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Transverse-Sigmoid Colon Synchronous Volvulus in a Patient with Lennox-Gastaut Syndrome: Case Report

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ABSTRACT

Introduction: Lennox-Gastaut syndrome (LSG) is an epileptic encephalopathy characterized by a triad: 1). tonic seizures during sleep with atonic and atypical absence seizures, 2). cognitive and 3). behavioral It is estimated that SLG affects between 1 to 2% of patients with epilepsy. In general, LGS often occurs in young children. The etiology of LGS can be classified as cryptogenic or symptomatic. Symptomatic etiology of LGS can be secondary to hypoxic ischemic encephalopathy, congenital brain malformation, vascular malformation, genetics (tuberous sclerosis), trauma, brain tumor, or perinatal meningoencephalitis. Clinical Case: This is a 29-year-old woman, who has a significant history of prolonged prostration; severe cognitive disorder, Lennox-Gastaut syndrome since childhood. She was admitted to the emergency department with symptoms of abdominal distension of seven days of evolution and generalized pain in all abdominal quadrants. A simple computed tomography of the abdomen was performed, where distension of bowel loops was observed at the expense of mesenteric torsion with data of colonic volvulus with little free fluid. It

is evaluated by the general surgery service who determines emergency surgical management by means of exploratory laparotomy, the following findings are addressed and obtained: toxic megacolon from the left end of the transverse colon to the middle third of the sigmoid colon, with multiple necrotic patches, as well as volvulus of the transverse and sigmoid colon, proceeding to perform a left hemicolectomy with colostomy of the transverse colon. Discussion: In children, the incidence is estimated at 2 per 100,000, and it is common in men. LGS is one of the most commonly encountered drug-resistant epileptic encephalopathies, with mortality rates ranging from 3-7%. There are many treatment options for these patients, such as antiepileptic drugs, ketogenic diets, VNS therapy (vagus nerve stimulation) callosotomy, and resective epilepsy surgery.

Keywords: Lennox-Gastaut syndrome, Volvulus, Toxic megacolon, Surgery.

INTRODUCTION

Lennox-Gastaut syndrome (LSG) is an epileptic encephalopathy characterized by a triad: 1) tonic seizures during sleep with atonic and atypical absence seizures, 2) cognitive and 3) Behavioral. It is estimated that SLG affects between 1 to 2% of patients with epilepsy. In general, LGS often occurs in young children. The etiology of LGS can be classified as cryptogenic or symptomatic. Symptomatic etiology of LGS can be secondary to hypoxic ischemic encephalopathy, congenital brain malformation, vascular malformation, genetics (tuberous sclerosis), trauma, brain tumor, or perinatal meningoencephalitis (1).

LGS generally has an onset before the age of eight, however there are exceptions to the rule, such as the case presented by Sixian Li et al., (2) in a 19-year-old man who debuted with this condition at the age of eight years old with mild to moderate symptoms, however at the age of 18 he experienced a worsening of his condition and was monitored with stereotactic electroencephalography. LGS with onset after 8 years of age, known as late-onset LGS, can be caused by focal lesions. Late-onset LGS and classic LGS have some differences in terms of clinical manifestation, cognitive with changes in the prognosis and its treatment. MRI is important in determine the etiology of late onset and is useful for treatment. Compared to craniotomy, RFTC (Radiofrequency thermocoagulation) is limited and less effective in lesion damage. However, clinical improvement after RFTC would support proper identification of the starting zone location indicated by the SEEG results and thus help with surgical planning for cortical resection.

CLINICAL CASE

This is a 29-year-old woman, who has a significant history of prolonged prostration; severe cognitive disorder, Lennox-Gastaut syndrome since childhood, on oral treatment with topiromate and risperidone every twelve hours; gastroparesis with a year of diagnosis being managed with pinaverium bromide, denies use of other drugs, no allergies and no previous surgical history. She was admitted to the emergency department with symptoms of abdominal distension of seven days of evolution and generalized pain in all abdominal quadrants. Her vital signs were taken, reporting blood pressure within normal parameters for age, without tachycardia or tachypnea. Upon indirect questioning, the mother refers intolerance to food intake for five days and vomiting of gastrobiliary content, denies fever, current uresis and

absence of bowel movements for seven days. On physical examination, neurological deficits were found, presence of a nasogastric tube with liquid distal intestinal discharge, pale integuments and dehydration of the mucous membranes, cardiopulmonary without compromise, abdomen with tension distension and pain on superficial and deep palpation, as well as a sign of positive rebound, intact, ankylosed and hypotrophic extremities, distal pulses present and delayed capillary refill of more than five seconds. Laboratory blood studies were taken with hemoglobin of 14.6 g/dL, hematocrit of 43% and leukocytes of $30.7x10^3$/uL. A simple computed tomography of the abdomen was performed, where distension of bowel loops was observed at the expense of mesenteric torsion with data of colonic volvulus with little free fluid. It is evaluated by the general surgery service who determines emergency surgical management by means of exploratory laparotomy, the following findings are addressed and obtained: toxic megacolon from the left end of the transverse colon to the middle third of the sigmoid colon, with multiple necrotic patches, as well as volvulus of the transverse and sigmoid colon, proceeding to perform a left hemicolectomy with colostomy of the transverse colon. In the postoperative period, he was admitted to the intensive care unit where he was reported severe, requiring assisted mechanical ventilation and the use of vasoactive amines for four days, with subsequent management by the general surgery service, where he evolved adequately tolerating the oral diet, presence of fecal waste from colostomy and adequate wound healing in the midline, she was discharged home after five days due to improvement (Figs. 1, 2).

Volvulus at any age is frequent and the objective of making this report with Lennox-Gastaut syndrome is due to the infrequency of this complication in any person regardless of the association of any other disease and that its solution requires the corresponding surgical management.



Figure 1: Toxic megacolon at the expense of the left colon.

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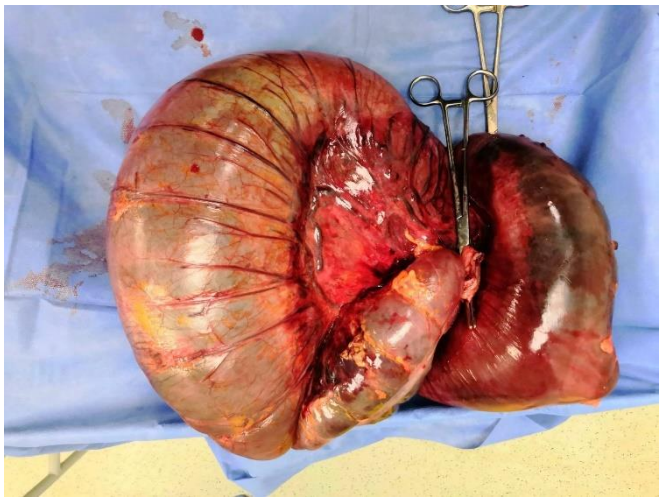


Figure 2: Synchronous volvulus of the transverse and sigmoid colon

DISCUSSION

Lenox-Gastaut syndrome (LSG) is one of the most severe epilepsy phenotypes associated with drug-resistant seizures and significant cognitive alterations such as Deep Brain Stimulation (DBS), in such a way that due to the complexity of the disease there are many the treatments for its management depending on the states and the same and other variables that affect its management (3).

In children, the incidence is estimated at 2 per 100,000, and it is common in men. LGS is one of the most commonly encountered drug-resistant epileptic encephalopathies, with mortality rates ranging from 3-7%. There are many treatment options for LGS patients, such as antiepileptic drugs, ketogenic diets, VNS therapy (vagus nerve stimulation) callosotomy, and resective epilepsy surgery (4).

Deep brain stimulation (DBS) is an emerging therapy for drug-resistant epilepsy. It involves the implantation of electrodes in subcortical structures with the aim of gradually modulating both the stimulation site and the connected circuits, generating seizures. In other diseases, particularly in movement disorders, significant progress has been made in understand the optimal objectives of DBS (5).

Purified cannabidiol (CBP) is a new drug that can be used to treat drug-resistant epilepsy, as its administration can reduce seizure frequency. Mild adverse events such as somnolence and gastrointestinal symptoms were reported. CBD exerts anticonvulsant effects, while another component of cannabis, Δ9-tetrahydrocannabinol (THC), has psychoactive functions. Efforts by the media, patient groups, and non-governmental organizations informed the public that CBD medicines contained a very small amount of THC, with very low potential for addiction and psychoactive effects. These acts led to the approval of CBD by the Ministry of Food and Drug Safety for the treatment of DS or LGS in people older than two years (6).

“Lennox-Gastaut syndrome (LGS), a childhood-onset severe developmental and epileptic encephalopathy (DEE), is an entity that encompasses a heterogenous group of etiologies, with no single genetic cause. It is characterized by multiple seizure types, an abnormal EEG with

generalized slow spike and wave discharges and cognitive impairment, associated with high morbidity and profound effects on the quality of life of patients and their families" (7).

"Lennox-Gastaut syndrome (LGS) is a pharmaco resistant epileptic encephalopathy. Herein reported is a case of LGS that combination therapy with levetiracetam, lamotrigine and valproate culminated in control of all seizure types and resolution of epileptic discharges in electroencephalography. This case indicates that logical combination therapy may provide seizure control and improvement of electroencephalographic pattern in patients with LGS even in cases at which epileptic surgery fails." (8)

Therapeutic management of seizures in LGS is challenging due to the high demand for medications. And resistance to antiepileptic drugs. Valproate, lamotrigine, and topiramate are first-line drug therapies, followed by rufinamide, clobazam and felbamate. Recently, newer treatments such as cannabidiol and fenfluramine have shown efficacy in these patients. Despite the increasing number of therapeutic options available, long-term outcomes remain poor due to drug-resistant seizures and impaired psychosocial functioning (9).

Most recently, the ELEKTRA study achieved its primary endpoint, with soticlestat, obtaining a statistically significant reduction in seizure frequency compared with placebo (10).

CONCLUSION

Colonic volvulus is not associated with any specific disease, but it is related to pathological states of decreased intestinal transit. It is a surgical emergency that must be attended to as soon as possible because it is associated with intestinal distress ischemia, perforation, and sepsis.

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