Journal of Scientific and Innovative Research 2019; 8(1): 4-6
Available online at: www.jsirjournal.com

Research Article

ISSN 2320-4818
JSIR 2019; 8(1): 4-6

Received: 20-03-2019
Accepted: 09-04-2019

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Juvenile Myasthenia gravis presenting with recurrent aspiration pneumonitis in a Nigerian child

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Abstract

Myasthenia gravis (MG) is a rare and chronic neurologic disease of neonates and older children. It commonly presents with progressive skeletal muscle weakness. Presentation with aspiration pneumonitis is uncommon and more so with recurrent aspiration pneumonitis. Diagnosis of recurrent aspiration secondary to MG can thus, be slow to make, especially at in a resource limited setting. A five year old boy presented with features suggestive of recurrent food aspiration and underlying Myasthenia gravis at the paediatric emergency unit of a Nigerian hospital. The case is reported in order document the occurrence of MG in our resource limited setting with the view to provide information that may aid the diagnostic acumen of physicians practicing in similar settings. The challenges associated with managing Myasthenis gravis in a resource limited setting are also discussed.

Keywords: childhood, aspiration, myasthenia gravis.

INTRODUCTION

Juvenile Myasthenia gravis (MG) is a rare chronic childhood disease manifesting with rapid fatigability of the striated muscles [1]. It occurs secondary to autoimmune reaction against acetylcholine receptors and it is uncommon in paediatric practice [2]. Reports on MG in Nigerian children are scarce as most of the few available are on adults, which may suggest that MG in the Nigerian child is rare or missed or under-reported [3, 4, 5]. The three clinical variants of MG in childhood are Juvenile myasthenia gravis in late infancy and childhood, congenital myasthenia and transient neonatal myasthenia.

Clinically the early signs of Juvenile Myasthenia gravis are ptosis and some degree of extraocular muscle weakness. Other features of MG in infants and older children include dysphagia, feeding difficulties and weakness of the limb girdle muscles and distal muscles of the hands with use. Patients tent to be more symptomatic later on during the day, especially in the evening or night. Symptoms are also more pronounced when the patient is exhausted or tired [1].

Mysthenia gravis is usually progressive if left untreated and it can become life threatening with involvement of the bulbar muscles and respiratory system, which may be compounded with the risk of aspiration. Respiratory involvement is particularly pronounced when the child has an upper respiratory tract infection or is otherwise unwell [1, 2].

Recurrent aspiration pneumonitis in children are commonly due to gastrooesophageal reflux, trachea-oesophageal fistulas, altered consciousness, cleft palate, laryngeal cleft, dysautonomia, immaturity of swallowing, vascular rings and oesophageal foreign body [6]. The rarity of myasthenia gravis leading to recurrent aspiration pneumonitis, coupled with the challenges associated with making obtaining an accurate diagnosis in a resource limited setting informed the decision to report the present case.

CASE PRESENTATION

A five-year-old boy was brought by his maternal grandmother to the childrens' emergency unit of Ladoke Akintola University of Technology, Osogbo with loss of consciousness. He had lapsed into unconsciousness 2hours prior to presentation following a bout of cough and a single episode of vomiting part of the feeds being ingested. The patient was only taking his food and not speaking before developing these symptoms. The grandmother did not witness these symptoms because she was in an adjacent room

however, she rushed to view the patient, when she heard the bouts of cough and surmised that the patient probably choked while feeding.

In addition to the above symptoms the grandmother witnessed the child sneeze and cough several times, while holding his chest, before lapsing into unconsciousness. Palm oil, water and pepper were the main fluid discharge noticed in the nostril of the child during the bouts of coughing and sneezing. There was also a transient period of cessation of breathing which compelled the caregivers to rush the child to a nearby private hospital, where he was given two unknown injections. The private hospital was said to have referred the child verbally to Ladoke Akintola University of Technology, Osogbo for further care after giving the injections.

The father was a security personnel who died five years previously at the age of 34years, while the mother is a 31year old petty trader, residing at Abuja with three older siblings of the child aged 13,10 and 8years. There is no family history of similar illness in the parents or other siblings. The mother earns about $40 per month which is barely enough to sustain her and the children at Abuja.

On examination at presentation the patient was conscious but weak and in respiratory distress. He was a febrile with a temperature of 36.1^0C and weighed 14kg. The patient was well hydrated. Examination of the respiratory system revealed dyspnoea with intercostals recession and diffuse coarse crepitations. The pulse rate was 142beats per minute and the first and the second heart sounds were normal. Neuromuscular examination revealed generalized weakness, normal tone and deep tendon reflexes were elicited. Examination of the other system were essentially normal.

An assessment of aspiration pneumonitis was made and a chest radiograph done at admission showed generalized evidence of pneumonitis. The SPO_2 at admission was 54% and this improved to 90% after suctioning the airway intermittently and administering humidified oxygen at the rate of 2L/min. Random blood sugar assessment at admission was reported to be 15.2mmmol/L. The full blood count, electrolytes, urea and creatinine results were normal.

Oxygen administration was continued and the patient was put on 70mg of intravenous hydrocortisone six hourly for 24hours. He was also treated with 750mg of Cefuroxime 12hourly and 100mg of metronidazole 8 hourly. The patient was allowed nil per oris for the first 48hours and put on intravenous fluids at maintenance rate. Improvement in the clinical condition of the child was assessed to be good at 48hours of admission. The respiratory rate had then decreased to 32 breaths per minute, the breath sounds were vesicular and of good intensity and a pulse oximetry of 98% was obtained in room air. Administration of oxygen was discontinued and the patient was restarted on oral feeds.

The patient was however noticed to have developed bouts of cough and choking had re-appeared on re-commencement of oral feeds at 72hours of admission. In addition, generalized weakness and drooping of both eyelids were noticed on general examination. Systemic examination was normal. Thus the patient was commenced on naso-gastric tube feeding and the diagnosis of Myathenia gravis as the possible cause of recurrent choking and aspiration was entertained.

The patient tested positive to the tensilon test with a 1cm increase in the distance between the upper and lower eyelid on administration of 0.6mg of neostigmine. Further tests such as single fibre electromyography and serology tests to detect antibodies to acetylcholine receptors or muscle-specific kinase antibodies were not done because they are not available in the hospital or other centers in the state. CT scan of the thymus could not be done because of financial constraints.

Thus the patient was presumptively started on prednisolone at a dose of 1mg/kg after one week of admission. The drooping of the eyelids ceased and the patient became active and playful around and on the ward without developing weakness during the course of the day. Oral feed were recommended and this was well tolerated without choking. The patient sustained the good recovery and was discharged to the neurology clinic in good health after two weeks of admission and treatment with oral prednisone. We intend during outpatient follow up to combine Azathioprine with the prednisolone and ultimately wean off prednisolone.

DISCUSSION

Recurrent aspiration is uncommon in neurologically intact children because of the protection of the airway from foreign objects or chemicals by the gas reflex. Aspiration tends to be recurrent in children who have lost this protective reflex either as a result of cerebral palsy or coma [6]. Congenital defects such as tracheooesophageal fistulas, cleft palate or laryngeal cleft can also predispose to recurrent aspiration of gastric contents and foods [6, 7]. Similarly, disease affecting the functioning of the gastro-oesophageal sphincter such as gastroesophageal reflux disease may be responsible for recurrent aspiration and pneumonitis [8]. In our and similar, societies forceful administration of feeds and drugs are known causes of serious and even fatal aspiration.

Poliomyelitis and Gullian Barre Syndrome are very rare causes of recurrent aspiration in children with neurological defects in under developed countries, Nigeria inclusive. Cerebral palsy is probably the most common cause of recurrent aspiration in children with neurological defects [6, 8]. The incidence of Myasthenia gravis varies based on the geographical location. The incidence of myasthenia gravis in Nigeria is not known and our clinical experience in Nigeria also suggests that the incidence of MG is much less compared with those of poliomyelitis and Gullian barre syndrome. A high index of suspicion and a good clinical acumen are evidently vital for the diagnosis of an uncommon presentation of a rare disease.

The challenges associated with the diagnosis of MG have already been highlighted. None of the electrophysiological or serological antibody tests was available or accessible at the district or province where the case was seen. The Tensilon test and the computerized tomography were the only available tests and the computed tomography test was beyond the financial reach of the patient. Yet all these tests need to be conducted in order to distinguish auto-immune from congenital myasthenia. The distinction is very important because of the differences in the treatment options, prognosis and genetic implications [1].

Managing a rare and chronic disease poses some daunting challenge. The social disadvantages in this child such as being raised by a single parent and the low socioeconomic status of the family likely contributed in no small way to the inability of this child to access quality optimum care, evidenced by the fact that only one of the necessary investigations for management could be conducted. Socially disadvantaged children have been found to be unable to access optimum care and consequently often need extra support from the attending physician, the government or philanthropists in order to get the best care [9,10]. The National health Insurance Scheme has been proffered as an important alternative for accessing affordable quality health care in developing countries by the poor and socially disadvantaged [11]. It is hoped that access to care and the repackaged exemptions under the National Health Insurance Scheme (NHIS) may probably be the way forward in accessing comprehensive care among the poor and socially disadvantage with chronic diseases which are also expensive to manage [12].

In the mean time the family of the patient may need to result to taking loans or seeking some other forms of assistance in order to ensure that this child obtains quality care. It is concluded that a high index of suspicion and good clinical acumen are vital ingredients in making the diagnosis of a rare disease like MG presenting unusually with recurrent aspiration. Availability of diagnostic kits or reagents for proper work up of patients with MG is a challenge in resource limited settings like Nigeria.

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