatory myopathies should be considered in any patient with oropharyngeal dysphagia.

Key words: deglutition disorders; diagnosis; myositis; dermatomyositis; gastrostomy; enteral nutrition; treatment outcome.

Background

Dysphagia can be present as a serious problem at any time during inflammatory myopathies. It is commonly observed in the acute inflammatory phase of these conditions, and may be associated with nutritional deficit, aspiration pneumonia, decreased quality of life, and poor prognosis. In cases of dysphagia grade 4, rehabilitation procedures, and interventional measures (cricopharyngeal or esophageal dilation, cricopharyngeal myotomy, botulinum injections of the upper esophageal sphincter) do not give desirable effects. In such cases non-oral feeding is needed. Swallowing disorders are considered a major cause of both morbidity and mortality in polymyositis (PM) and dermatomyositis (DM) and may lead to life threatening complications (cachexia related to severe swallowing disorders, and recurrent aspiration infectious
pneumonia)\(^6,7\). Dysphagia in PM/DM has not been evaluated systematically, especially for the striated muscle dysfunction. Indeed, the subject has been focused on problems in the esophagus and scant attention has been paid on the oropharynx which may be equally affected in PM/DM\(^8,10\).

**Case report**

A 60-year-old man had had difficulty in swallowing a month before his admission to our hospital. In that time he also had pain and weakness in the proximal parts of his extremities. Ten years ago he was treated for alcohol abuse, but otherwise was healthy. Skin manifestations demonstrated erythematous maculopapular eruption in his cheeks and forehead, mild periorbital edema and scarce Gottron's papules overlying dorsal metacarpophalangeal surfaces of his hands (Figures 1 and 2).

![Fig. 1 – Erythematous maculopapular eruption on the cheeks and forehead](image1)

![Fig. 2 – Skin manifestations (Gottron’s papules) seen in the patient with dermatomyositis](image2)

Muscle strength on the Medical Research Council scale was grade 4 in the proximal arms and neck flexors, and 3 in the legs. We did not notice swelling and painful movement of his oral floor. The strength of the tongue was intact. Laboratory findings showed increased serum muscle enzymes. Serum creatine kinase (CK) was 1 526 IU/L, with the fraction CK reflecting myocardial injury (MB) between 36 - 40 ng/mL, MB mass fraction 5.1 µg/L, and aldolase 16.2 IU/L. There were also pathological levels of liver enzymes – lactate dehydrogenase (LDH) 1047 U/L, aspartat aminotransferase (AST) 18 U/L, alanin aminotransferase (ALT) 72 U/L, and gamma glutamine transpherasis (gamma GT) 67 U/L. Sedimentation rate (SE) was 90. Autoantibody screen was positive for extractable nuclear antigens (ANA HEP 2) with a value of 1:40, and negative for antinuclear antibody (ANA), C-reactive protein, and anti-Jo IgG. Electromyography demonstrated mild proximal myopathy in all extremities. A muscle biopsy was obtained from the right deltoid muscle, because electromyographic examination revealed mild neuromyopathy in this area. Biopsy findings presented the infiltration of lymphocytes and plasma cells mainly in perivascular areas. (Figure 3). A diagnosis of dermatomyositis was made. Severe dysphagia-related symptoms progressed over several days (“food sticking in the throat”, “deglutitive coughing”, “choking”, and “postnasal regurgitation”). Chest X-ray was normal. Abdominal ultrasonography showed no pathological findings. A barium sulphate swallow test (a contrast swallow X-ray film) was remarkable for regurgitation, aspiration, and no absolute emptying in the region of pharynx. There was also evidence to nasal emerging of barium (Figure 4). Nuclear magnetic resonance (MRI) images

![Fig. 3 – Myositis (hematoxylin-eosin staining × 200)](image3)

![Fig. 4 – A barium sulphate swallow X-ray test – no absolute emptying in the region of pharynx](image4)
of cranium revealed bilateral cortical and subcortical reduction of both cerebrum and cerebellum (Figure 5).

Fig. 5 – Nuclear magnetic resonance images of cranium: cortical and subcortical reduction

No expansive processes were found. We performed tests to exclude an underlying malignancy. Carcinoembryonic antigen (CEA) was 8.7 ng/ml and carbohydrate antigen 19–9 (Ca 19–9) was 73,3 U/mL. Esophagogastroscope was normal and histopathological findings from the biopsies performed from several parts of gastroduodenum showed only multiphocal atrophic pangastritis (MAG – Houston) grade 2, stage 2, as well as chronic duodenitis. Colonoscopy and prostate examination were not performed because of dramatic clinical deterioration. The patient was treated promptly with prednisolone 1 mg/kg daily. Although muscle pain, muscle enzyme values, and skin manifestations partially improved, dysphagia and dysphonia progressed over several days until he was unable to swallow liquids. A necessity for introduction of non-oral feeding was obvious. Seven days after his first visit, enteral feeding was performed introducing percutaneous endoscopic gastrostomy (PEG) (Figure 6). Unfortunately, the patient developed febrile state with aspiration pneumonia, and died two months afterwards due to respiratory failure.

Discussion

Polymyositis and DM are the two major idiopathic inflammatory myopathies, and usually affect the skeletal musculature of the body 11. Skin lesions may precede the development of the myopathy and may persist after the control of myositis. Proximal muscle disease is usually symmetrical, and symptoms are fatigue, weakness and sometimes myalgia 6. Proximal dysphagia reflect an involvement of striated muscle of the pharynx or proximal esophagus. The symptom is frequent and may occur in 60 - 73% of patient with the inclusion of body myositis 2,3 and 12 - 54% of patient with PM/DM 1,5. The overall frequency of esophageal involvement in PM/DM has been reported to be 25 - 60 %, and it is considered a major cause of both morbidity and mortality 5,12. Both striated and smooth muscles of the oropharynx and esophagus could be affected. These events lead to delayed emptying and dysmotility of these structures, cachexia, and recurrent aspiration infectious pneumonia 1,6,7. In a retrospective 5-year study of dysphagia and inflammatory myopathy of elderly, Terry et al. 2 found that the highest 1-year mortality was in those with DM (31%). Among them the highest mortality was in patients who required PEG (64%). However, dysphagia in PM/DM has not been evaluated systematically especially for the striated muscle dysfunction 6,9. Most of the available studies are less detailed and based in personal experiences or reviews of records 12–14. Furthermore, authors are usually focused on problems in the esophagus and scant attention has been paid on the oropharynx which may be equally affected in PM and DM 8–10. In spite of the prevalence of dysphagia in inflammatory myosi-
tis reported among 29 - 44% of children, symptom can be overlooked until it becomes severe 15. X-ray videofluoroscopic swallowing study is the procedure of choice in children to delineate pharyngeal and/or oesophageal phases of swallow. In juvenile DM even minimal swallow abnormalities recognition is crucial to avert aspiration and lung damage, and also preventable by nasogastric or parenteral feeding 16. In latest investigations of Otao at al. 17 prompt and non-invasive recognition of inflammation in the muscles of oral floor was done using T2-weighted fat-saturated horizontal and coronal images of MRI. This is the first report of oral muscle inflammation in DM confirmed by MRI though there are similar cases that have been reported without MRI findings 18.

**Conclusion**

In cases of dermatomyositis with severe dysphagia PEG should be performed in surgical ward as soon as possible. It is very important to detect the first minimal signs of the swallowing dysfunction. According to newest approaches to this serious problem, this should be done with fast and harmless methods (like MRI). In such cases it is possible to prevent the lung damage, as well as damage of other organs and systems. Mortality is high, and overall survival rate is low, even if an adequate therapy is administrated. On the other hand, inflammatory myopathy should be considered in any patient with unexplained oropharyngeal dysphagia.

**REFERENCES**


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