Case Report

Unusual Presentation of Myasthenia Gravis as Barium Aspiration Pneumonitis

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Abstract

Isolated dysphagia is a rare form of presentation of myasthenia gravis (MG), resulting in discoordinated peristalsis. Accidental aspiration of barium sulfate may occur during radiological examinations of the upper gastrointestinal system using contrast media in conditions affecting the anatomical and functional integrity of the oropharynx and the esophagus. Though inert, barium may result in severe pulmonary inflammatory reaction and acute respiratory distress syndrome (ARDS). Thus, it is important to identify patients at risk of aspiration. Timely consideration of MG during evaluation of dysphagia is crucial to prevent complications and improve the quality of life. Whenever neuromuscular weakness is suspected, videofluroscopic technique for barium swallow needs to be advocated. We, hereby, report an unusual case of MG who presented with dysphagia only and developed fatal barium aspiration pneumonitis.

Key words: Aspiration pneumonitis, barium aspiration pneumonitis, barium swallow, dysphagia, myasthenia gravis

Introduction

Myasthenia gravis (MG) is an autoimmune neuromuscular disorder, with incidence of one in 5,000.[1] Rarely, it may present with isolated dysphagia affecting the functional integrity of the upper gastrointestinal tract. Aspiration of barium sulfate can accidentally occur during contrast-enhanced studies of the upper gastrointestinal tract. We report an unusual case of MG who presented with dysphagia only and developed fatal barium aspiration pneumonitis.

Case Report

A 31-year-old lady presented to our hospital with severe respiratory distress. Her clinical history revealed upper respiratory tract infection followed by dysphagia for which barium swallow evaluation was done. Following that, she developed respiratory distress and was intubated in view of hypoxemia. Bronchoscopy was performed that ruled out tracheoesophageal fistula. X-ray of her chest revealed multiple high-density nodules [Figure 1]. Computed tomography (CT) of her chest confirmed multiple high-density nodules with bilateral lower lobe consolidation. The patient was managed with ventilator support with high positive end-expiratory pressure (PEEP), antibiotics, analgesics, fluids, nasogastric (NG) feeds, and supportive care. As CT scan and bronchoscopy ruled out structural anomalies, we suspected pharyngeal muscle weakness as the cause of aspiration. After ruling out stroke, vasculitis, and hypercoagulable states, we checked antiacetylcholine receptor antibodies that were positive and the diagnosis of MG was arrived at. On day 10, she was tracheostomized in view of the need for prolonged ventilation. She was started on steroids and pyridostigmine and gradually weaned off ventilator support by day 35 and shifted out of the intensive care unit (ICU). Unfortunately, she developed severe respiratory distress 10 days later, requiring reintubation. X-ray of her chest showed atelectasis and consolidation, suggesting gastric aspiration despite tracheostomy and NG feeds. She succumbed to sepsis, multiorgan failure, and septic shock.

Discussion

MG is an autoimmune antibody-mediated T cell-dependent neuromuscular junction disorder,[1] presenting with a fluctuating and painless weakness involving specific muscle groups. Our case was unusual as dysphagia was the first and only manifestation of MG. During the course of MG, 40%[1,2] of patients suffer from dysphagia with 6-20%[3] of them presenting with dysarthria or dysphagia. Weakness of the oropharyngeal and the esophageal smooth muscles results in discoordinated peristalsis, increasing the risk of aspiration. Dysphagia and respiratory involvement continue to be significant sources of morbidity and mortality.[3] Factors such as infection, physical

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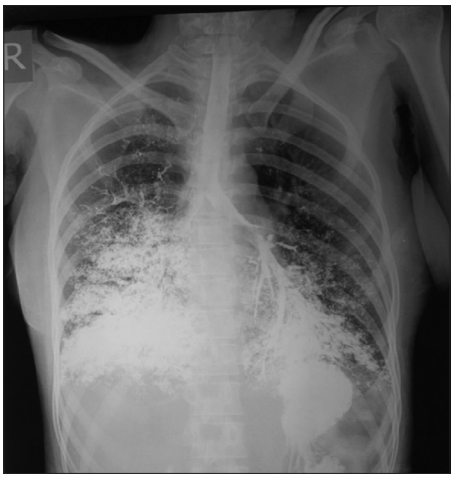

Figure 1: X-ray of the chest showing multiple high-density nodules in the lungs

or emotional stress, pregnancy, or drugs can trigger episodes of muscle weakness. In our case, we suspected respiratory infection to be the triggering factor for dysphagia.

Aspiration of barium sulfate into the lungs accidentally occurs during contrast-enhanced examinations of the upper gastrointestinal tract. The exact incidence is not known. The overall mortality rate associated with massive barium aspiration is approximately 30% but can exceed 50% in patients with initial shock or apnea, secondary pneumonia, or adult respiratory distress syndrome.[4,5]

Conditions affecting the anatomical and functional integrity of the oropharyngeal and esophageal segments have been described as the predisposing factors for aspiration. These are extreme age, neurological disorders such as cerebral infarction, brainstem infarction, intracranial hemorrhage, MG, parkinsonism, poliomyelitis, multiple sclerosis, and structural lesions such as tracheoesophageal fistula, neoplasm, congenital web, Zenker diverticulum, connective tissue disorders such as polymyositis, muscular dystrophy, alcoholism, psychological illness, and functional gastrointestinal disorders.[4-6]

Barium sulfate is a radiopaque contrast medium used for imaging studies of the gastrointestinal tract with high density, i.e., 4.5 g/cm^3.[3] Aspiration of barium is not expected to cause severe lung injury due to its relatively inert and nonirritant nature. But severe manifestations like pulmonary edema and acute respiratory distress syndrome (ARDS) may be more frequent than those suggested by the literature.[4,5]

It has been suggested that aspiration of gastric contents concomitant to barium aspiration plays an important role in severe pulmonary inflammatory reaction and progression to death. Hypersensitivity reactions may be caused by additives to commercial barium preparations. Complications of barium sulfate aspiration depend on the quantity, trachea bronchial distribution, and general physical condition of the patient.[4,5]

The aspirated barium is eliminated by coughing and the mucociliary apparatus. The remaining barium is removed by macrophages and will accumulate in the alveolar spaces and tracheobronchial lymph nodes that appear opaque for years. Particles may also pass directly across the alveolar epithelium into the alveolar or peribronchial interstitial tissue, thereby leading to fibrosis.[4,5]

The radiographic pattern is typically striking, high-density opacities as a result of the high atomic number of barium (56). The pattern visible in x-ray or in CT scan of the lung may be time-dependent due to a slow, progressive clearance of the barium particles. Plain x-ray of the chest remains the diagnostic technique of choice in the acute stage. Long-term fibrotic changes with crazy paving appearance can be detected on high-resolution CT scan.[4,5]

The treatment is based on clinical judgment. Bronchoscopy is recommended in the presence of arterial hypoxemia and dyspnea after massive aspiration to eliminate as much barium as possible and to obtain aspirates for microbiology testing. Bronchoalveolar lavage is not advisable due to the danger of dissemination of the contrast medium into the bronchoalveolar system. Aspiration of the gastric contents necessitates treatment with antibiotics. Aspiration of nonionic, iodinated contrast material may lead to pulmonary edema requiring aggressive treatment including oxygen, ARDS ventilation, Lasix, and steroids as in our patient.[4-6]

Myasthenic patients may require mechanical ventilator support for multiple reasons such as acute ventilator failure in myasthenic crisis, aspiration pneumonitis, or stridor due to bilateral abductor paralysis. Incidence of silent aspiration, as suggested by clinical experience, is quite high.[3] Aspiration may result in atelectasis, consolidation, and hypoxic respiratory failure.

Prevention of aspiration should be focused on early recognition of the predisposing factors, pretreatment with antireflux medications, and correct choice of the contrast media.[4] Iopydol, used for bronchography, and iodixanol, an iso-osmolar contrast medium used for intrarterial and intravenous contrast studies, demonstrate no pulmonary harm.

Prediction of the risk of dysphagia and aspiration is important in the management of MG. The modified barium swallow (radiographic videofluoroscopic technique) and fiber optic endoscopy help in functional evaluation.[2,7] Electromyographic studies and bedside clinical tools like speech pathology assessment and quantitative MG score correlate well with the risk of aspiration.[7]

Further management should aim toward swallowing safety and efficiency with diet modification, behavioral techniques, postural techniques, and nonoral routes of feeding. Compensatory swallowing skills like effortful swallow, Mendelson maneuver, and super-supraglottic swallow improve pharyngeal clearance. Compensatory postures like head rotating to the weak side divert the bolus to the contralateral

stronger side.[2,3] In our patient, a reaspiration could have been avoided by timely advocation of postural and feeding modifications.

Conclusion

A high index of clinical suspicion is required to identify patients at risk of aspiration. Timely consideration of MG during evaluation of dysphagia is crucial. Videofluroscopic barium swallow enhances safety in suspected neuromuscular weakness and minimizes the risk of accidental aspiration. Timely behavioral and postural modifications may help to avoid aspiration.

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