May 2nd, 12:00 AM

Case Report: Dysphagia in Inclusion Body Myositis Leading to Respiratory and Gastrointestinal Complications

Veroneka Mikhail
Jefferson Health NJ

James A. Espinosa
Jefferson Health NJ

Alan Lucerna
Jefferson Health NJ

Case Report: Dysphagia in Inclusion Body Myositis Leading to Respiratory and Gastrointestinal Complications

Veroneka Mikhail DO, James Espinosa MD, Alan Lucerna DO

Emergency Medicine Residency and Department of Emergency Medicine, Jefferson Health NJ

Abstract:

Inclusion body Myositis (IBM) stands as a rare and complex neuromuscular disorder (NMD) characterized by progressive muscle weakness and atrophy. Among its cardinal symptoms are dysphagia and respiratory distress, which are the most common cause of death in this disease. While the differential diagnoses of respiratory distress is vast and includes aspiration, pneumonia, acute coronary syndrome, emphysema, and congestive heart failure, a clinician should recognize that respiratory distress can also be secondary to dysphagia in NMDs like IBM and can quickly become life threating. Here we present the case of a 68-year-old female with a history of IBM who presented for respiratory distress, was found to have severe dysphagia, and subsequently required intubation and percutaneous endoscopic gastrostomy (PEG) tube placement.

Case Presentation:

68-year-old female with a history of inclusion body myositis presented to the emergency department (ED) in respiratory distress with chief complaint of sudden shortness of breath that occurred prior to arrival after she aspirated while eating cheese and crackers, and drinking wine. At that time, patient (pt) denied any chest pain, coughing, nausea, and vomiting. Pt explained that approximately six years prior to this visit she underwent series of tests including multiple barium swallow studies, electromyography, and anti-nuclear antibody (ANA) testing and was diagnosed with IBM. She also explained that she had multiple hospitalizations in the past due to aspiration pneumonia and even a tracheostomy in 2019.

Upon arrival to the ED, pt's vitals were as follows: Heart rate 117 beats per min, respiratory rate 30 breath per minute, oxygen saturation (O2) 86%, blood pressure 162/75 mmHg, and temperature 99.2 degrees F. The physical exam showed mild tachypnea, intercostal retractions, bilateral expiratory wheezing, and diminished breath sounds at the right lung base. Lactate 2.6. no leukocytosis. All other labs within normal limits. Electrocardiogram showed sinus arrhythmia 99 beats per minute, no ST changes, no QT prolongation. Pt was immediately placed on 6L nasal cannula (NC) with minimal improvement in O2 status, then transitioned to 11L oximyzer with O2 stat 96%. Arterial blood gas showed: pH 7.44, pCO2 40, pCO2 81, bicarb 27. Chest x-ray (CXR) showed bibasilar lung infiltrates with mild perihilar interstitial edema. Computerized tomography angiogram (CTA) scan preformed and showed mild enlarged pulmonary trunk, and patchy airspace opacities in the right upper, right lower, and left lower lobes representing multi-lobar pneumonia (figure 1). Pt was given antibiotics (doxycycline and ceftriaxone), duonebs, tapering dose of solumedrol placed on placed on nothing by mouth (NPO), code status confirmed as full code, and pt was admitted to the intensive care unit for sepsis and acute hypoxic respiratory failure secondary due to multi-lobar aspiration pneumonia.

In the ICU, nasogastric tube (NGT) was placed, pt was started on trickle feeds at 20ml/hr and O2 saturation stabilized and pt was weaned down to 6LNC. Overnight, pt de-satted to 80%, was titrated up to 10L oxymizer without improvement, then placed on 50L at 50% high flow nasal cannula (HFNC) but continued to show signs of respiratory distress including O2 stat ~80%, increased work of breathing, copious oral secretions, and bilateral wheezing despite multiple suctioning. Pt was subsequently intubated, Infectious disease consulted, and she was switched to eravacycline. Pt was successfully extubated 3 days later to 50L HFNC and safely tapered down to 2L NC and wanted NGT removed. Pt was kept NPO, incentive spirometer, aspiration precautions, head of bed elevated, Physical therapy, Occupational therapy, and Speech and Swallow therapy were placed. Speech evaluation preformed via Videofluroscopic swallow studies showed severe pharyngeal dysphagia, pooling of food consistencies, and moderate esophageal dysphagia (figure 2). It was determined that pt is unable to tolerate by mouth (PO) diet and is a high aspiration risk. Pt declined NGT, dobbhuff (DHT), and PEG despite multiple detailed discussion and wanted to have nutrition PO as she did at home. The patient's family gave her Jello PO against medical advise, and soon after pt developed another episode of respiratory distress where she became hypoxic SpO2 86-89% on HFNC 100% FiO2 55lpm. Stat portable CXR shows complete atelectasis of the left side with suspected mucus plugging (figure 3). Pt was agreeable to intubation, temporary DHT placement, and eventual a PEG tube due to malnutrition in the setting of recurrent aspiration. Sputum cultures grew pseudomonas and pt was started on 7 day course of meropenem, 3% saline nebs, and chest physical therapy. Gastroenterology was consulted and PEG tube was placed once pt's respiratory status stabilized. Pt was extubated 4 days after PEG tube placement, tube feeds were up-titrated slowly until goal of 41ml/hr was met. Pt continued to clinically improve and was stable for discharge home 1 week later.

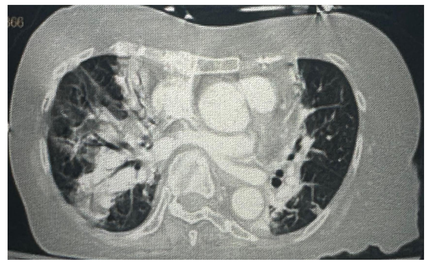

Figure 1: CTA of the chest showing mild enlarged pulmonary trunk measuring 3.2cm. Patchy airspace opacities in the right upper, right lower, and left lower lobes presenting multi-lobar pneumonia.

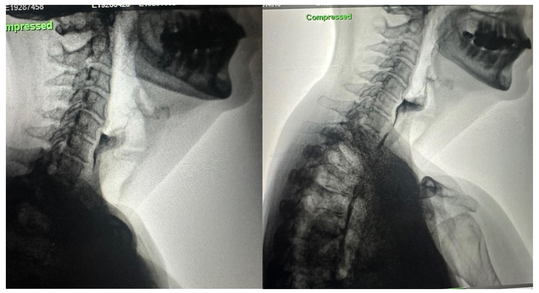

Figure 2: FL SLP video fluoroscopic swallow study showing moderate to severe pharyngeal dysphagia. Pooling of consistencies within vallecula. Piriform sinus retention of thin and thick liquids. Moderate to severe esophageal dysmotility.

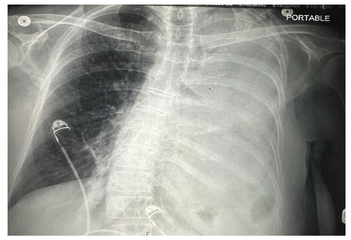

Figure 3: Chest x-ray showing complete opacification of the left lung with mild mediastinal deviation to the Left.

References:

1. Panginikkod S, Musa R. Inclusion Body Myositis. [Updated 2023 Jul 17]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 Jan-. Available from: https://www.ncbi.nlm.nih.gov/books/NBK538200/
2. Naddaf E. Inclusion body myositis: Update on the diagnostic and therapeutic landscape. Front Neurol. 2022 Sep 27;13:1020113. doi: 10.3389/fneur.2022.1020113. PMID: 36237625; PMCID: PMC9551222.
3. Naddaf E, Barohn RJ, Dimachkie MM. Inclusion Body Myositis: Update on Pathogenesis and Treatment. Neurotherapeutics. 2018 Oct;15(4):995-1005. doi: 10.1007/s13311-018-0658-8. PMID: 30136253; PMCID: PMC6277289.
4. Greenberg, Steven A.. Pathogenesis of inclusion body myositis. Current Opinion in Rheumatology 32(6):p 542-547, November 2020. | DOI: 10.1097/BOR.0000000000000752
5. Mohannak N, Pattison G, Hird K, Needham M. Dysphagia in Patients with Sporadic Inclusion Body Myositis: Management Challenges. Int J Gen Med. 2019 Dec 5;12:465-474. doi: 10.2147/IJGM.S198031. PMID: 31824189; PMCID: PMC6901064.
6. Nagy S, Khan A, Machado PM, Houlden H. Inclusion body myositis: from genetics to clinical trials. J Neurol. 2023 Mar;270(3):1787-1797. doi: 10.1007/s00415-022-11459-3. Epub 2022 Nov 18. PMID: 36399165; PMCID: PMC9971047.
7. Oh, Terry H. MD; Brumfield, Kathlyn A. OTR; Hoskin, Tanya L. MS; Kasperbauer, Jan L. MD; Basford, Jeffrey R. MD, PhD. Dysphagia in Inclusion Body Myositis: Clinical Features, Management, and Clinical Outcome. American Journal of Physical Medicine & Rehabilitation 87(11):p 883-889, November 2008. | DOI: 10.1097/PHM.0b013e31818a50e2
8. Middelink M, Voermans NC, van Engelen BGM, Janssen MCH, Groothuis JT, Knuijt S, Zweers-van Essen H. Indications for Tube Feeding in Adults with Muscular Disorders: A Scoping Review. J Neuromuscul Dis. 2023;10(5):777-785. doi: 10.3233/JND-230014. PMID: 37483025; PMCID: PMC10578233.

Discussion:

Pathophysiology of IBM:

The pathophysiology of IBM, is not yet completely understood however, it is classified as an idiopathic myopathy with components of inflammatory, autoimmune, and muscle degeneration (1,2). It is thought that IBM muscles contain abundant inflammatory molecules which upregulate MHC1 and contain restricted T-cell receptor sequences which response to a specific antigen leading to increased destruction of myofibers (3,4). The Gold standard for diagnosis of IBM is muscle biopsy which shows inflammatory cells surrounding and invading non-necrotic muscle fibers, presence of vacuoles rimmed by a membranous cytoplasmic material, atrophic fibers, and mitochondria showing an increased number of cytochrome c oxidase negative fibers (3,5,6). Seropositive for anti-cytosolic 5'-nucleotidase 1A (anti-cN1A) autoantibodies are 76% sensitive and 92 to 96% specific for IBM (3,6)

Presentation and epidemiology of IBM and the presence of dysphagia:

IBM is the most common acquired myopathy after the age of 50, effecting 5-200 cases per million adults with a 2:1 male to female ratio (1,2). IBM presents with painless progressive weakness, often asymmetric, mainly affecting finger flexors and knee extensors (1,3). 15-50% of patients have atypical presentation such as dysphagia, dysphonia, foot drop, proximal upper limb weakness, or head drop (1,2,3). The most common cause of death in IBM is dysphagia leading to respiratory complications such as aspiration pneumonia (2,3). The underlying etiology of dysphagia in IBM is not well understood however, it is commonly found that spasm of the upper esophageal sphincter, fibrosis of cricopharyngeal muscle, and suprahyoid muscle weakness lead to pyriform fossa stasis and diminished descending bolus forces (5).

Laboratory and imaging studies:

Elevated leukocytosis, and creatinine kinase (CK) may be present in pts with IBM, however, a normal leukocytosis or CK does not rule out the disease. Electrolyte abnormalities due to vomiting or malnutrition may be seen on a basic metabolic panel. ABG, lactate, CXR, CT chest, and blood cultures should be obtained in patients to assess for aspiration pneumonia and sepsis. The gold standard in the diagnosis of dysphagia in IBM is Videofluroscopic swallow studies or flexible endoscopic evaluation of swallowing (5).

Management of dysphagia in IBM:

Treatment of dysphagia in IBM is categorized into non-invasive and invasive treatments. Non-invasive strategies include: swallow exercise/ Mendelsohn maneuver to helps strengthen lingual muscles, intravenous immunoglobulin/ immunosuppressive both were found minimally beneficial with mild symptom resolution only lasting up to 1-2 month, diet modifications including enteral feeds, and alternating solids and liquids. Invasive management includes Balloon dilation of the pharyngoesophageal segment to decrease cricopharyngeal retraction, Botulinum toxin injection to the cricopharyngeal muscle, cricopharyngeal myotomy, and insertion and use of a PEG feeding tube for dysphagia leading to malnutrition/weight loss and respiratory infections (5,7,8).

Conclusions:

In this study, we reviewed how aspiration in a patient with Inclusion Body Myositis (IBM) presented, was managed, and what course it took. Dysphagia and respiratory distress represent the most life threating complications of IBM. Healthcare professionals must remain vigilant, considering IBM as a potential underlying cause in patients presenting with dysphagia and respiratory distress, particularly in the absence of more common etiologies. Ultimately, by enhancing awareness and understanding of IBM, we can strive to optimize patient care and enhance the quality of life for those living with this challenging neuromuscular disorder.