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Early-Onset Dysphagia Leading to the Diagnosis of Dermatomyositis and Polymyositis-Scleroderma Overlap: Report of Two Cases

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Abstract

Dysphagia can be the first presenting symptom of autoimmune myopathies, often leading to delayed diagnosis and treatment. We report two cases of severe dysphagia leading to the diagnosis of inflammatory myopathies. The first patient, a 26-year-old man, presented with progressive oropharyngeal and esophageal dysphagia and classic dermatomyositis skin manifestations. He responded well to intravenous methylprednisolone, IV immunoglobulin (IVIG), cyclophosphamide, and maintenance therapy with mycophenolate mofetil, achieving full recovery. The second patient, a 48-year-old woman, developed progressive dysphagia and weight loss and was diagnosed with polymyositis-scleroderma overlap syndrome. Treatment with corticosteroids, IVIG, rituximab, and mycophenolate led to significant improvement in swallowing and functional status. These cases highlight the importance of early recognition of autoimmune dysphagia and a multidisciplinary approach to management to prevent complications such as aspiration pneumonia, malnutrition, and long-term disability.

Keywords: Dysphagia, Dermatomyositis, Polymyositis-Scleroderma, Oman

Introduction

Dysphagia is a significant but often under-recognized manifestation of autoimmune rheumatologic diseases. It may result from myositis affecting striated muscles of the oropharynx and esophagus, mucosal involvement, or extrinsic compression from vasculitis. Early recognition is crucial to prevent malnutrition, weight loss, aspiration pneumonia, and impaired quality of life.

Autoimmune dysphagia may be the initial symptom of conditions such as dermatomyositis, polymyositis, overlap syndromes, Sjögren's syndrome, systemic lupus erythematosus, and systemic sclerosis. Accurate diagnosis requires exclusion of structural and primary motility disorders through endoscopy, imaging, and manometry.

We report two cases demonstrating the spectrum of autoimmune dysphagia and discuss their clinical course, diagnostic evaluation, and management.

Case Presentations

Case 1

A 26-year-old Omani man presented with a six-month history of progressive dysphagia to solids, resulting in an 18 kg weight loss. Initially investigated for gastrointestinal causes, including OGD and urea breath test, all results were normal. ANA was weakly positive.

Upon presentation at Suhar Hospital, he developed skin lesions consistent with dermatomyositis, including heliotrope rash, Gottron's papules, and periorbital edema. Laboratory tests revealed CK 13,671 U/L, ALT 87 U/L, and AST 200 U/L. ANA and ENA were negative. Tumor markers, viral screening, and CT scans were unremarkable.

He was started on IV methylprednisolone (1 g daily × 3 days), azathioprine (2–3 mg/kg), and oral prednisolone (1 mg/kg). Dysphagia worsened, with absent gag reflex and proximal muscle weakness, necessitating NG-tube feeding.

He received IVIG (0.4 g/kg × 5 days) and intravenous cyclophosphamide (500 mg every 2 weeks for 6 doses) with PCP prophylaxis. Maintenance therapy included mycophenolate mofetil 1 g BID. Over several weeks, dysphagia resolved, weight increased from 44 kg to 64 kg, CK normalized, and he returned to normal oral intake and work.

Case 2

A 48-year-old female nurse assistant with hypothyroidism presented with progressive dysphagia, nasal regurgitation, dysphonia, and significant weight loss (78 kg → 39 kg) over approximately one year. She had previously undergone extensive gastrointestinal and oncologic evaluations, including OGD, barium swallow, and pan-CT scans, all unremarkable. ANA was strongly positive (>1:640), and anti-PM-Scl antibodies were detected. CK and troponin were elevated, and chest HRCT showed mild interstitial lung disease.

She was diagnosed with polymyositis-scleroderma overlap syndrome. Treatment included IV methylprednisolone (1 g × 3 days), IVIG (0.4 g/kg × 5 days), Rituximab (2 g in two divided doses), mycophenolate mofetil 1 g BID, and supportive physiotherapy. NG-tube placement was refused. Over subsequent months, swallowing and dysphonia improved, weight increased to 43 kg, CK decreased to 400 U/L, liver enzymes normalized, and she resumed daily activities with ongoing Rituximab maintenance every six months.

Discussion

Autoimmune dysphagia arises from immunologically mediated muscle or neural involvement of the swallowing apparatus. It may precede other systemic manifestations, delaying diagnosis. Both reported cases illustrate the need for a multidisciplinary approach, including rheumatology, gastroenterology, ENT, nutrition, and physiotherapy.

IVIG is effective for steroid-resistant esophageal involvement, with studies showing rapid improvement in swallowing and early removal of feeding tubes. Corticosteroids remain first-line therapy, while immunosuppressive agents such as cyclophosphamide, mycophenolate, methotrexate, and rituximab serve as steroid-sparing or rescue therapy. Interventional procedures (cricopharyngeal myotomy, esophageal dilation) and temporary feeding tubes may be required in severe cases.

Early recognition of autoimmune dysphagia is crucial to prevent malnutrition, aspiration pneumonia, and long-term disability. Comprehensive evaluation and prompt initiation of immunosuppressive therapy are key to favorable outcomes.

Conclusion

Dysphagia may be the first presenting symptom of autoimmune myopathies. Clinicians should maintain a high index of suspicion, particularly when structural and motility causes are excluded. Early, aggressive, multidisciplinary management can restore function, prevent complications, and improve quality of life.

Patient Consent

Written informed consent was obtained from both patients for publication. Identifying details have been withheld.

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