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Case Report

Sandifer syndrome: the mis-interpretable disorder

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ABSTRACT

Sandifer syndrome (SS), a movement disorder which is characterised by spasmodic torsional dystonia with back arching and rigid opisthotonic posturing, negatively impacting predominantly the neck, back, and upper extremities. Symptomatic gastro-esophageal reflux disease, esophagitis, or the presence of a hiatal hernia are all associated with Sandifer syndrome. The cause of Sandifer syndrome being uncertain, lifestyle adjustments and modifications highlights as the appropriate mode of treatment. To treat the condition and help relax the baby after feeding, dietary changes or medications can be administered. The case report of a patient with Sandifer Syndrome is considered for observation. Upon arrival, the child was stable, and an Electro-encephalogram (EEG) test revealed nothing abnormal. The child was taking several Anti-epileptic drugs (AED's), which were stopped in favour of Sodium valproate and Pyridoxine. An opinion from a Gastro-enterologist was sought in light of the epilepsy and possible Gastro-esophageal reflux disease (GERD), and they suggested a milk scan. Rantac was then started, and breastfeeds were thickened. Milk can indicate mild GERD and a reduction in episode frequency. So, sodium valproate was discontinued. Haemodynamically stable child was discharged from the hospital with Pyridoxine and Carnisure. Studies shows most cases of SS improve over time, within the first 24 months in general.

Keywords: Sandifer syndrome, Esophagus, Gastro-esophageal reflux disease, Epilepsy, Opisthotonic posturing

INTRODUCTION

The movement disorder known as Sandifer syndrome (SS) causes paroxysmal spasms of the head, neck, and back but spares the limbs. In children, SS and Gastro-esophageal reflux disease (GERD) are frequently linked.[1] When a patient has extra-esophageal symptoms, usually neurological ones, it is a rare complication of Gastro-esophageal reflux disease (GERD).[2] SS affects the children generally up-to the ages of 18 to 24 months.[3] These signs and symptoms could be so severe as to resemble epileptic convulsions or seizures.[4] Opisthotonic posturing and eye movements are also possible. These strange and abnormal movements, which are typically non-existent while you sleep, have been associated with food intake.[5] With hardly supportive evidence, this syndrome was first mentioned in the medical literature more than 50 years ago.[6]

CASE REPORT

A five-month year old baby came with history of multiple episodes of left sided jerky movements from 30 January 2022 (lasting to 15 to 30 seconds) which resolves spontaneously. No history of post ictal drowsiness. After each episode the child cries well and shows good appetite. Parents denied any history of fever was shown along with the episodes mentioned above when admitted and evaluated at JIPMER and then subjected to multiple anti-epileptics. Lab investigations, MRI scanning and ECG were normal.

At term, the baby was born normally. He was breastfed shortly after birth, and he had never previously experienced physiological jaundice. There was no notable family history of these symptoms or any chronic illnesses. The motor and mental development were

normal. The neurological and physical examinations came out normal.

Initially on arrival at Apollo Children's Hospital, the patient was conscious and crying. Physical and neurological examination were normal. The child was on multiple AED' and one of those were stopped. Sodium valproate and Pyridoxine were continued. On the following day, opisthotonic posturing was decreased on evaluation, and started thickened breast milk. On day 3, baby was conscious, reported one episode of facial twitching initially with excessive crying and oral secretion. Gradually multiple episodes of jerky movements and arching were shown. Therefore, started Carnisure and Rantac syrup. On the next day, in view of epilepsy and suspected GERD, gastro-enterology opinion was obtained and advised for milk scan. Reports showed mild GERD and also frequency of episodes were decreased. Hence Sodium valproate was stopped. Child was haemodynamically stable and discharged on day 6 with Pyridoxine, Carnisure, Ranitidine and Vizylac GG drops.

DISCUSSION

Sandifer's syndrome (SS) is an uncommom juvenile form of gastro-esophageal reflux syndrome which is characterized by unusual and aberrant movements of the eyes, head, neck and trunk.[12] It is uncertain what specifically causes Sandifer syndrome. The stomach's contents can flow back into the food pipe because of an under-developed connection between the food pipe (esophagus) and the stomach in new-borns, which can cause gastro-esophageal reflux disease (GERD). The baby's dystonic movements or muscle spasms are a response to GERD related pain or discomfort. The baby is attempting to stop the internal movement of the contents of their stomach and esophagus moving in the wrong direction when they quickly jerk their body or arch their back.[3]

The main symptoms of SS include Torticollis (neck movements that are involuntary) and dystonia (involuntary muscle contraction) along with other symptoms such as choking, slow feeding, constant irritability, coughing, breath-holding episodes, slow feeding, recurrent pneumonia, anaemia, weight loss, wheezing or stridor, upper abdominal bulking (hiatal hernia), etc.[3,7,10] Although these are the symptoms mis-diagnosis can happen in certain cases. In the study conducted by Uldall et al, it was reported that 39% of kids with epilepsy diagnosis were misdiagnosed, and around 47% of these diagnoses included non-epileptic paroxysmal events (NEPEs).[8] And therefore, it is essential that doctors carefully consider each observational finding in order to provide an appropriate diagnosis.

To confirm that it is a case of SS, endoscopy, esophagram (while the child is swallowing, X-ray images of their esophagus can be captured to observe its functioning), electro-encephalogram (EEG), gastro-esophageal scintigraphy, gastric pH monitoring.[7]

Gastro-esophageal scintigraphy (GERS): This is a quick procedure with only the least amount of discomfort for the patient and is an accurate method for identifying gastro-esophageal reflux (GER). A sensitive, non-invasive, physiological, and direct method to detect GER is gastro-esophageal reflux scintigraphy. Ultra-sonography and 24-hour pH monitoring are displayed by GERS, and also offers information on abnormal esophageal contractions, aspiration, and gastric emptying. It can also identify reflux that is non-acidic.[9]

Most cases of Sandifer syndrome improve over time and growth, generally within the first 24 months. As initial modes of treatment, modification of lifestyle, feeding adjustments (such as reducing volume, feeding more frequently, and avoiding over-feeding), keeping the baby in a semi-upright position for 30 minutes following a feeding, using a protein or amino-acid-based formula that has been extensively hydrolyzed, with or without thickening, blending up to 1 tablespoon of rice cereal with every 2 ounces of infant formula, etc. can be executed. Later, in severe conditions, administration of drugs that block H2 receptors, such as Ranitidine and Famotidine, Antacids, drugs that inhibit proton pumping, such as Lansoprazole are effective in improving the condition.[7]

CONCLUSION

Sandifer syndrome is a condition that requires a high index of caution. Primary care providers (PCP) should be competent to recognize and treat SS. When the clinical picture is not apparent, the PCP should send the patient to a pediatric neurologist for additional assessment to ascertain the underlying cause. Early diagnosis facilitates prompt treatment and prevents any complication. It is necessary to conduct further studies to ascertain the exact cause of Sandifer syndrome.

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