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Diaphragmatic Flutter Presenting as Inspiratory Stridor*

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Diaphragmatic flutter is a rarely reported disorder in which the diaphragm involuntarily contracts at a rapid rate. We report a unique case in which diaphragmatic flutter was associated with inspiratory stridor and was severely disabling. A new approach to the treatment of this condition, phrenic nerve crush, provided an optimal outcome, with resolution of symptoms and the return of normal diaphragmatic function. Pathophysiology and treatment of this condition are discussed.

(Chest 1995; 107:872-75)

Key words: diaphragm; flutter; hiccups; myoclonus; nerve crush; phrenic nerve; respiratory dysfunction; singultus; stridor; vocal cord dysfunction

Diaphragmatic flutter (myoclonus) is a rare disorder in which involuntary contractions of the diaphragm occur one to eight times per second. It was first described in 1723 by Leeuwenhoek,[2] the inventor of the microscope who was personally afflicted with this disorder. Since then, with the exception of a review of 42 cases,[3] this disorder has been rarely reported. We present a unique case in which the rapid myoclonic activity of the diaphragm presented as inspiratory stridor.

Case Report

A 13-year-old white girl developed a frequent, severe cough during an upper respiratory tract infection. She was hospitalized for bronchitis and cough-variant asthma and was treated with frequent nebulized β-agonists, intravenous aminophylline, and corticosteroids. Despite this therapy, her condition failed to improve and she proceeded to develop loud inspiratory stridor preceding her cough. The cough and associated stridor became more frequent. Then, she abruptly developed an unusual breathing pattern characterized by tachypnea and inspiratory stridor. This breathing pattern was relentless while awake, resolved during sleep, and was made worse by anxiety. She was dyspneic and limited in her speech and physical activity. Several diagnoses were considered, including a seizure disorder, stress conversion disorder, Tourette's syndrome and vocal cord dysfunction. In an effort to treat these conditions, lorazepam (Ativan), clonazepam, carbamazepine (Tegretol), diazepam (Valium), fluoxetine (Prozac), clonidine (Catapres) patches, and biofeedback were all tried, with no improvement. She was then referred to our institution for further evaluation.

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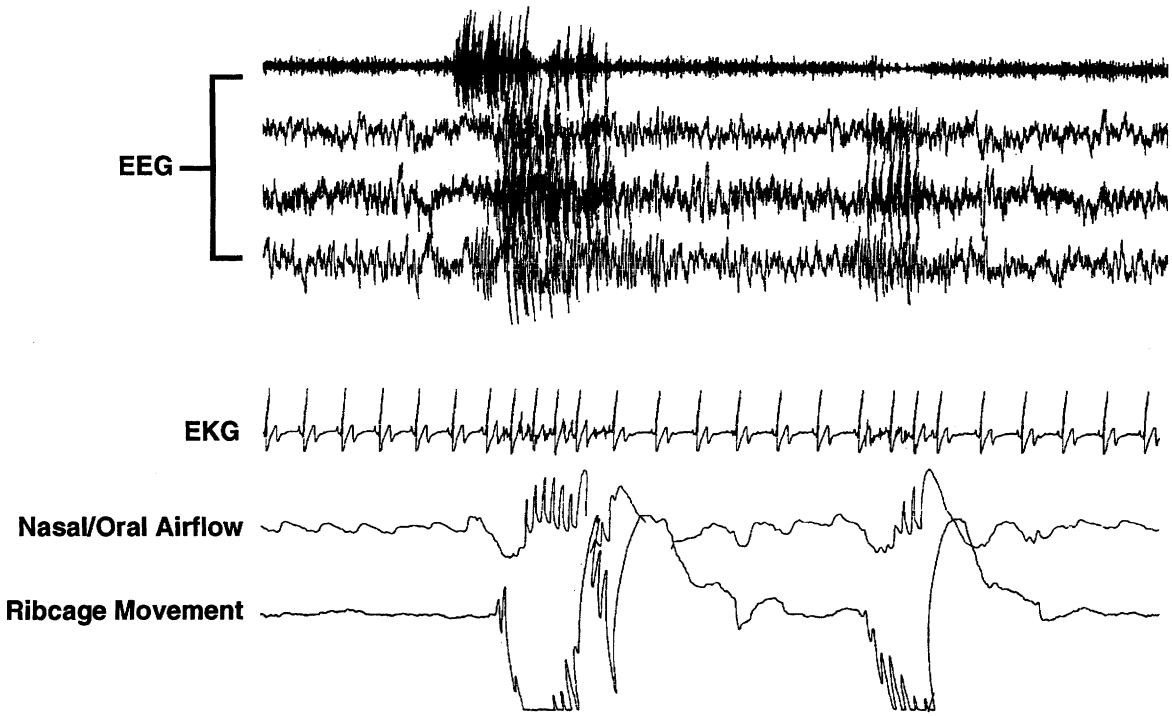

FIGURE 1. This segment of our patient's sleep study shows a normal breathing pattern during sleep, except for two periods of increased EEG activity lasting several seconds. During these brief arousals, the patient did not awaken, but the return of her abnormal breathing pattern is evident in the simultaneous recordings of nasal/oral airflow and ribcage movement.

Her medical history was remarkable for a diving accident that occurred 16 months prior to the above events, when she hit the right side of her head on the edge of a pool. While she did not suffer a concussion, skull fracture, or spinal injury, she had neck, back, and shoulder muscle spasms, and required a Philadelphia collar for 6 weeks.

On physical examination, her breathing was audible as a rapid, stridulous panting sound. Her respiratory rate was 140 to 160 breaths/min in a cycle of 13 to 16 shallow inspirations followed by one normal expiration. Her neck had decreased range of motion to the left. She occasionally exhibited facial tics. Results of the remainder of her examination were unremarkable. An arterial blood gas value was consistent with hyperventilation (pH 7.65/$PaCO_2$ 19 mm Hg/PaO_2 100 mm Hg/HCO_3 21 mEq/L), while results of other blood tests (including CBC, erythrocyte sedimentation rate, and biochemistries) were normal. Fiberoptic laryngoscopy revealed abnormal closure of the true and false vocal cords occurring multiple times during inspiration, but in a manner inconsistent with vocal cord dysfunction, which typically involves only the true vocal cords. A sleep study confirmed the history of normalization of her abnormal breathing pattern during sleep. This study also revealed rare periods during sleep of electroencephalographic activity, lasting only several seconds, when her abnormal breathing pattern would return (Fig 1). This indicated the involuntary nature of her disorder. The EEG, computed tomographic and magnetic resonance imaging scans of her head, and radiographs of her cervical spine, thoracic spine, and chest were all normal. An ECG provided evidence for *noncardiac* electrical activity occurring during inspiration, in discrete bursts at a rate of about 200/min (Fig 2, top), and disappearing during expiration (Fig 2, bottom). Presuming that this electromyographic activity may be emanating from the diaphragm, fluoroscopy of her diaphragm was performed that revealed rapid myoclonic contractions of the left hemidiaphragm, primarily on inspiration, and superimposed on normal diaphragmatic excursion.

To eliminate the possibility of a focal lesion that may explain our patient's diaphragmatic dysfunction (*eg*, tumor or enlarged lymph node pressing on the left phrenic nerve), a chest computed tomographic scan was added to the previous imaging studies of her head and neck, and was normal. We then considered all medications previously reported to be useful in treating diaphragmatic flutter. Many of these medications (listed above) had already been tried in our patient as therapy for other suspected conditions and were unsuccessful in improving her condition. At our institution prior to the diagnosis of diaphragmatic flutter and in an effort to treat Tourette's syndrome, valproic acid (Depakene), haloperidol (Haldol), pimozide, and clonidine patches were tried, but were also not helpful.

Having exhausted all previously reported pharmacologic therapies, we investigated the effect of direct anesthesia of the left phrenic nerve. Local infiltration under fluoroscopic guidance of the left C4 root with bupivacaine (Marcaine) resulted in paresis of her left hemidiaphragm and complete, although temporary, resolution of her abnormal breathing pattern. We proceeded with repeated local nerve blocks over a 2-week period to see if this might lead to sustained resolution of her symptoms. Such therapy has had reported success in the treatment of refractory hiccups.[4] We also injected methylprednisolone (Solu-Medrol) at the left C4 root, hypothesizing that local inflammation due to her diving accident was responsible for phrenic nerve irritation that would cause diaphragmatic flutter. Neither of these solutions resulted in

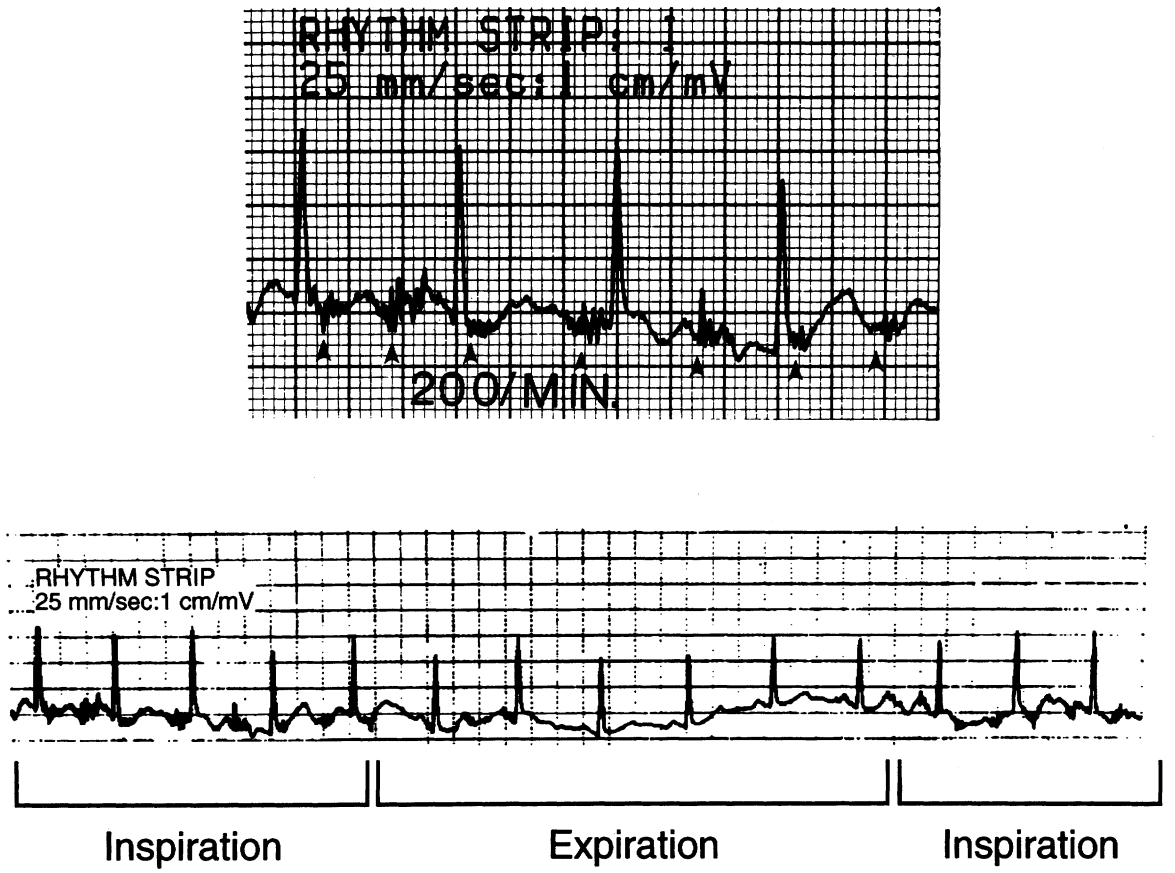

FIGURE 2. *Top*, ECG rhythm strip (lead II) shows discrete bursts of noncardiac electromyographic activity, identified by arrowheads, at a rate of about 200/min. *Bottom*, ECG rhythm strip (lead II) shows variation in the abnormal electromyographic activity with the respiratory cycle.

a more sustained resolution of her condition. Finally, we performed a left phrenic nerve crush, which usually allows for the return of diaphragm function within 6 months.[5] This resulted in complete resolution of her symptoms. After this operation, results of pulse oximetry and pulmonary function tests, both at rest and during exercise, were all normal. Fluoroscopy 2 months after the phrenic nerve crush showed the return of normal left hemidiaphragm excursion with respiration, and 1 year postoperatively, our patient continues to be asymptomatic.

DISCUSSION

Diaphragmatic flutter is a rarely reported disorder characterized by rapid, involuntary contractions of the diaphragm, often superimposed on normal diaphragmatic excursion. It is considered to be a form of myoclonus, which has been defined as repetitive, involuntary contractions of a muscle or group of muscles, usually superimposed on voluntary contractions. Diaphragmatic flutter is usually episodic in nature, but in our patient, it was continuous for nearly 8 months. The most common presenting complaints are shortness of breath and epigastric pulsations, fatigue, and/or pain of the abdominal wall muscles. In a review of 42 cases, Rigatto and De Medeiros[3] found that two thirds of the patients had bilateral involvement of the diaphragm. The unilateral cases usually involved the left hemidiaphragm. This left-sided predominance has also been noted in singultus or hiccups.[6] Their review also found that the rate of contractions averaged 150/min, but ranged from 35 to 480. In only 6 of 43 cases was there tachypnea at the same rate as the flutter. Only two patients had documentation of respiratory alkalosis, as was seen in our patient.

Perhaps the most unusual aspect of our patient's presentation was her "stridor" with each inspiration. To our knowledge, this is the first reported case of diaphragmatic flutter associated with stridor. We suspect that the audible sound in our patient was related to compensatory closure of the glottis in response to the rapid and forceful contractions of the diaphragm, as is seen in singultus or hiccups.[6] Laryngoscopy findings and resolution of her stridor with left phrenic nerve anesthetic blockade and crush support this conclusion.

Possible etiologies of this disorder include abnormal excitation of the phrenic nerve caused by disturbances of the central nervous system, irritation of the nerve itself, or irritation of the diaphragm. The most common central nervous system disturbance is encephalitis.[3] Irritation of the phrenic nerve may occur anywhere along its path; however, irritation of the left phrenic nerve by the heart, resulting in synchronous flutter with cardiac systole, is most common.[3] Other causes include mass lesions impinging on the phrenic nerve or trauma to the cervical root of the phrenic nerve, which may have occurred in our patient during her diving accident. Pleurisy, peritonitis, or ischemia of the diaphragm have also been reported causes of diaphragmatic flutter.[3] Metabolic disturbances may be important cofactors to any of the above causes.[7] Where the

cause is not clear, hysteria or psychogenic causes have been implicated.[8] However, many of the reported cases of diaphragmatic flutter were initially misdiagnosed as hysterical conversion disorders, perhaps because it is episodic, it resolves during sleep, and has a tendency to worsen with anxiety or exertion.

Multiple pharmacologic trials in our patient, including many medications anecdotally reported to be successful in treating diaphragmatic flutter, failed. Because the myoclonic activity of the diaphragm was unilateral, and because anesthetic blockade of the C4 root provided complete although temporary relief, we decided to crush the left phrenic nerve. This procedure, once used for the treatment of tuberculosis[9] and occasionally used in the treatment of singultus,[10] was chosen because return of diaphragmatic function due to regeneration of the phrenic nerve usually occurs within 6 months.[5] Phrenic nerve crush is often associated with a decrease in lung volumes;[11] however, our patient's lung volumes and function during exercise after left phrenic nerve crush were normal. We attribute this maintenance of lung function to the preservation of left hemidiaphragmatic tone, demonstrated by the lack of paradoxic diaphragmatic movement seen on fluoroscopy postoperatively. Two months later, our patient had recovered normal function of her left hemidiaphragm while remaining asymptomatic.

In conclusion, an abnormal stridulous disorder may actually be a manifestation of a diaphragmatic abnormality such as diaphragmatic flutter. In patients with hemidiaphragmatic flutter unresponsive to medication, phrenic nerve crush may provide an optimal outcome. Phrenic nerve anesthetic blockade may identify those patients with diaphragmatic flutter most likely to benefit from phrenic nerve crush.

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Self-injection With Olive Oil*

A Cause of Lipoid Pneumonia

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A 48-year-old man with unipolar depression and a psychosexual problem concerning his body image was injecting his scrotum repeatedly with olive oil to increase the size of his genitals. He developed respiratory failure following accidental intravenous injection of olive oil and was found to have lipogranulomatous lesions in the lung and the scrotum.

(Chest 1995; 107:875-76)

Key words: genital self-injection; lipoid pneumonia; olive oil

Exogenous lipoid pneumonia is usually due to aspiration of lipids or rarely because of intravenous use of oil-based dyes for diagnostic radiologic procedures.[1] Although a clinical rarity, exogenous lipoid pneumonia has aroused interest because radiologically it can mimic bronchogenic carcinoma.[1,2] Further associations of excessive use of lip gloss,[3] chapstick,[4] and Jamaican tobacco[5] with lipoid pneumonia have helped maintain interest. We report a case of lipoid pneumonia in a man who had been frequently injecting his scrotum with olive oil; he presented following an accidental intravenous injection.

Case Report

A 48-year-old man was transferred to our hospital in a stuporous state. The patient was found unconscious and pulseless (blood pressure 60/40 mm Hg) with a 22-gauge needle stuck in his scrotum and a syringe lying nearby. In the local hospital, he received intravenous saline solution and 60 U of insulin since his serum glucose level was 16 mmol/L. With this, although his blood pressure and serum glucose level (4.6 mmol/L) returned to normal, there was no improvement in his level of consciousness.

The patient had undergone a vasectomy 8 years earlier and a circumcision necessitated by phimosis 2 years before. At the time of circumcision, the surgeon observed a firm swelling in the upper scrotum; a biopsy specimen was taken that revealed lipogranulomata. This had upset the patient and he had signed himself out

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