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Acute respiratory failure induced by belly dancer's syndrome: A glance to a rare case report

Ruth Elizabeth Abraham[1], Gan Lay Min[1], Ahmad Luqman Bin Md Pauzi[2]*, Noor Hafizah Abdul Salim[2], Iskasymar Ismail[2]

[1]Department of Medicine, Newcastle University Medicine Malaysia, [2]Department of Medicine, Faculty of Medicine and Health Sciences, University Putra Malaysia, Selangor, Malaysia
**Corresponding author*

Abstract:

Respiratory myoclonus, also known as belly dancer's dyskinesia (BDD), is a rare manifestation of movement disorder characterized by repetitive choreiform involuntary movements involving the anterior abdominal muscles, the diaphragm, and other respiratory muscles. Currently, there is no definite pathophysiology that clearly explains this condition. A 25-year-old male with a known case of BDD presented with an exacerbation of involuntary and continuous writhing movements of the abdominal wall muscles associated with abdominal pain and shortness of breath over the past 2 days. Subsequently, he was intubated due to worsening respiratory distress a few days after his admission. He was then put on ultrasound-guided botulinum toxin A injections of 25 units over the left hemidiaphragm regularly. His symptoms markedly improved since then as the attacks had reduced to 5–6 monthly intervals. Administration of ultrasound-guided botulinum toxin A injections may help to control the exacerbation of BDD and might be an option for cases refractory to medical treatment and phrenic nerve ablation.

Keywords:

Abdominal wall dyskinesia, belly dancer's dyskinesia, diaphragmatic flutter, respiratory myoclonus

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ORCID:
REA: 0000-0002-9204-1269
GLM: 0000-0003-2054-4104
ALBP: 0000-0003-4086-9248
NHAS: 0000-0002-3057-4852
II: 0000-0002-8892-1378

Address for correspondence:
Dr. Ahmad Luqman Bin Md Pauzi, Department of Medicine, Faculty of Medicine and Health Sciences, University Putra Malaysia, 43400 Serdang, Selangor, Malaysia.
E-mail: ahmadluqman@upm.edu.my

Introduction

Respiratory myoclonus, also known as belly dancer's dyskinesia (BDD), is a movement disorder characterized by repetitive choreiform involuntary movements involving the anterior abdominal muscles, the diaphragm, and other respiratory muscles innervated by cervical nerve roots.[1] This syndrome is an extremely rare disorder with an incidence and prevalence of <8.6 cases per 100,000 people.[2]

There are various cases reporting a wide variety of underlying etiologies. However, there is no exact underlying pathophysiology, thus making the diagnosis and management of BDD extremely challenging.[3] The causes of belly dancer's syndrome vary including disruptions within the central or peripheral nervous systems, anxiety, nutritional imbalances, and administration of certain pharmaceutical agents or drugs.[3] The prognosis of this condition is uncertain as so far no specific management has been recommended. In this report, we present a case of a young male patient who presented with respiratory failure during an acute attack of respiratory myoclonus and some treatment options to manage this condition.

Case Report

The patient was a 25-year-old male who presented to the emergency department

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with a complaint of increasing, involuntary, and continuous writhing movements of the abdominal wall muscles over the past 2 days. This was associated with generalized abdominal pain, shortness of breath, fatigue, and sleep disturbances as the muscle contractions failed to cease even while he was asleep. He denied symptoms suggestive of respiratory infections such as fever or cough. He also had no chest pain, orthopnea, or bilateral leg swelling to suggest it could be cardiac in origin. He was able to carry out his daily living activities as usual unless the intensity of the pain increased. On examination, the patient was observed to have continuous rapid movements of the anterior abdominal muscles in both supine and sitting positions. Vitals were stable, and oxygen saturation was 95% on room air. Apart from the abnormal movement, other systemic examination was normal. The chest X-ray [Figure 1] reveals an elevated right hemidiaphragm. The electrocardiogram and blood investigations were all within normal range. He was given a nasal prong 3 L/min, intramuscular (IM) pethidine 50 mg for the pain, and intravenous (IV) diazepam 10 mg administered to control the abdominal movement. However, the movements failed to subside, and he was subsequently admitted to the cardiothoracic ward for observation.

The patient was first diagnosed with BDD in 2014 when he presented multiple times with rapid movements of the abdominal wall, abdominal pain, and shortness of breath. He reported no known triggers or prodromes. A fluoroscopy revealed bilateral diaphragmatic flutters. Since then, he had been on several treatments with a combination of multiple anti-epileptic drugs which all failed to control the symptoms. He subsequently underwent a right phrenic nerve neurectomy in 2017. A fluoroscopy postprocedure showed right hemidiaphragm paralysis as no movement of the right hemidiaphragm was noted during the study [Video 1].

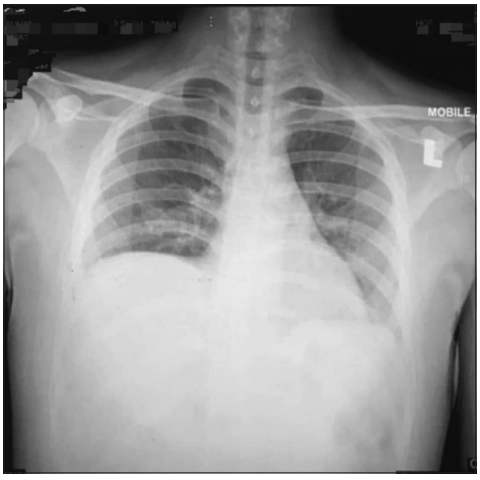

Figure 1: The chest X-ray reveals elevated right hemidiaphragm postright phrenic nerve ablation. There was no evidence of consolidation or edema in the lungs

However, the symptoms were still not controlled, and he had multiple recurrences of diaphragmatic flutters that caused him to be admitted to the hospital for a total of 14 times due to similar problems.

On day 2 of admission, the patient developed worsening respiratory distress, which required IV diazepam and IM pethidine to control the symptoms. His oxygen saturation was 96% on a nasal prong 3 L/min with a normal cardiorespiratory examination. His abdomen appeared to have rapid irregular movements that made him appear tachypneic. He was put on noninvasive ventilation to control his respiration. In the next few days, the patient was noted to be lethargic and had an ineffective breathing pattern. His blood gas showed type 2 respiratory failure with a pH of 7.21, PCO_2 of 65 mmHg, PO_2 of 180 mmHg, and bicarbonate of 18 mmol/L. He was electively intubated and was on IV midazolam infusion and promethazine. His abdominal flutters subsequently subsided. He was extubated 2 days later and was discharged 17 days after admission. Since discharge, the patient had undergone multiple treatments with ultrasound-guided botulinum toxin A injections of 25 units over the left hemidiaphragm at the rectus abdominis muscle. His symptoms markedly improved since then as the attack had reduced to 5–6 monthly intervals. Consent for this publication was obtained from the patient.

Discussion

We report a case of a patient with underlying BDD who presented with frequent attacks of diaphragmatic flutter despite multiple treatments given to the patient. As presented in this patient, this type of dyskinesia commonly presents with involuntary, visible, or nonvisible, undulating movements of the epigastrium or the abdominal wall.[3,4] This movement can disrupt the spontaneous breathing of the patient; however, oxygen saturation usually remains within the normal range.[5] Interestingly, during this recent attack, the patient developed respiratory distress and further deteriorated, which required advanced airway management and intubation.

There are two mechanisms that can explain the production of diaphragmatic flutter. The first mechanism is due to the abnormal excitation of the phrenic nerve, either by disturbances of the central nervous system or by the presence of irritating factors anywhere along the phrenic pathway. The second mechanism is due to irritation of the diaphragm itself.[4] In more than 50% of cases reported in the literature, the area of the diaphragm affected in BDD was noted to be more on both hemidiaphragm. This is evidenced by 15 cases affecting both hemidiaphragm compared to 9 cases involving only 1 hemidiaphragm, where predominantly

the left hemidiaphragm is more affected than the right hemidiaphragm.[4]

During an exacerbation of diaphragmatic flutter, the normal breathing of the patient will usually be affected. There are a few mechanisms to explain why BDD can affect normal respiration and possibly lead to respiratory failure. This is possibly due to the three types of breathing patterns that happen during the diaphragmatic flutter. The first breathing pattern is tachypnea without any disruption of breathing patterns. The second pattern is dysrhythmic breathing superimposed with high-frequency waves, and the third breathing pattern is diaphragmatic flutter possibly associated with apneustic respiration. This apneustic pattern has no clear apneustic period; however, the original breathing rhythm seems to have lost its regularity.[3-6] The patient in our case developed the third pattern of dysrhythmic breathing, which led to ineffective breathing and deterioration of respiration. It is speculated that during an episode of diaphragmatic flutter, the repetitive stimulation of vagal stretch receptors or chest wall proprioceptors by the flutter suppresses the reflexogenic spontaneous respiratory rhythm, and the respiratory muscles start to synchronize with the diaphragmatic flutter.[6]

There are a few studies in the literature that show that diaphragmatic flutter may have a possible effect on respiratory functions. Katz *et al*.[7] reported three neonates who developed flutter of the diaphragm soon after delivery, which later progressed to respiratory failure and required nasal continuous positive airway pressure immediately to normalize the breathing pattern. These infants were given pharmacological therapy with chlorpromazine. This caused the termination of the respiratory flutter, which permitted weaning of ventilatory support within a few hours. There were also several cases of patients who developed diaphragmatic flutter after undergoing a coronary artery bypass graft. There were difficulties in weaning these patients off the mechanical ventilation as the flutter had affected their respiratory functions.[8] Apart from that, BDD can also cause respiratory distress due to acute stridor. This stridor is suspected due to the audible sound related to compensatory closure of the glottis in response to the rapid and forceful contractions of the diaphragm.[9]

At present, there are no proper guidelines detailing the management options of BDD. Thus, it always relies on professional opinions. Diazepam has been shown to control the symptoms of BDD. However, in this case, this treatment has not managed to completely control the symptoms.[3] The treatment options for those with BDD who do not respond to pharmacotherapy include phrenic nerve block or a complete transaction of the phrenic nerve on the affected side. This treatment is effective but associated with a recurrence of symptoms and paralysis of the hemidiaphragm.[3] In our patient, transection of the right phrenic nerve caused a temporary cessation of his symptoms, but this was not a permanent solution as the flutter, then started in the left hemidiaphragm causing a return of his symptoms. The other option is ultrasound-guided botulinum toxin A injections. The botulinum toxin A mediates its effects by inhibition of acetylcholine release at the presynaptic nerve endings of the motor endplates and can successfully cause a complete cessation of dystonia.[10] This is similar with our case where the patient's condition improved with the cessation of his symptoms using botulinum toxin A injections. He was planned for regular injections of botulinum toxin A during clinic follow-ups and has showed a good response as it reduced the frequency of his exacerbations of the diaphragmatic flutter.

Conclusion

Belly dancer's syndrome is a rare condition that may affect respiratory function. The exact mechanism is still unknown, thus making it refractory to medical therapy. The administration of ultrasound-guided botulinum toxin A injections may help control the exacerbation of BDD and might be an option for cases refractory to medical treatment and phrenic nerve ablation.

Authors' contributions

AL provided the concept of manuscript. RE, GH, AL, NH and II contributed to the concept of manuscript and wrote the paper.

AL provided the setting according to CARE Guidelines. All Authors Contributed to this manuscript by editing, revisioning and taking care of references.

Conflicts of interest

None Declared.

Consent to participate

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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