Pseudocyst of Pancreas Presenting in Respiratory Failure Due to Myasthenia Crisis

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Case Report

Abstract: We report a case history of a 43 yr old lady, who presented with acute abdominal pain and then went into respiratory failure. The patient was not a known case of myasthenia gravis. The Chest X-ray was normal, blood gas analysis showed respiratory acidosis, CT scan of abdomen showed pseudocyst of pancreas with necrosis in tail of pancreas. Myasthenia crisis can be triggered by a variety of factors, in this case by acute pancreatitis. In our case myasthenia gravis was diagnosed since we had difficulty in weaning her off from the ventilator. Myasthenia crisis with pseudocyst of pancreas and acute pancreatitis is rare. After putting her on ventilator, there was improvement in blood gas, but she could not be weaned off the ventilator even after repeated attempts because of poor respiratory muscle activity. Serum Acetylcholineesterase receptor auto antibodies were found to be high, and she was then diagnosed as having Myasthenia gravis. Myasthenia gravis should be suspected in a patient with unexplained respiratory failure.

Keywords: Myasthenia gravis, Pseudocyst of pancreas, Respiratory failure

Abbreviations: **SIMV**—Synchronized intermittent mandatory ventilation, **CT-SCAN**—Computerized tomography scan

Introduction

Acute pancreatitis with pseudocyst of pancreas can precipitate Myasthenia crisis. Myasthenia gravis patients may remain undiagnosed and can present directly in Myasthenia crisis. A variety of triggers have been described. We report this case of previously undiagnosed Myasthenia gravis with pseudocyst of pancreas who presented in Myasthenia crisis. Increased fatiguability and difficulty in weaning off from ventilator, led to the diagnosis of Myasthenia gravis with crisis.

Case Study

A 43 yr old lady, non alcoholic, non smoker, presented with acute pain in abdomen, vomiting, constipation, generalized distention of abdomen since 2 days. The abdominal pain was generalized and radiating to back. After 2 hours of admission, she developed acute severe breathlessness and was shifted to ICU. The diagnosis of acute Pancreatitis was made. She went into acute respiratory failure and was put on ventilator. She was conscious but not fighting with the ventilator

On examination: patient was febrile, Pulse: 140/min, Blood pressure: 150/80 mm Hg

Respiratory system: Intensity of breath sounds equal on both sides. No adventitious sounds

Cardiovascular System: Heart sounds were normal, no murmurs per abdomen: on admission, severe tenderness in epigastric region, with guarding and rigidity. Central nervous system: conscious, obeying commands, no sensory-motor neurodeficit Plantar flexion, Pupils of both eyes reacting to light.

Investigations: Hemoglobin: 7 gm/dl, Total leucocyte count: 19500(day 1st) Serum amylase: 258 U/L (0-90 U/L), Serum lipase: 142.92 U/L (5.6-51.3 U/L) Arterial Blood Gas Analysis showed Respiratory Acidosis. (pH – 7.3, pCO2 - 80mm Hg, HCO3 – 26, pO2-60mm Hg). Serum electrolytes, Liver Function and Renal Function Tests were normal, Creatinine phosphokinase enzyme was normal, Chest X ray Posteroanterior view: normal. Thyroid function tests were normal the patient was put on ventilator on volume control mode (without muscle relaxant as patient was not fighting with the ventilator), in view of diagnosis of acute respiratory failure. The day after admission: Arterial Blood Gas Analysis was normal (pH-7.42, pCO2-42, HCO3-24, pO2 -102) Total leucocyte count: 30,000. Per abdomen: no tenderness, no guarding, no rigidity. Central Nervous System: Patient conscious, obeying commands, and no sensory-motor neurodeficit. No ptosis, no nasal regurgitation. Respiratory system: few crepitations (bilateral basal) On third day we tried weaning from Synchronized intermittent mandatory ventilation (SIMV) mode to T Piece, but she had bradycardia (pulse: 40/min) and severe breathlessness and was put back on SIMV mode. There was no trigger for respiration. On SIMV, Arterial Blood Gas Analysis was normal. Total leucocyte count: 15,000Per abdomen:. no tenderness present. Central Nervous System: Normal. Respiratory system: bilateral crepitations present CT Scan of ABDOMEN: pseudocyst of pancreas with necrosis in tail of pancreas (figure 1)

CT scan of THORAX: bilateral basal segmental atelectasis with bilateral minimal pleural effusion and bilateral anterior segment of upper lobe consolidation. No thymoma (figure 2) After 2 days, with improved Arterial Blood Gas Analysis, we repeated an attempt of weaning which failed again. She was getting easily fatigued and was unable to take her own breaths. 2 days after this, we repeated another attempt of weaning which failed again. A neurophysician was consulted, who advised serum acetylcholine esterase receptor auto antibodies, which came to be 8.36 nMol/l (N = 0.25-0.4 nMol/l) She was then diagnosed as a case of myasthenia gravis in crisis with pseudocyst of pancreas with pancreatitis. Pyridostigmine with neostigmine was given, with corticosteroids. Her condition improved after 1week and we weaned her off the ventilator.

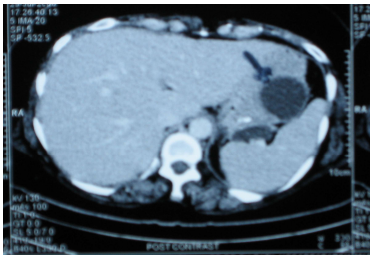

Figure 1*:* CT SCAN OF ABDOMEN - Pseudocyst of pancreas with necrosis in tail of pancreas

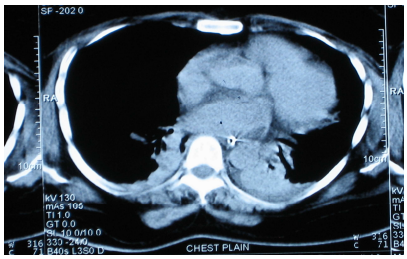

Figure 2: CT Scan of THORAX: bilateral basal segmental atelectasis with bilateral minimal pleural effusion

Discussion

Myasthenia crisis occurs when weakness from myasthenia gravis becomes severe enough to necessitate intubation for ventilatory support or airway protection. 12-16 %. Myasthenic patients may develop crisis usually within 2-3 weeks after diagnosis[1]. Possible triggers include infection[2], aspiration, physical and emotional stress and changes in medication. Crisis is most likely in patients who have previous crisis, oropharyngeal weakness or thymoma. Patients may present with respiratory failure only. Acute dysphagia with respiratory failure in myasthenia gravis has also been reported.[3] we must have an organized approach to assess and manage myasthenia crisis.

Assessment

1. Confirmation of diagnosis of myasthenia gravis.
2. Identify triggers
3. To determine whether the patient needs intubation or swallowing restrictions.

Confirmation of diagnosis of myasthenia gravis.

By levels of acetylcholineesterase auto receptor antibodies and also by ruling out other conditions which mimic myasthenia gravis like: myopathies secondary to electrolyte abnormalities, Neuropathies such as GBS-Guillain Barre Syndrome, Other Neuromuscular junction conditions, Organophosphorus toxicity, Eaton lambert syndrome, Central lesions including cervical cord or brain stem compression.

The Neurologist can advise any other investigation for further diagnosis.

Identify triggers

Infections: 30-40 % of crises are due to Upper Respiratory Tract Infections, bronchitis and bacterial pneumonia, 10% crises are due to aspiration pneumonitis, Physical stress like surgery or trauma, Change in medication may cause crisis, in 30-40%, no triggers can be identified.

Evaluation of Respiratory Function

Patient anxiety, bradycardia, tachypnoea: are early signs of respiratory failure

Criteria for intubation:

Vital Capacity < 15 ml/kg
Tidal Volume < 5ml/kg
Negative aspiratory force < 20 cm H2O
Positive expiratory force < 40 cm H2O

Evaluation for weaning begins when patients are free of crisis triggers, comfortable and objectively getting sharper on examination.

Vital Capacity >10 ml/kg
Tidal Volume > 5ml/kg
Negative aspiratory force >20 cm H2O
Positive expiratory force > 40 cm H2O

Anxiety, respiratory rate, pulse and Tidal Volume are to be frequently checked during weaning.[4] Incidence of cardiac arrhythmias are high in myasthenia crisis, so all patients should receive continuous cardiac monitoring.

Management of myasthenia crisis

1. Removing of triggers.
2. Supportive care- intubation.
3. Respirator
4. Nutrition

Weaning should be attempted as early as possible.

1. Treatment of hypokalemia, anemia
2. Acetylcholine esterase inhibitor- i.v. pyridostigmine infusion 1-2 ml/hr.
3. Plasma exchange
4. Intravenous immunoglobulins
5. Corticosteroids

Chronic pancreatitis may lead to an internal pancreatic fistula with leakage of pancreatic secretions anterior to

the peritoneal sac, forming pancreatic ascites or posterior to the retroperitoneal space, forming a pseudocyst. In our case myasthenia crisis is precipitated by pseudocyst of pancreas with acute pancreatitis. Our patient presented with acute abdomen and respiratory failure. CT Scan of abdomen and serum amylase and lipase levels confirmed acute pancreatitis with pseudocyst of pancreas. On conservative management of pancreatitis and mechanical ventilation, the patient showed a good improvement of Arterial Blood Gas Analysis and symptoms of pain in abdomen. Our inability to wean her off the ventilator using four separate attempts was unsuccessful, as she was getting increasingly fatigued even on taking breaths. This led us to suspect a neuromuscular disorder, and on consultation with a neurologist, we diagnosed her as having Myasthenia gravis after her acetylcholine esterase receptor auto antibodies were found to be significantly high. Treatment of Myasthenia gravis with pyrodistigmine and neostigmine improved her condition and serum amylase and lipase levels also returned to normal. In critically ill patients who presented with unexplained respiratory failure, myasthenia gravis should be suspected. There are no reported cases in literature of pseudocyst of pancreas and myasthenia gravis occurring together in a patient. Hence we report the case history of this patient who presented in myasthenia crisis.

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