Diaphragmatic Flutter: Dyspnea Associated with Violent Abdominal Pulsations

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Summary:
1. Diaphragmatic flutter is characterized by rhythmic involuntary contractions of the diaphragm; other respiratory muscles may be involved.
2. It is a rare cause of dyspnea. Diagnosis requires a high degree of clinical suspicion.
3. Previously, diagnosis was confirmed by fluoroscopy and/or electrophysiological studies of the diaphragm.
4. Bedside ultrasonography provides a convenient and rapid alternative method of diagnosis.
5. Phenytoin, carbamazepine, clonazepam may ameliorate symptoms. Procedures such as phrenic nerve block with bupivacaine or methylprednisolone or procedures to crush the phrenic nerve are reserved for cases refractory to pharmacologic management.

Case Presentation:
Mr. A, an 80 year old gentleman, former smoker with >110 pack years with COPD, diabetes, chronic kidney disease and aortic aneurysm status post stent presented with increasing dyspnea over the past few months. He reported recent intermittent pounding and “jerking” of his abdomen. Mr. A had persistent shortness of breath with showering, bending, and carrying heavy objects despite compliance with his inhalers (fluticasone propionate/salmeterol, tiotropium, and albuterol). Recent pulmonary function tests revealed a post-bronchodilator FEV1 of 2.26 (66% of predicted), FEV1/FVC ratio of 50% and DLCO of 16.5 (51% of predicted). He denied nausea, vomiting, abdominal pain, and chest pain. He was able to complete all activities of daily living on his own, and walked on a treadmill for 20 minutes several times a week.

Vital signs were stable with pulse oximetry of 92% on room air. Examination revealed right lower lobe basilar inspiratory crackles. During the examination, intermittent violent abdominal pulsations lasting seconds, without associated tachycardia, were observed. Bedside ultrasound revealed rapid fluttering of the hemi-diaphragms corresponding to the abdominal pulsations.

Discussion:
Diaphragmatic flutter is also known as diaphragmatic myoclonus, van Leeuwenhoek’s disease, or Belly dancer’s syndrome. It is characterized by rhythmic involuntary contractions of the diaphragm and other respiratory muscles innervated by the cervical nerve roots. The first recorded case was that of Anton van Leeuwenhoek, “father of the microscope”.

From 1723 to the end of the 20th century, 48 documented cases were reported.

In the past, in the setting of characteristic signs and symptoms, diagnosis was confirmed with fluoroscopy or electrophysiological studies of the diaphragm. Our case highlights the ease and utility with which bedside ultrasound can confirm this diagnosis. In this case report, bedside ultrasonography was performed at the time of the visit and revealed fluttering of the diaphragm corresponding to the abdominal pulsations observed clinically.

Diaphragmatic flutter is a rare diagnosis. There are no randomized studies addressing treatment options for this disorder. Reported treatment options include pharmacological treatment with phenytoin, carbamazepine, clonazepam and procedures such as phrenic nerve block with bupivacaine or methylprednisolone or procedures to crush the phrenic nerve. Phrenic nerve crush has been reported to provide symptom relief lasting up to 6 months.

Mr. A was started on clonazepam 0.5 mg three times a day. There was improvement in both the severity and frequency of the myoclonic episodes and corresponding dyspnea. Due to increasing fatigue, unsteadiness and orthostatic hypotension, Clonazepam was decreased to 0.5 mg daily and other antihypertensive medications were also reduced. Symptoms of orthostasis resolved with this intervention. Myoclonus symptoms remained substantially decreased and although not completely resolved, no longer concerned the patient.

References: