Intractable singultus: a case report

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ABSTRACT

Background: Intractable singultus is a rare disorder, and there are no guidelines for its management. More than 100 etiologies are described in the literature, but the etiology cannot be defined in as much as 20% of patients. Acute singultus needs no further treatment, but intractable singultus warrants thorough evaluation and treatment because it has high morbidity and mortality.

Case Presentation: We present a 29-year-old male with intractable singultus accompanied by a sliding hiatal hernia, refractory to chlorpromazine and bacoifen treatment, and hiatal hernia repair. The patient underwent hiatal hernia repair and Nissen fundoplication, which provided 3 days free of singultus. On the fourth postoperative day, the singultus begins 3 times a day, with episodes lasting up to 5 minutes, with a rate of 10-25 singultus per minute. The patient was discharged on postoperative day-5, hemodynamic was stable and no signs of infection were reported.

Conclusion: Symptomatic treatment for intractable singultus is often unsuccessful, with serious side effects that may impair the patient’s quality of life. The cornerstone of the management of intractable singultus is finding the etiology.

Keywords: Intractable Singultus, Hiatal Hernia, Case Report.


INTRODUCTION

Sliding hiatal hernia is a stomach protrusion and sliding up of Gastroesophageal Junction (GEJ) through the esophageal hiatus.1 Most cases are asymptomatic but may result in Gastroesophageal Reflux Disease (GERD).1 Singultus or hiccups is an involuntary contraction of the inspiratory muscle, followed by the abrupt closure of the glottis, which produces the characteristic “hic” sound.2 This reflex is mediated by the phrenic nerve, vagus nerve, and reflex center in the brainstem. They can occur in adults, children, and infants.3 Often, these episodes are transient and resolved within 48 in acute hiccups, persistent lasting over 2 days. Intractable lasts over 2 months that have a significant impact on quality of life by interfering with eating, sleeping, speaking, and social activities, and can be a harbinger of severe medical pathology.3,4

The causes of singultus or hiccups have been traced to the central nervous system (CNS), the neck, thorax, and belly, namely along the phrenic and vagus nerve pathways. Hiccups are most commonly caused by CNS lesions or irritation of the phrenic and vagus nerves.5-7 Trauma, infections, inflammations, chemicals, vascular illness, demyelinating disease, arteriovenous malformations, tumors, and hysteria have all been identified as causal factors in several studies.5-7 Hiccups can also be caused by medications, blood disorders, and cardiac pacemakers.8-10

Based on those mentioned above, this case study aims to evaluate the current management of intractable singultus accompanied by a sliding hiatal hernia as well as the follow-up evaluation during the study period.

CASE PRESENTATION

A 29-year-old male with a history of intractable singultus, worsening in the last 6 months. Initially, the singultus was for intermittent onset, gradually they became more consistent, occurring daily with a frequency of about 30 per day. The singultus worsened over time and the patient developed GERD, possibly due to the ongoing singultus. Since the last 2 months, the singultus has become non-stop, occurring every 5 seconds and continuing during sleep. There were no symptoms of heartburn and regurgitation. It causes severe distress to the patient who became progressively anorexic (losing weight 6 kg in a month), irritated, short-tempered, fatigued, depressed, hard to talk, and choking secondary to glottis spasm resulting in pneumonia aspiration. He has been prescribed chlorpromazine for the last 4 months, 50 mg every 8-12 hours daily, but ineffective, resulting in headaches, vision loss, blank facial expression, dry mouth, restlessness, and agitation, which made him unable to work. The patient underwent countless medical evaluations; physical and neurological examinations including ENT (ear, nose, and throat), chest and abdomen; laboratory test including blood urea nitrogen and transaminases; imagining including MRI of brain and neck, CT of abdomen and thorax, EEG (electroencephalography), and upper gastrointestinal endoscopy. Electrocardiogram, head and neck MRI, and EEG were within normal limit, alanine transaminase was 189 unit/L, aspartate transaminase was 161 unit/L, total bilirubin was 0.3 mg/dL, the endoscopy shows a wide-open esophageal hiatus with an upward displacement of GEJ from diaphragmatic crura indentation (Hill classification grade IV),

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and grade C esophagitis (Los Angeles classification). Histopathology from GEJ mucosa reveals no cardiac mucosa, goblet cell, malignant cell, or Helicobacter pylori. Thoracoabdominal CT shows type I hiatal hernia (Figure 1).

The patient underwent hiatal hernia repair and Nissen fundoplication, which provided 3 days free of singultus. On the fourth postoperative day, the singultus begins 3 times a day, with episodes lasting up to 5 minutes, with a rate of 10-25 singultus per minute. The patient was discharged on postoperative day-5, hemodynamic was stable, no signs of infection, the laboratory results (complete blood count, electrolyte serum, and renal function were still in normal limits; ALT (175 unit/L) and AST (159 unit/L)), with prescription of baclofen 20 mg three times daily. On the 14th day postoperatively, the patient came to the emergency department with ARDS (acute respiratory distress syndrome) due to food aspiration and the patient did not survive.

DISCUSSION

The cornerstone in the management of intractable singultus is identifying the etiology. The singultus in our patient turns up without preceding signs, continuing singultus causing dysphagia, without symptoms of heartburn and regurgitation. There was no sign of infection, history of accident or operation, or history of past illness. The CNS symptoms were drowsiness and inability to focus after taking chlorpromazine for more than 2 months, increasing liver transaminases. There were no other CNS symptoms or peripheral nerve symptoms. Hiatal hernia is found during the endoscopy and CT examination, and biopsy reveals esophagitis in GEJ mucosa. The patient was given chlorpromazine without improvement of the singultus. Omeprazole 20 mg two times daily for 2 months also yielded no improvement in the singultus symptom. The esophagitis is presumed to be the consequence of a hiatal hernia or intractable singultus, not its cause. The acid perfusion test may identify whether the acid provocation (mimicking acid refluxate) on GEJ induces the singultus or not, but we don't have this examination in our institution. Previous studies present a hiatal hernia as a cause of intractable singultus due to diaphragm irritation.1-3 There are no reports of whether a singultus may cause the formation of a hiatal hernia.

Persistent singultus after the operation might be because the etiology has not been identified. Electrocardiogram and CNS evaluation (CT scan and EEG) yielded normal results. And there was no history of past illnesses, trauma, or surgery before. The only medicine approved by FDA is chlorpromazine, but there were many other drugs in different case reports, including GABA-derivatives, baclofen, dopaminergic antagonists, and anticonvulsants.11-15 Site of action of pharmacologic therapy is in the central nervous system that blocks the singultus reflex arc, with the resultant neurologic side effects that potentially impair quality of life.16 Non-pharmacological options such as phrenic nerve blocking, crushing and pacing, percutaneous nerve stimulation, and cervical epidural block has shown limited success due to the occasional presence of an accessory phrenic nerve and the occasional presence of an accessory phrenic nerve bilateral diaphragmatic contraction etiology of centrally originated hiccups.17 Other treatments as acupuncture, massages, and hypnosis are also widely used with different clinical outcomes.

CONCLUSION

Symptomatic treatment for intractable singultus is often unsuccessful, with its serious side effects that may impair the patient's quality of life. The cornerstone of the management of intractable singultus is finding the etiology. Very little research is available on the cure of intractable singultus and the pathogenesis of singultus is less understood. Future research will help us better understand singultus and its treatment.

CONFLICT OF INTEREST

The author declares there is no conflict of interest regarding the publication of the current report.

ETHICS CONSIDERATION

The patient provided written, informed consent for their anonymized data to be used for study purposes. In addition, this case study has followed the ICMJE and COPE protocols for the publication ethics guidelines.

FUNDING

This report received no funding from any sources.

AUTHOR CONTRIBUTIONS

Both authors equally contribute to this study from the conceptual framework, data acquisition, until administering appropriate intervention as well as follow up the outcome of the case study.

REFERENCES