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Case Report
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Sleep Disorders in a Patient with Asternia: A Case Study

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Abstract

Introduction: Asternia, also known as complete congenital sternal cleft, is a rare chest wall abnormality caused by a failure in sternal fusion during embryonic development. The role of this disease and its complications on the patient's sleep is not clear yet.

Case Report: The patient was a 20-year-old female with a history of congenital sternum cleft referred to the Sleep Clinic of Rasoul Akram Hospital in Tehran for a split-night polysomnography test due to daytime sleepiness, morning headache, and insomnia.

Conclusion: This case report aimed to investigate the parameters and sleep disorders in a patient with asternia. Considering the complications of this rare disease, it is essential to pay more attention to sleep disorders, especially sleep-related breathing disorders, in these patients.

Keywords: Asternia; Sleep disorder; Obstructive sleep apnea; Iran

Introduction

Asternia or complete sternal cleft is a very unusual chest wall deformity caused by a sternal fusion deficiency during embryonic development. This anomaly can cause serious consequences and, like other congenital anomalies, can be linked to other birth disorders [1].

Asternia carries several risks, including mediastinal injury, hypothermia, and insensible fluid losses. Also, respiratory and dynamic complications may occur. Paradoxical breathing pattern can lead to cyanosis and recurrent chest infections due to impaired gas exchange and inadequate secretion clearance. In addition, changes due to intrathoracic pressure can decrease venous return [2].

Another sternal cleft abnormality includes Pentalogy of Cantrell (POC), which involves abnormalities of the sternum, pericardium, heart, diaphragm, and abdominal wall. Seemingly unrelated processes such as PHACE (posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies, and eye anomalies) syndrome can also be present [3].

The optimal time to conduct sternal cleft repair is during infancy, when the chest wall is extremely malleable and can therefore be initially closed without considerable cardiac constraint. As the patient ages and the chest wall stiffens, closure might become progressively more challenging as venous return and lung flexibility are reduced [4].

Given that respiratory disorders induced by defects in the chest wall muscles during sleep are likely to worsen and no study has been conducted on sleep disorders in patients with sternal defects, this study aimed to evaluate sleep parameters in patients suffering from this rare disease.

Case Presentation

The patient was a 20-year-old female with a history of congenital sternum cleft and repair in childhood. The patient referred to a pulmonologist and complained of daytime shortness of breath. The patient was referred to the Sleep Clinic of Rasoul Akram Hospital in Tehran, Iran to be assessed for obstructive sleep apnea (OSA). All necessary tests were performed and paraclinical parameters, including spirometry were normal.

According to the patient's records, she complained of fatigue, excessive daytime sleepiness, and morning headaches. The patient did not report snoring or breathing problems during sleep and complained of frequent inability to sustain sleep, which raised the possibility of OSA. The patient got into bed at 12 AM, fell asleep in about half an hour, and woke up at 10 AM in the morning. She did not report such symptoms and disorders as restless legs syndrome, sleep paralysis, cataplexy, hypnagogic or hypnopompic hallucinations, sleepwalking, nightmares, and other parasomnias.

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TST (Total Sleep Time) = 380 min
SE (Sleep Efficacy) = 85% WITH 3 sleep cycles
Sleep Latency = 16 min
REM Latency = 141 min
PLM index = 2/hr sleep
Average Heart Rate = 57 bpm
Without SPO2<90%
AHI = 7/hr sleep
Duration of Sleep Stage: REM = 27 min (7%)/N1 = 19 min (5%)/N2 = 207 min (54%)/N3 = 127 min (34%)

Table 1: Polysomnography.

There was no history of psychiatric disorder, suicidal ideation, delirium, hallucinations, or medical conditions. In her childhood, she had sternal reconstructive surgery and tonsillectomy. Also, there was no history of using medications, smoking cigarettes, using drugs, or a family history of sleep or psychiatric disorders.

The patient's blood pressure was normal (80/110), Body Mass Index (BMI) was 18 kg/m^2, neck circumference was 34 cm, the distance between the thyroid cartilage and the chin was 9 cm, and Mallampati score was 2.

Her scores in the Restless Legs Syndrome Screening Questionnaire, STOP-BANG Questionnaire, Insomnia Intensity Index, Epworth Sleepiness Score, and Pittsburgh Sleep Quality Index were zero, one, nine, four, and three, respectively (Table 1).

In polysomnography, various sleep parameters were assessed. The sleep parameters, including total sleep time, sleep latency, REM latency (the time between the onset of sleep and the first stage of REM sleep), and Periodic Leg Movement index (PLMS; average frequent movements of the lower limbs in one hour of sleep) were normal. Heart rate was also normal and no snoring was reported. The number of sleep cycles, including both the REM and non-REM sleep, was also normal for the age of the patient. However, sleep efficacy (the ratio of a person's total sleep time to bed time) decreased slightly. AHI (the average of apnea and hypopnea per hour of sleep) was equal to seven, which is classified as mild OSA syndrome.

Discussion

The present study examined sleep parameters in a patient with asternia, which is an extremely rare condition that can develop alone or in conjunction with other malformations of abdominal wall, diaphragm, pericardium, or heart. The baby will have the highest chance of survival if diagnosed and treated early [5]. The prevalence of asternia has been reported as one per 100,000 live births [6].

In fetal life, the sternum develops from the mesoderm's outer surface. In the tenth week, the cells from the two mesoderm bands on either side of the anterior chest wall migrate to the midline and fuse together to form the sternum. Primordia between the ventral ends of the growing clavicle produce the manubrium. The sternum columns fuse in a cephalocaudal manner. The sternal columns do not always unite in the middle, resulting in a complete sternal cleft [7]. The etiology of the sternal cleft is unknown.

External clefts can be classified into three main groups: (A) sternal cleft without associated abnormalities; (B) thoracic or true ectopia cordis with varying degrees of sternal cleft where the heart is outside the thoracic wall; and (C) thoracoabdominal ectopia cordis, also known as POC [8].

In her childhood, the patient under study had her congenital sternal cleft corrected by posterior sternal wall plus chondral grafts plus total correction surgery.

During infancy and youth, the chest wall and upper airway alter to accommodate the growing organism's physiological needs. Compression is possible in the newborn's chest wall, which aids in the removal of pulmonary fluid. The adaptation rate of the chest wall in infants is three times that of the lung [9]. This causes paradoxical movement of the chest inward, especially during Rapid Eye Movement (REM) sleep, which is associated with increased respiratory activity and decreased intercostal muscle activity during inhalation. The sternum and vertebrae ossify in the womb and continue until the age of 25, resulting in chest wall rigidity.

In our patient, the look of the chest was unaffected and there were no paradoxical chest motions. Children with upper airway obstruction sleep with more inconsistency compared to adults. Normal adolescents do not experience paradoxical internal chest movement during inhaling. The form of chest might also change during childhood. The ribs are placed horizontally in babies, resulting in a circular chest with limited expansion potential. As a result, at one month of age, the share of chest breathing current in non-REM sleep is just one-third of that in adults [10].

Tomohisa Nagasao et al. concluded that when the full length of the sternum is missing, chest breathing is substantially hampered, and that correction of the defect is required to treat the disorder. However, since the upper half of the sternum is more crucial for respiration, the upper part should be prioritized in reconstruction [11].

There are some limited studies on the respiratory disorders associated with sternal cleft. However, no studies have been carried out on sleep disorders in patients with asternia so far. Although some respiratory illnesses, such as sleep apnea, exclusively occur during sleep, practically all respiratory disorders, including upper airway obstruction, central hyperventilation, and chronic lung disease, get worse during sleep than wakefulness. Thus, the impact of sleep on breathing is a crucial concern because when we are awake, we breathe more deeply than sleeping; also, ventilation is reduced during sleep [12].

Respiratory drive is reduced and breathing becomes irregular, especially during the REM sleep. The respiratory rate and current volume are variable and are associated with recurrent central apnea. REM sleep is also associated with decreased intercostal and upper airway muscle tone. Therefore, breathing is impaired during sleep compared to wakefulness, and REM is more impaired during sleep. This is especially important in children, as they sleep more than adults and have relatively more REM sleep.

The patient under study was diagnosed as having normal sleep parameters, except for sleep efficacy and Apnea-Hypopnea Index (AHI). The patient's AHI was indicative of mild OSA [13].

OSA is the most common sleep disorder characterized by recurrent collapse of the soft tissue in the rear of the throat during sleep. According to the latest research, its prevalence is 17% in women and 35% in men [14]. These intermittent collapses reduce airflow, which lead to two consequences: Arousal in sleep to resume airflow, and gas exchange disorders caused by hypopnea (decreased breathing) or apnea (stopped breathing) [15]. Frequent arousals in OSA lead to neurological cognitive consequences, such as decreased memory, daytime drowsiness, and decreased quality of life. However, the mechanisms contributing to these adverse outcomes are complex and not yet fully understood.

Also, Mallampati score was equal to 2 in our patient. This has been identified as an independent risk factor for OSA in previous studies, so that for each degree of increase in Mallampati score, the chance of OSA doubles [17]; this should be considered in the analysis of results.

Conclusion

This case report evaluated the parameters and sleep disorders in a patient with asternia. Considering the complications of this rare disease, it is essential to pay more attention to sleep disorders, especially sleep-related breathing disorders, in these patients. Further studies are needed to investigate the chest anatomy defects causing OSA syndrome in patients with congenital sternum cleft.

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