An Unusual Presentation of Dysphagia in a Young Male with Myositis

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Published on 30th December 2024

ABSTRACT

Dysphagia is a rare presentation in patients with viral myositis and only 10%-20% of patients with myositis developed weakness of oropharyngeal, laryngeal, and oesophageal musculature that leads to impairment in the oropharyngeal phase of swallowing. The proper mechanism of dysphagia in patients with myositis remains unclear and it is thought to be due to impaired muscle contractions and decreased hyo-laryngeal excursion that leads to impaired relaxation of the upper oesophageal sphincter. Delayed treatment, bulbar involvement, respiratory involvement, old age, and occurrence of malignancy are associated with poor prognosis. Here we are describing a case of a 25-year-old gentleman who presented with fever, myalgia, and dark-colored urine, who was diagnosed as viral myositis with dysphagia, and a holistic approach in the management of the disease.

Keywords: Dysphagia, Myositis, Inflammation

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INTRODUCTION

Dysphagia is also a known complication during the course of the disease and has been reported in 10% to 20% of patients with viral myositis.[1] As the pharynx and upper oesophageal sphincter (UES) consist of skeletal muscle, reduced pharyngeal contraction and dysfunction of the Upper oesophageal sphincter are the two principal swallowing problems observed in these patients. The primary treatment for viral myositis is corticosteroids.[1] Other immunosuppressants and immunomodulatory therapies have been used for steroid-refractory cases.[2] Together with medical treatment, swallowing rehabilitation therapy is common.

Dysphagia in viral myositis patients usually gradually progresses over several days to months. To our knowledge, patients with viral myositis who visit the emergency room with sudden-onset dysphagia without notable aggravation of other symptoms have rarely been reported.[3] In addition, severe dysphagia with liquids, solids and even saliva (aphagia) is also uncommon.[3] 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, neurologist, and rehabilitation doctor was necessary for the diagnostic process to rule out potential acute structural obstruction and/or neurological conditions. After the exclusion of head and neck cancer, laryngeal oedema, infection, gastrointestinal causes and neurological causes, the diagnosis was acute severe dysphagia due to exacerbation of viral myositis.

Typical symptoms of myositis-related dysphagia are coughing, choking, bolus-sticking in the pharynx, and swallowing problems with dry and solid food consistencies. Early diagnosis and specific therapeutic management of dysphagia is crucial as it can lead to aspiration pneumonia with respiratory failure, which is the leading cause of mortality in patients with myositis

Cite this article as: James J, Shahanas PS, Philips GM, Thomas J, Sana A, Ittiachen MM, et al. An Unusual Presentation of Dysphagia in a Young Male with Myositis. Kerala Medical Journal. 2024 Dec 30;17(4):216–19. | DOI: https://doi.org/10.52314/kmj.2024.v17i4.682

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Figure 1a & 1b.**Saliva Pooling, Valleculae Pooling**

CASE PRESENTATION

A 25-year-old male with no known comorbidities presented with complaints of generalized diffuse muscle pain associated with stiffness of two weeks duration. He also had prodromal flu-like symptoms one week before the onset of presenting complaints. It was also associated with sore throat. There was no history of cough, breathlessness, joint pain, early morning stiffness, burning micturition or bleeding manifestations.

He was admitted with the above-mentioned complaints and clinical findings in the Medical intensive care unit. Routine Blood investigations showed elevated blood counts (Total count -20 x 109 /L), abnormal liver function test (SGOT/SGPT-250/780 U/L), elevated creatinine phosphokinase levels (CPK) (36650 U/I), CRP and ESR levels on day 1. In view of severe weakness of bilateral upper limbs(left>right), he was started on supportive medications. There were elevated inflammatory markers, for which he was started on intravenous antibiotics and analgesics. A T2 weighted MRI scan revealed extensive abnormal high signals involving skeletal muscles of bilateral upper and lower extremities, trunk, and muscles of head and neck with oedema in myofascial plains and patchy areas of subcutaneous oedema. He was adequately hydrated with Intravenous fluids (NS/RL) at 75 ml/hr. Due to progressive weakness in bilateral upper and lower limbs, nerve conduction study was done on day 3 which was normal. Swelling and weakness over bilateral upper and lower limbs gradually increased for which he was started on 3 % saline dressing. A tropical panel was sent (Dengue/ Lepto/malaria/scrub) and it was negative. Arterial and venous doppler of the bilateral upper limb was done to rule out a thrombotic event in view of the progressive swelling and erythema and it was normal. In view of the clinical suspicion of myositis, he was started on Intravenous pulse steroids. CPK levels were monitored daily and there was a gradual elevation in levels. CPK level which was 36650 U/L at the time of admission raised to 43000 U/L on the subsequent day and then to 69000 U/L. It reached a range of 85,221 U/L on day 4. CPK levels crossed 1 lakh (102,700 U/L) on day 6. His urine gradually became a dark brown colour over the successive days. A Myositis profile was sent, which turned out to be negative. Blood culture and urine culture were negative for any growth. There was a gradual elevation in Liver enzymes with each passing day. Liver enzymes (SGOT/SGPT) reached a level of 1833/659 U/L on day 5. A serology panel was sent and it turned out to be negative. A muscle biopsy was taken which showed inflammatory myopathy suggestive of viral myositis. In view of poor response to steroids, gradual elevation in CPK levels and clinical deterioration, he was started on Immunoglobulin (IVIG) infusion after discussing with the rheumatology team.[2] There was an improvement in his existing muscle pain and weakness noted three days after the initiation of therapy. A fall in the levels of CPK level was noted on day 6 and the value decreased from 102,700 to 83245 U/L. Surprisingly his renal functions were stable throughout the course of hospital stay. He was noted to have severe dysphagia and nasality of voice. On detailed evaluation by the swallowing pathology team, it was concluded that there was a pooling of saliva **(Figure 1a and 1b)** and nasal regurgitation was detected.

Gag reflex was normal bilaterally and hyolaryngeal elevation was delayed. Oral and palatal strengthening exercises were initiated by the swallow pathology team. Masoko maneuver and pharyngeal exercises were regularly done to reduce the pooling of secretions. In view of dysphagia, he was started on Ryles tube feeding. He was medically stabilized and was shifted to Ward. The patient had an episode of fever secondary to immunosuppression by steroids. Blood culture sent at this point of time had growth of Multidrug resistant Klebsiella. Antibiotics were then hiked to a broad spectrum category. He was simultaneously started on swallowing therapy and physiotherapy.

He improved symptomatically and his CPK levels came to normal levels. He was advised to do out-of-bed mobilizations and physiotherapy. He gradually regained muscle power over these successive days and his swallowing improved. Ryles tube was then removed and he started having blended semi-solids, at frequent intervals with dietary consultation to ensure nutritional supply. He was advised to continue palatal strengthening exercises to improve his voice. The patient improved symptomatically and was discharged with advice to follow up as an outpatient in OPD.

Given its complexity, diagnosing and treating dysphagia in the setting of Viral myositis requires a multidisciplinary approach, including a gastroenterologist, neurologist, rheumatologist, otolaryngology surgeon, nutritionist, and rehabilitation medicine doctor. Our experience with this patient emphasizes these management principles.

DISCUSSION

Myositis is a recognized complication of numerous systemic viral infections including influenza.[3] In adults, the typical pattern is characterized with myalgia and marked proximal muscle weakness in upper and lower limbs and resolves slowly over weeks rather than days. Viral infections cause mild myalgias to severe rhabdomyolysis, leading to renal failure and cardiac arrhythmias.[1] The exact mechanism of viral myositis is still vague, but some attributions include direct viral invasion and disruption of myositis resulting in acute myositis, viral-mediated myotoxic cytokines, and viral-induced autoimmune reaction.[1] Viral and post-viral myositis can be a potentially life-threatening disease with extensive muscle involvement. Microorganisms induce myositis via immune mechanisms rather than directly infecting the muscle tissues, for example, viruses and parasites cause diffuse generalized myalgias or multifocal myositis.[1] Viral myositis has been reported with a wide range of viruses but the most common include Influenza A and B viruses, enteroviruses, HIV, hepatitis viruses, parainfluenza; coxsackie. In adult patients infected with SARS-CoV-2, necrotizing autoimmune myositis has been reported in up to 10% of SARS-CoV2- infected patients.[1] The clinical presentation of extremely high CPK levels (>10 000 U/L), associated myalgia, and weakness is typical. Elevated CPK levels without associated rhabdomyolysis and acute kidney injury have also been reported in adult patients. There have also been reports in adult patients of SARS-CoV-2-associated autoimmune myositis presenting with elevated serum CPK levels and acute kidney injury, although not requiring Renal replacement therapy.[1] Delayed onset necrotizing myositis, confirmed by muscle biopsy, has also been reported in an adult patient following an infection with SARS-CoV-2.[1]

MRI appears streaky or patchy infiltration of muscle by abnormally high T2-weighted signal.[4] Heterogeneous or diffuse enhancement of muscle lesions is often seen. Interestingly the time frame between viral fever to the onset of myositis can vary up to 3 weeks. Most common clinical feature is muscle weakness accompanying severe pain especially in the limb girdles and paraventricular musculature, pelvic girdle and shoulders. The neck flexors are commonly affected and, in some patients, the neck extensors.[5]

There are only few studies that investigated the pathophysiological characteristics of myositis related dysphagia.[6] Typical symptoms of myositis related dysphagia are coughing, choking, bolus-sticking in the pharynx and swallowing problems with dry and solid food consistencies. Prompt diagnosis of dysphagia is crucial to avoid complications like nutritional deficits, decreased quality of life, aspiration pneumonia with respiratory failure eventually leading to poor overall prognosis.[7] Serum CPK elevation has a high positive predictive value. Instrumental assessments, e.g., flexible endoscopic evaluation of swallowing (FEES) or videofluoroscopy (VFSS) are considered the diagnostic gold standard in the diagnosis of dysphagia.[7] Therapy must be an individualized combination of pharmacological and non-pharmacological regimens. Diet and food consistency modifications, swallowing maneuvers, and speech therapies have a positive outcome for the patient.[7]

Dysphagia in viral myositis is much more common in steroid-resistant patients. For steroid-resistant dysphagia, high-dose IVIG and cyclophosphamide have been effective in a few cases.[8] However, unlike many previous reports, there was a gradual improvement of dysphagia in our patient. Therefore, it is difficult to discuss the therapeutic effect of a certain drug in this case. Our case demonstrates that complete improvement of life-threatening steroid-resistant dysphagia can occur if aggressive medical and rehabilitative therapy is maintained.[9]

IVIG at 2 g/kg had significant improvement in muscle strength, decrease in CPK levels and helped to reduce corticosteroid dosage. In one study, approximately 82% of 73 patients were able to return to oral feeding.[8] Proposed mechanisms of IVIG include blocking Fc receptors on phagocytic cells, inducing inhibitory Fc-γ-RIIB receptors on effector macrophages through interleukin (IL)-33,7,8 interaction with dendritic cell

differentiation and maturation, reducing proinflammatory subsets of peripheral blood monocytes (CD14+ CD16++), inhibiting leukocyte adhesion molecule binding to the endothelium, blocking of Fas ligand-mediated apoptosis.[8]

This is a case of viral myositis that caused severe inflammation of muscles, mainly proximal muscles and even muscles involving deglutition. Usually, viral myositis presents with rhabdomyolysis and renal failure. One peculiarity of this case is even with high degrees of rhabdomyolysis, there was no renal involvement. The patient had a history of flu-like illness 1 week before the onset of symptoms. Also, his COVID antibody was positive which is attributed to a viral illness that occurred before the onset of symptoms. His High level of CPK level above 1 lakh shows the severity of muscle involvement in this case. He also had dark coloured urine which shows evidence of myoglobinuria. Even though he had myoglobinuria, his renal functions were preserved. In this case, his muscles of deglutition were also involved, which is a rare pattern of muscle involvement and is not usually seen in post-viral myositis.[10] The treatments we gave here were IV steroids and IVIG. The patient well responded to the treatment and he improved symptomatically after IVIG administration.

CONCLUSION

Typical presentation of myositis includes severe myalgia, proximal muscle weakness, and myoglobinuria with renal failure, leading to renal replacement therapy. This case is a different presentation of viral myositis with severe involvement of proximal muscles, muscles of deglutition, and myoglobinuria. The patient had normal renal functions throughout the course of the hospital stay. He responded well to IVIG therapy.

This unique presentation of dysphagia following myositis in a critically ill patient highlights the significance of prompt diagnosis at a crucial stage of the disease-preventing further complications which can be attributed to the clinical judgment that identified the early symptoms of dysphagia secondary to myositis. The management approach involving a multidisciplinary team including physicians, intensivists, rheumatologists, clinical pharmacists, speech and swallowing pathologists, and clinical nutritionists significantly created a positive outcome in the patient's recovery. In conclusion, Viral myositis can present with the involvement of muscles of deglutition and with stable renal functions. Prompt treatment with steroids and IVIG can save the life of the patient and thus can reduce mortality rates.

END NOTE

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Financial and Competing Interests' Disclosure: Nil

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