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Oropharynx Dysphagia In Dermatomyositis Patients

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ABSTRACT

Introduction: Oropharyngeal dysphagia is one of the clinical manifestations of dermatomyositis, which can impact the patient's health and quality of life. Dermatomyositis is characterized by progressive muscle weakness and characteristic skin lesions. Oropharyngeal dysphagia is a significant complication, occurring in 10-73% of dermatomyositis patients. This case report describes the treatment of oropharyngeal dysphagia in a dermatomyositis patient with cyclophosphamide. Case report: A 38-year-old man with the main complaint of being unable to swallow saliva and solid food for 3 weeks, accompanied by weakness in the extremities and a reddish rash around the eyes. Laboratory examination showed an increase in serum CK. EMG showed demyelinating motor polyneuropathy. Skin biopsy was consistent with dermatomyositis. Initial FEES showed severe oropharyngeal dysphagia with penetrating aspiration. The patient was given high-dose methylprednisolone without significant improvement, followed by cyclophosphamide 1 gram for 7 cycles. Methods: A literature search was conducted in PubMed with the keywords “dysphagia” OR “swallow” AND “dermatomyositis” OR “polymyopathy” AND “cyclophosphamide”. Results: Cyclophosphamide administration showed clinical improvement after the second cycle. FEES evaluation after the sixth cycle showed improvement to mild oropharyngeal dysphagia. Two months post-therapy, patients reported significant improvements in swallowing and muscle strength. Conclusion: This case illustrates the manifestation of oropharyngeal dysphagia in dermatomyositis and significant improvement after administration of cyclophosphamide. FEES evaluation plays an important role in assessing the degree of dysphagia and response to therapy. Cyclophosphamide may be an option in cases of dermatomyositis with steroid-resistant dysphagia.

Keyword*:* *Disfagia orofaring, Dermatomiositis, Siklofosfamid, FEES*

1. INTRODUCTION

Dysphagia comes from the Greek word which means eating disorder. Oropharyngeal dysphagia is a swallowing disorder that occurs in the oral and pharyngeal phases. This condition can cause serious complications such as malnutrition, dehydration and aspiration pneumonia.[1] Neurological conditions that can cause oropharyngeal dysphagia include dermatomyositis, stroke, Parkinson's, poliomyelitis, myasthenia gravis, head malignancies and neck.[2] Dermatomyositis is part of a group of idiopathic inflammatory myopathies characterized by progressive proximal muscle weakness and characteristic skin lesions.[3] This disease can affect various organ systems, including the digestive system, where oropharyngeal dysphagia is one of the significant clinical manifestations.[4] Dermatomyositis is found in adult patients with an age range of 40-60 years, and are more common in women than men with a ratio of two to one.[5]

The prevalence of oropharyngeal dysphagia in dermatomyositis patients is quite significant. Epidemiological studies show that around 10-73% of dermatomyositis patients experience dysphagia.[6] The prevalence in Jakarta shows that of the 47 dermatomyositis patients studied, 18 patients (38.3%) experienced dysphagia.[7]

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dermatomyositis patients studied, 18 patients (38.3%) experienced dysphagia.[7] Oropharyngeal dysphagia in dermatomyositis patients occurs due to weakness and dysfunction of the muscles involved in the swallowing process. Chronic inflammation of the muscles of the oropharynx, including the muscles of the tongue, soft palate, pharynx, and upper esophagus, leads to impaired coordination and strength necessary for effective swallowing.[8]

Treatment of oropharyngeal dysphagia in dermatomyositis patients requires a multidisciplinary approach that includes pharmacological and non-pharmacological therapy. The first line of pharmacological therapy is glucocorticoids (corticosteroids).[9] Patients who do not respond to corticosteroids can administered immunosuppressants (methotrexate, azathioprine, or mycophenolate mofetil, cyclophosphamide), intravenous immunoglobulin and biologic agents (rituximab). [10] A cohort study of 123 dermatomyositis patients has demonstrated a significant improvement in disease activity with administration of cyclophosphamide. [9]

The purpose of writing a case report is to report one case of oropharyngeal dysphagia in dermatomyositis.

2. CASE REPORT

A 38-year-old man referred from Soewandi Hospital Surabaya came to the Emergency Room of the Regional Dr. Soetomo General Academic Hospital Surabaya with the main complaint of not being able to swallow saliva since 3 weeks ago and unable to eat solid food, requiring effort to swallow, feeling of a lump in the throat and often choking when drinking since 2 weeks ago, another complaint is weakness in all four extremities since three months ago. Complaints begin with weakness in both legs, followed by weakness in both arms, worsening over time so that the patient cannot walk.

Five months before entering the hospital, the patient complained of a purplish red rash around the eyes which felt darker, a reddish rash was also found on the patient's chest and the appearance of the rash was not influenced by sun exposure. The patient complained of intermittent fever. The patient has complained of difficulty swallowing and slurred speech in the past month for the last 2 weeks. The patient has no previous history of hypertension, diabetes, stroke, trauma or autoimmune disease. The patient works as a driver.

Physical examination revealed a weak general condition, compos mentis consciousness, weight 55 kg, height 165 cm with vital signs within normal limits. Lung and heart examinations were within normal limits. Examination of the limbs revealed weakness in all four extremities with muscle strength in the upper extremities 3/3 and lower extremities 2/2 without sensory abnormalities. Local ORL-HNS status showed that the ears, nose, throat and neck were within normal limits, on the face there were purplish red lesions around both eyes. The patient was diagnosed with oropharyngeal dysphagia, the patient had a Naso Gastric Tube installed for nutrition with the suggestion of a Flexible Endoscopic Evaluation of Swallowing (FEES) examination at the ORL-HNS Outpatient Unit.

The patient was treated together with a neurology colleague who diagnosed suspicion of dermatomyositis with a differential diagnosis of Chronic inflammatory demyelinating polyradiculoneuropathy (CIPD) and the patient underwent laboratory examinations (serum C3, C4, and Ck), ANA test (Anti Nuclear Antibody), Electromyography (EMG) and FEES. ANA test was negative, EMG examination showed demyelinating motor polyneuropathy with suspicion of muscle disease. The patient received methylprednisolone 1 gram every 24 hours intravenously for three days, there was no improvement, followed by methylprednisolone 62.5 mg every 24 hours intravenously for 3 days. The clinical condition did not improve after six days of treatment. The patient was given 1 gram of cyclophosphamide immunosuppressant, but the patient refused and was forced to go home and was given 16-16-0 mg methylprednisolone tablets.

One day after leaving the hospital, the patient returned to the emergency room at Dr. Soetomo General Academic Hospital because the NGT had fallen off. It had been reinstalled, and a laboratory examination was carried out. Laboratory results showed leukocytosis (16,170/mm3) so the patient was treated again and given an Asering infusion: D5%: Kalbamine 1:1:1 in 24 hours, Cyclophosphamide 1gr in 4 hours intravenously for 1 day, Ceftriaxone 1gr every 12 hours intravenously and vitamin B Complex 1 tablet every 24 hours.

The patient underwent a skin biopsy from a skin lesion on the chest by a Skin and Venereology colleague. The biopsy results showed vacuolar degeneration in some of the basal cells. The dermis layer showed a mild infiltration of lymphocytes at the dermo-epidermal junction and melanophage distribution, with conclusions consistent with systemic lupus erythematosus. The patient was treated for 6 days and went home with oral medication Methylprednisolone 16-16-0 mg, and oral hydroxychloroquine 200 mg every 8 hours. The patient was planned to undergo six cycles of Cyclophosphamide therapy and was scheduled for a Magnetic Resonance Imaging (MRI) examination of the neck and a Flexible Endoscopic Evaluation of Swallowing (FEES) examination. The MRI results showed no lesions or masses in the nasopharynx, oropharynx, or hypopharynx, and no lesions or changes in intensity were seen in the neck muscles.

The patient came to ORL-HNS after discharge for a FEES examination. The examination results showed pre-swallowing, poor oral hygiene, weak lip strength, and swollen cheeks leaking on the right and left. The results of the Fiber Optic Laryngoscope (FOL) examination showed weak velopharyngeal movement symmetrically on both sides, heavy standing secretion, right and left leakage, symmetrical vocal cord movement, no hypopharyngeal sensitivity. In the swallowing assessment, the first food was given, namely strained porridge; heavy residue was found in the valleculae and piriform sinuses; the patient was asked to cough, but the patient could not cough, and the FEES examination was stopped. The

results of the examination concluded that the patient had severe oropharyngeal dysphagia with heavy aspiration penetration; NGT diet advice, medical rehabilitation consultation for bio feeding and cough therapy, and internal medicine therapy were continued.

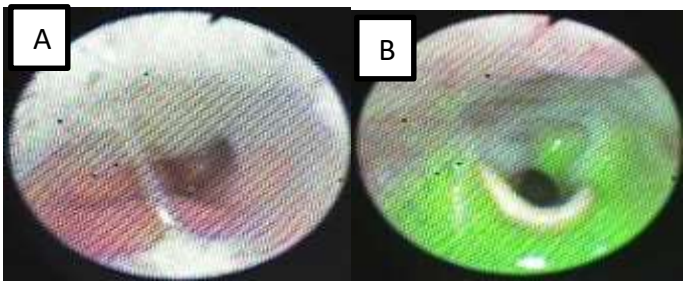
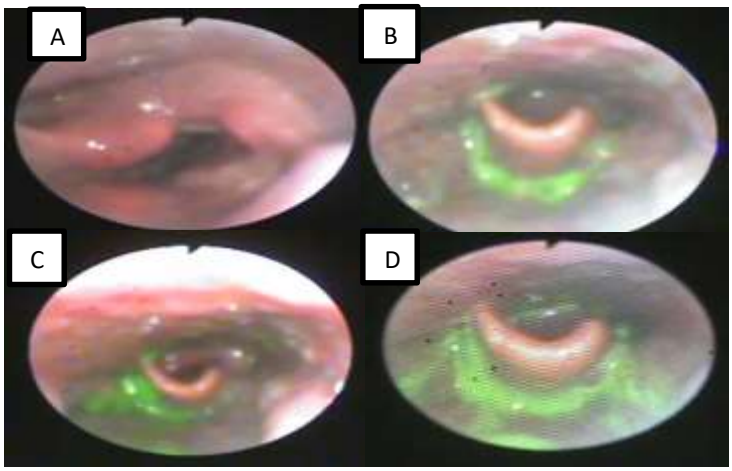


Figure 1. A. Heavy standing secretion in the piriform sinus and vallecula.

B. Bolus residue fills the vallecula and priform sinus

The patient continued cyclophosphamide therapy. The patient admitted that the complaint had improved since the second cycle of cyclophosphamide so that the patient could eat fine porridge slowly, but the weakness in the limbs was still there. In the fifth cycle of cyclophosphamide, the patient could walk without assistance and eat soft rice slowly without using an NGT, which internal medicine colleagues had removed since the fourth cycle. After cyclophosphamide therapy for 6 cycles, treatment was continued with oral Imuran 50 mg every 12 hours, methylprednisolone 8 mg every 24 hours, and the patient was advised to evaluate FEES.

The patient was taken to ORL-HNS Outpatient for a FEES evaluation, the results of the pre-swallowing examination were obtained, and oral hygiene was adequate. The results of the FOL examination of the right and left arytenoids are udim, the vocal cords are hyperemic, the epiglottis is edematous, right-sided leakage, the movement of the velopharynx is symmetrical and equally strong, the movement of the vocal cords is normal, light standing secretion in the vallecula/pyriform sinus, the muscle tone of the lateral walls of the pharynx is symmetrical, hypopharyngeal sensitivity is present. The results of the swallowing assessment showed that patients were given 6 types of food, with minimum residue after five swallows (Figure 3). The results of the FEES examination can be concluded that the patient has improved with the conclusion that the patient is experiencing mild oropharyngeal dysphagia



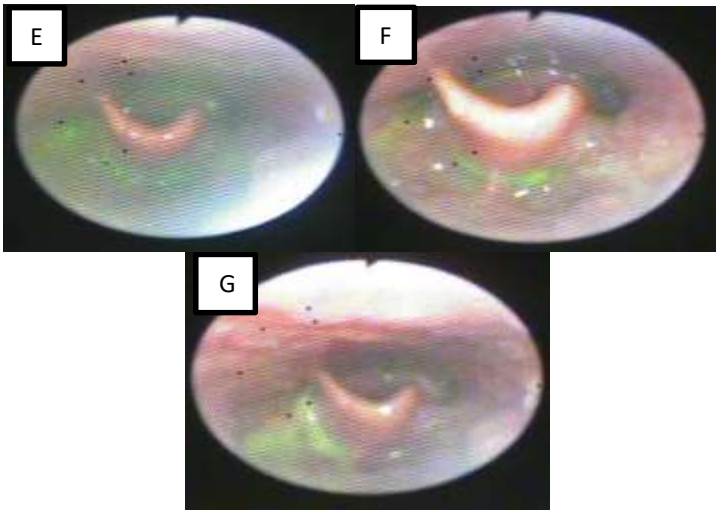


Figure 3. A. Right and left arytenoid edema, hyperemic vocal cords, epiglottis edema.

B. Residue in strained porridge, C. Rice porridge residue, D. Flour slurry residue

E. Liquid milk residue, F. Water residue colored with green dye, G. Biscuit residue.

Clinical Question

How does cyclophosphamide respond to dermatomyositis patients with oropharyngeal dysphagia?

Methods

Literature search was conducted in November 2022 in PubMed with the keywords "dysphagia" OR "swallow" AND "dermatomyositis" OR "polymyopathy" AND "cyclophosphamide" OR "cytophosphane". There were 4 case reports that were relevant to the topic and available in full text, from a total of 66 literature search results.

3. RESULT

Management of dermatomyositis cases differs in each case characteristic. Age, complications of dermatomyositis and patient condition affect the differences in treatment. In general, management of dermatomyositis is the administration of high-dose steroids and adjuvant therapy. Adjuvant therapy can be in the form of cyclophosphamide.

Table. 1 Literature Review

	Problem	*Intervention*	*Control*	*Outcomes*
Elmdaah A et al., 2021	Patients with dermatomyositis who experience acute	Combination therapy intravenous methylprednisolone, oral prednisolone, and cyclophosphamide	-	Improvement of dysphagia symptoms Increased muscle strength Decreased inflammatory markers (CRP, ESR) and muscle enzymes (CK LDH)
Senju A et al, 2011	A boy with dermatomyositis who presented with acute dysphagia and oropharyngeal ulcer	cyclophosphamide	-	Improvement muscle strength and ulcers improved
Deaken CT et al, 2018	Patients with Severe juvenile	Treatment with cyclophosphamide	Comparing patients who received cyclophosphamide	□ Effectiveness of treatment (improvement of

	dermatomyositis (JDM)		with those who did not receive cyclophosphamide	symptoms and clinical signs) □ Safety of treatment (side effects)
Ali et al, 2021	Description of atypical dysphagia Complaints in a Patient with dermatomyositis	Administration of high-dose prednisolone 1 mg/kg (60 mg once daily orally for four weeks) and cyclophosphamide cycles 500 mg	-	Dermatomyositis patients with dysphagia can be treated with combination therapy.

4. DISCUSSIONS

The patient in this case was a 38-year-old man. Based on prevalence data in several studies, it was found that the most dysphagia sufferers were in the 45-54 year age range (27%), followed by the 55-64 year age range (25%) and the 35-44 year age range (19%). [11] Dermatomyositis has a bimodal age distribution, with the first peak in children aged 5-15 years and the second peak in adults aged 45-64 years. [12]

The main complaint felt is unable to swallow saliva and unable to eat solid food, requires effort to swallow, a feeling of obstruction in the throat and often choking when drinking. Symptoms in patients are in accordance with oropharyngeal dysphagia because complaints appear within a few seconds after swallowing, such as a feeling of obstruction in the throat, frequent dripping saliva, coughing, nasal regurgitation, aspiration or choking. [11] Patients with dermatomyositis with dysphagia are caused by the oropharyngeal swallowing phase involving the tongue, epiglottis, palatine tonsils, hard palate and soft palate which experience tongue weakness, poor palatal movement, and accumulation of secretions in the hypopharynx. Studies say that dysphagia in patients with inflammatory myopathy seems to be largely caused by impaired pharyngeal muscle contraction originating from suprahyoid muscle weakness. [13]

Initial complaints in this patient Initial complaints of this patient were weakness in all four extremities, starting from both legs to both arms so that he could not walk which was preceded by a purplish red rash on the chest, around the eye area. This complaint is in accordance with the clinical manifestations of dermatomyositis, namely progressive muscle weakness, especially in the proximal muscles, and typical skin lesions in the form of a purplish red rash around the eyes, known as heliotrope rash, which is a typical sign of dermatomyositis.[14]

Laboratory examination results in this patient showed an increase in C3, C4 and serum Creatinine kinase (CK) and hypoalbumin. Increased serum CK levels are a sensitive indicator for the diagnosis of dermatomyositis because more than 90% of patients with classic dermatomyositis have significant increases in CK enzyme levels. [15] Anti-nuclear antibody (ANA) tests in this patient were negative. Negative ANA results do not exclude the diagnosis of dermatomyositis, as 40% of dermatomyositis patients can have negative ANA results, in patients with strong clinical symptoms for dermatomyositis it can indicate a certain subtype of the disease or a different prognosis. [16]

EMG examination shows demyelinating motor polyneuropathy. EMG results show demyelinating motor polyneuropathy with suspicion of muscle disease in accordance with the characteristics of EMG in dermatomyositis. EMG in dermatomyositis can show myopathic features such as short polyphasic motor unit potentials with low amplitude, and increased spontaneous activity.[17]

The biopsy results in this patient are vacuolar degeneration in some basal cells, the dermis layer shows a mild infiltration of lymphocyte cells at the dermo-epidermal junction accompanied by the distribution of melanophages according to the description of dermatomyositis, namely in dermatomyositis there is an increase in dermal mucin, vacuolar changes in the basal cell layer, and mild to moderate inflammatory mononuclear cell infiltrate. 10 The biopsy results in this patient are in accordance with the literature describing vacuolar changes in the basal layer.

The first FEES examination results obtained pre-swallowing poor oral hygiene, weak lip strength, puffed cheeks leaking right and left. The results of the FOL examination of velopharyngeal movement, weak symmetrically on both sides, heavy standing secretion, symmetrical vocal cord movement, hypopharyngeal sensitivity no leakage right and left. According to research, leakage is the entry of food bolus into the hypopharynx before the swallowing process begins. This is caused by impaired tongue function (1/3 posterior) which forms the glossopharyngeal valve so that the glossopharyngeal muscle is not strong enough to contract (approximation) to the posterior pharyngeal wall. Neurogenic dysphagia (post-stroke or head trauma), leakage occurs due to damage to the glossopharyngeal nerve (n.XI) which carries sensory components to the center and damage to the motor component by the hypoglossal nerve (n.XII).[18]

In the swallowing assessment, the first food given was strained porridge, heavy residue was found in the vallecula and

piriform sinus and penetration-aspiration occurred. This occurs because of a decrease in the elevation of the hyoid muscle and the elevation of the epiglottis that is lost or decreased, as well as weakness of the pharyngeal muscle contractions, causing difficulty in bolus clearance.[14] Decreased or weak closure of the laryngeal structure (retroversion of the epiglottis, glottis closed by the arytenoids and impaired closure or elevation of the vocal folds) causes penetration-aspiration.[14] The diagnosis of this patient is severe oropharyngeal dysphagia with penetration and aspiration so that the patient requires modification of food and drink through NGT.

The patient received methylprednisolone therapy with a dose of 1 gram which was then slowly reduced. The use of methylprednisolone in high doses (pulse therapy) is the right approach for initiating therapy in cases of severe dermatomyositis, aiming to induce rapid remission and then continue with dose reduction. The inadequate clinical response to single steroid therapy in this case suggests that dermatomyositis often requires a combination therapy approach, including the addition of other immunosuppressive agents.[19]

The patient was then given cyclophosphamide therapy for 6 cycles and showed an improvement response. A positive clinical response after the second cycle of cyclophosphamide showed the effectiveness of this drug in cases of refractory dermatomyositis. Deakin et al. in a large cohort study found that cyclophosphamide can improve skin and muscle manifestations in dermatomyositis. Cyclophosphamide administration in several cycles is often used to optimize effectiveness and minimize toxicity can be given 6-12 cycles for optimal therapeutic effect. [20]

FEES evaluation in this case after the sixth administration of cyclophosphamide without using NGT, showed changes in swallowing complaints. The results of the FEES evaluation were obtained in the pre-swallowing phase well. FOL standing secretion examination became mild although leakage was still found on the right side and epiglottis edema. Examination of the swallowing assessment phase, the patient was able to swallow 6 types of food, requiring five attempts to swallow and three rinses until clean and no aspiration and penetration were found. The diagnosis of this patient became mild oropharyngeal dysphagia with modification of soupy foods and avoiding flour porridge, milk.

The patient continued the seventh cyclophosphamide therapy. The evaluation results in this patient after two months after FEES evaluation showed significant improvements in the form of increasing body weight, no choking when eating and drinking, no complaints of a lump in the throat and muscle weakness in the limbs improved so that the patient could walk.

5. CONCLUSIONS

One case of a 39-year-old man with oropharyngeal dysphagia in a dermatomyositis patient. Administration of cyclophosphamide therapy showed significant improvement in this patient.

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