Clinical Case Report

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A case report of life-threatening acute dysphagia in dermatomyositis

Challenges in diagnosis and treatment

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Abstract

Rationale: Although dysphagia is a known complication of dermatomyositis, sudden onset of dysphagia without the notable aggravation of other symptoms can make the diagnosis and treatment challenging.

Patient concerns: A 53-year-old male diagnosed as dermatomyositis 1 month ago came to our emergency department complaining of a sudden inability to swallow solid foods and liquids. The patient showed generalized edema, but the muscle power was not different compared with 1 month ago.

Diagnoses: Serum creatine kinase level was lower than that measured 2 weeks ago. Computed tomography scan of the larynx, chest, abdomen, and pelvis, an esophagogastroduodenoscopy, and brain magnetic resonance imaging were unremarkable. A videofluoroscopic swallowing study revealed inadequate pharyngeal contraction and slightly decreased upper esophageal sphincter opening with silent aspiration.

Intervention: Treatment with oral prednisolone, intravenous methylprednisolone, azathioprine, and intravenous immunoglobulins was applied. During the course of medical treatment for life-threatening dysphagia, he continued with rehabilitative therapy.

Outcomes: He could swallow saliva at 2 months and showed normal swallowing function at 3 months from the onset of dysphagia. Dysphagia has not recurred for 3 years after recovery.

Lessons: A multidisciplinary approach is necessary to diagnose severe acute dysphagia due to exacerbation of underlying dermatomyositis rather than other structural or neurological causes. Appropriate supportive care is important because dysphagia can be life-threatening and last for a long time.

Abbreviations: CK = creatine kinase, IVIG = intravenous immunoglobulins, MRC = Medical Research Council, UES = upper esophageal sphincter, VFSS = videofluoroscopic swallowing study.

Keywords: conservative treatment, deglutition disorder, dermatomyositis, diagnosis

1. Introduction

Dermatomyositis is a systemic inflammatory disorder affecting diverse organs including skeletal muscle.[1,2] Dysphagia is also a known complication during the course of the disease and has been reported in 18% to 20% of patients.[1,3–5] As the pharynx and upper esophageal sphincter (UES) consist of skeletal muscle, reduced pharyngeal contraction and dysfunction of the UES are 2 principal swallowing problems observed in these patients.[1,3,5] The primary treatment for dermatomyositis is corticosteroids. Other immunosuppressants and immunomodulatory therapies have been used for steroid-refractory cases.[2] Together with medical treatment, swallowing rehabilitation therapy is common.[3]

We report a case of a 53-year-old man who visited an emergency room due to sudden-onset dysphagia. The unusual acute severe dysphagia in the patient diagnosed as dermatomyositis 1 month ago posed challenges to diagnosis and treatment. The case demonstrates that life-threatening acute pharyngeal dysphagia can develop by the exacerbation of the underlying myopathy. After the diagnosis, aggressive drug therapy and proper supportive care are indicated. This case report was reviewed and approved by the Institutional Review Board of Uijeongbu St. Mary's Hospital, College of Medicine, The Catholic University of Korea (Ethical approval number: UC16ZISE0097). This report was exempted from patient informed consent by the Institutional Review Board of our hospital.

2. Case report

A 53-year-old Asian man complained of myalgia, general weakness, and weight gain over 2 months. He showed proximal muscle weakness of Medical Research Council (MRC) grade 3 in upper and lower extremities.[6] Skin rash was apparent on the cheeks and neck. Serum creatine kinase (CK) levels were

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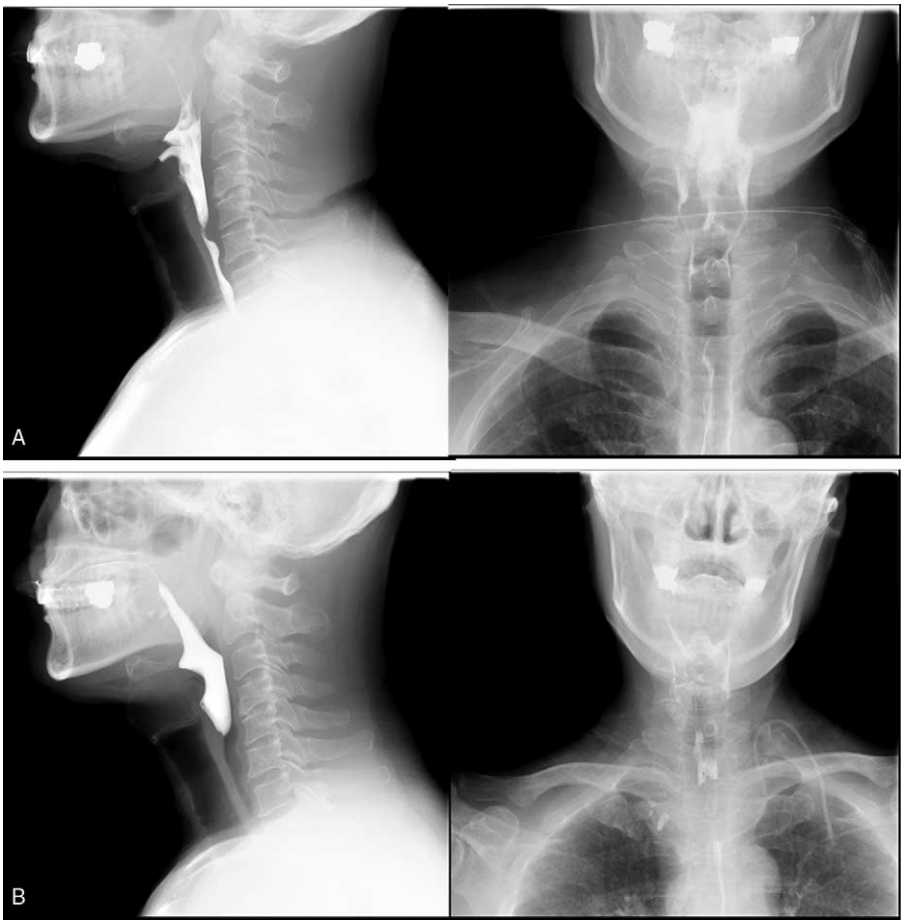

Figure 1. Videofluoroscopic swallowing study data. (A) Initial videofluoroscopic swallowing study. (Left) Lateral images showed inadequate hyolaryngeal movement, incomplete laryngeal closure, and laryngotracheal penetration and aspiration. (Right) Anterior-posterior images showed large amounts of residue in the valleculae and pyriform sinus fossa. (B) Videofluoroscopic swallowing study after 2 months from the onset of dysphagia. (Left) Lateral images showed improved hyolaryngeal movement and laryngeal closure. (Right) Anterior-posterior images showed decreased pharyngeal residue.

increased to 981 U/L (normal range: 43–244 U/L). Antinuclear antibodies were positive (1:400, speckled staining pattern) and anti-Jo antibodies were negative. Electromyography showed typical myopathic findings. A muscle biopsy showed focal perifascicular atrophy and perivascular lymphocytic infiltration in the perimysium and endomysium. These clinical findings and the results of laboratory tests supported a diagnosis of dermatomyositis. Treatment with low-dose prednisolone (5 mg/day) was begun due to a necrotic lesion of the penile skin. After 2 weeks, CK levels had declined to 605 U/L.

One month after starting treatment, he visited the emergency department complaining of a sudden inability to swallow solid foods, liquids, and even saliva with generalized edema. Serum CK levels were 574 U/L and the muscle power was unchanged with MRC grade 3. He was admitted to the rheumatology department. He showed poor laryngeal elevation on palpation and decreased gag reflex on both sides of the pharynx. Computed tomography scan of the neck, chest, abdomen, and pelvis and esophagogastroduodenoscopy were unremarkable, reducing the possibility of dysphagia due to an obstructing lesion, such as an abscess or a malignancy. Brain magnetic resonance imaging was performed to rule out stroke for acute dysphagia with decreased laryngeal elevation and the gag reflex. The imaging showed no definite abnormality in the brain. A nasogastric tube was inserted, allowing nutritional support and oral drug delivery. A videofluoroscopic swallowing study (VFSS) revealed inadequate hyolaryngeal movements, delayed triggering of swallow reflex, and slightly limited UES opening (Fig. 1A). Aspiration was noted for both puree and liquid, with large amounts of residue in the valleculae and pyriform sinus fossa.

Treatment with low-dose prednisolone was escalated to intravenous methylprednisolone, 1 mg/kg/day. The dose was increased to 2 mg/kg/day and azathioprine 50 mg/day was added. Dysphagia worsened and he complained of hypernasality during speaking after 2 weeks. Methylprednisolone and azathioprine were maintained and additional therapy with intravenous immunoglobulins (IVIGs) 400 mg/kg/day for 5 consecutive days was carried out. No rapid improvement in the dysphagia occurred. Two weeks later, *Pneumocystis jiroveci* pneumonia with leukopenia developed and he was treated with antifungals (fluconazole 200 mg/day for 24 days), antibiotics (intravenous cefazolin 3 g/day for 9 days), oral prednisolone 10 mg/day for 4 days, and intravenous hydrocortisone 75 mg/day for 6 days. With the improvement in the infection, at 2 months from the onset of dysphagia, he could swallow saliva again. A follow-up VFSS demonstrated improved pharyngeal contraction and decreased pharyngeal residues (Fig. 1B). Aspiration was not detected with puree, but he showed silent aspiration with liquid. Therefore, we

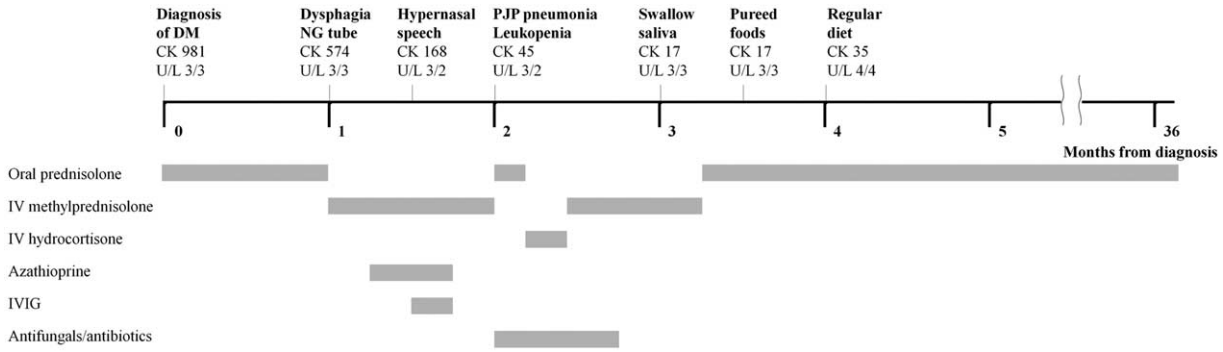

Figure 2. The clinical course of the patient. CK = serum creatine kinase level, DM = dermatomyositis, IV = intravenous, IVIG = intravenous immunoglobulins, NG = nasogastric, PJP = *Pneumocystis jiroveci*, U/L = Medical Research Council grades in upper and lower extremities.

started oral feeding with a level 2 dysphagia diet (pureed foods, with honey-like fluid).[5] He was discharged after 2 months of hospitalization with oral prednisolone 60 mg/day. VFSS performed at 3 months after the onset of dysphagia showed normal swallowing function. He started a regular diet without difficulty. During the longstanding course of dysphagia, he continued with rehabilitative dysphagia therapy, including compensatory maneuver training, oropharyngeal exercise, and neuromuscular electrical stimulation (VitalStim; Chattanooga Group, Hixson, TN). His normal swallowing function has been maintained for 3 years and he has continued on low-dose prednisolone, while the dose tapered down gradually to 2.5 mg/day. Figure 2 summarizes the clinical course of the patient.

3. Discussion

Dysphagia in dermatomyositis patients usually gradually progresses over several days to months.[3,7,8] To our knowledge, patients with dermatomyositis who visit the emergency room with sudden-onset dysphagia without the notable aggravation of other symptoms have rarely been reported. In addition, severe dysphagia with liquids, solids, and even saliva (aphagia) is also uncommon.[1,7–11] Our patient could not swallow saliva abruptly, without the evidence of disease aggravation such as progression of limb weakness and elevated CK levels. This unusual presentation made the diagnosis and treatment challenging. A multidisciplinary team with a rheumatologist, otolaryngologist, gastroenterologist, neurologist, and rehabilitation doctor was necessary for the diagnostic process to rule out the potential acute structural obstruction and/or neurological conditions. After the exclusion of head and neck cancer, laryngeal edema, infection, gastrointestinal cancer, and acute stroke, the diagnosis was acute severe dysphagia due to exacerbation of dermatomyositis.

Dysphagia in dermatomyositis is much more common in steroid-resistant patients.[8,9,11] For steroid-resistant dysphagia, high-dose IVIG and cyclophosphamide have been effective in a few cases.[7–9,11] We also used multiple drugs, including intravenous methylprednisolone, azathioprine, and IVIG due to the life-threatening condition of the patient, including saliva aspiration, malnutrition, and leukopenia. However, unlike many previous reports, there was no immediate improvement of dysphagia in our patient. Therefore, it is difficult to discuss the therapeutic effect of a certain drug in this case. However, our case demonstrates that complete improvement of life-threatening steroid-resistant dysphagia can occur if aggressive medical and rehabilitative therapy is maintained. Among the cases with severe dysphagia, only 1 patient showed delayed recovery at 3 months after use of intravenous methylprednisolone, IVIG, and methotrexate.[8] The clinical course of this patient was quite similar to our patient, but she showed severe dysphagia for more than 2 months before worsening, which made the diagnosis simpler than in our case. In cases of severe dysphagia in patients with dermatomyositis, aggressive therapy seems to be needed.

Our serial VFSS provided detailed information on the clinical course of severe pharyngeal dysphagia. Dysphagia in dermatomyositis has previously been attributed only to esophageal involvement.[1,3] A recent study using both VFSS and manometry suggested that dysphagia in patients with inflammatory myopathy appears to be due more to impaired pharyngeal muscle contraction, stemming from weakness in the suprahyoid muscles than the commonly held belief of failed UES relaxation.[4] For pharyngeal, rather than esophageal, motility dysfunction, the swallowing rehabilitation therapies, such as compensation techniques and oropharyngeal exercises, would be more applicable.[5] Although our patient stated that his swallowing had completely recovered at 2 months from the onset of dysphagia, VFSS performed at that time revealed silent aspiration with liquid.[5] Therefore, the subjective complaint of the patient may not be a reliable indicator of the presence of swallowing problems in myopathy patients.

4. Conclusion

We report a rare case of sudden-onset severe pharyngeal dysphagia due to exacerbation of underlying dermatomyositis without the aggravation of other symptoms. A multidisciplinary approach is necessary to exclude potential structural and neurological causes of acute dysphagia. It is imperative to maintain appropriate medical, dietary, and rehabilitative support because the control of the underlying disease can be delayed up to several months.

Author contributions

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Supervision: Jung Soo Lee, Yeo Hyung Kim.
Validation: Kyoung Min Kwon, Yeo Hyung Kim.
Writing – original draft: Kyoung Min Kwon, Yeo Hyung Kim.
Writing – review & editing: Jung Soo Lee, Yeo Hyung Kim.

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