Case Report

Triple “A” syndrome presenting as recurrent chronic sinusitis with Pneumonia, septic shock and meningoencephalitis in a child

Ramaning Loni*, Priyanka Agrawal**, Prashant Rajebhosale***, Mrutyunjaya Panda****, Prashant Darveshi***** Prakash Valse******

*Pediatric Intensivist & In charge Pediatric ICU, Dr. Bidari’s Ashwini Children Hospital, BLDEA’s Road, Vijayapura, Karnataka, India. **Senior Resident in Pediatrics, department of pediatrics, ***Pediatric Intensivist, ****Consultant Gastroenterologist, ***** Consultant in Pediatric surgery, *******Gastrointestinal Surgeon, Aditya Birla Memorial Hospital, Chinchwad, Pune, India.

Recived:22-Jun-18/Accepted:08-Aug-18/Published Online:30-Aug-18

ABSTRACT

Triple A syndrome (All grove syndrome) is a rare autosomal recessive disorder characterized by the clinical triads of adrenal insufficiency, achalasia cardia and alacrimia with variable association of autonomic and neurological manifestations. Nearly100 cases have been reported all over world however in India, only 2 to 3 cases have reported. We present 13 years old boy with chronic recurrent sinusitis with failure to thrive without lacrimation since birth who had presented recurrent chronic sinusitis with pneumonia, septic shock, and acute meningoencephalitis with adrenal insufficiency. Catastrophic complications can be prevented with adequate cortisol and specific measures such as cardiac pneumatic dilatation or myotomy along with other supportive management.

Key words: Triple “A” syndrome, child, Shock

Introduction

Triple “A” syndrome (All grove syndrome) is a rare autosomal recessive disorder characterized by the clinical triads of adrenal insufficiency, achalasia cardia, and alacrimia. It was first described by Allgrove in 1978 (1) but in 1995, it was diagnosed as a syndrome with the variable association of autonomic and neurological manifestations. This disorder manifests in the early childhood with life-threatening complications like severe hypoglycemia, shock, and recurrent infection. The syndrome usually presents during the first decade of life with dysphagia, while other signs may be delayed until adulthood. We present 13 years old boy with chronic recurrent sinusitis with failure to thrive without lacrimation since birth who has presented pneumonia, septic shock, and acute meningoencephalitis with adrenal insufficiency.

Case report

13 years old boy had presented to us with recurrent vomiting and fever on/off for last 8 months with significant weight loss and failure to thrive for last 6 months but acutely presented to us for fever for 2 days and altered sensorium with one episode of generalized tonic-clonic seizure for 1 day. History of hyper nasal speech was present with no tears secretions by birth whenever he cries.

He was wasted as well stunted. He was febrile with low sensorium, tachycardic with cold hypotensive septic shock with BP of 72/38 mmHg with mild respiratory distress. His investigations anemia with mild leukocytosis with high CRP levels (112 mg/dl). Chest X ray in figure 1 revealed right-sided pneumonia with parapneumonic effusion and X-ray PNS (Figure: 2) showed bilateral maxillary and frontal sinusitis. His CSF showed total cell count of 12 with lymphocytic predominance. Tuberculosis gene expert test of pleural fluid was negative. Immunological workup showed normal serum immunoglobulins levels and negative HIV antigen I and II.

He was given appropriate treatment for 14 days and also underwent functional endoscopic sinus surgery (FESS) for his sinusitis and discharged home with reasonable recovery but got readmitted after 11 days with recurrent nonprojectile vomiting, multiple episodes in a day leading to hypovolemic shock with hypoglycemia in the emergency. His blood sugars by dextrostix were 38 mg, 62 mg and 87 mg at 0 hr, 1hr and 6hrs of 2nd admission respectively and he was...
resuscitated for hypovolemic shock, hypoglycemia and stabilized. He underwent upper GI endoscopy (Figure 3) which showed narrowed LES with residual food. Upper GI series - Gastrografin dye (Figure 4) showed prominent and dilated lower two-thirds esophagus with retained contrast with the very minimal flow of contrast in the stomach. His esophageal manometry showed LES pressure of 37 mmHg (high) with complete relaxation to wet swallows with the generalized failure of peristalsis: Achalasia type 2 and recurrent hypoglycemia workup showed lower serum cortisol level 4.55 nmol/l with normal fasting insulin and thyroid function test. He was started on IV hydrocortisone after endocrinologist’s opinion and continued till 4 days after surgery and later started on oral hydrocortisone supplementation as per Endocrinologist’s advice.

He underwent laparoscopic cardiomyotomy with fundoplication on day 5th of readmission once child was stabilized after discussing pros and cons of both ballondilatation and myotomy to the parents. His repeat Gastrografin dye studies (upper GI series) showed resolution of achalasia cardia with the flow of dye into the stomach (Figure 5).

His sugars normalized, tolerating oral diet well and his weight improved from 19 kg with BMI 14.4 to 27 kg with BMI 14.8 within 3 months on OPD follow up and his serum cortisol was within normal range with oral hydrocortisone 10mg in the morning and 5 mg in the evening.

Discussion
The description of Allgrove syndrome is limited to case reports/series and thus prevalence is unknown. Allgrove’s syndrome is considered a rare autosomal recessive disorder with variable presentations.1,2 Recent studies have identified a mutation in the AAA syndrome of a candidate gene on chromosome 12q13 in such patients.3,4 In our index case, the child has presented with dry eyes since birth with recurrent sinusitis, respiratory infections due to reflux regurgitation.
leading to severe failure to thrive. Achalasia is most common presentation leading to failure to thrive, weight loss with recurrent sinopulmonary infections but alacrima was first to be noticed by parents in this case. Adrenal insufficiency and achalasia are usually manifested during the first decade of life. Achalasia of the cardia occurs in about 75% of cases so, in older children and adults it usually manifests as dysphagia especially for liquids. Symptoms of achalasia may appear in individuals as young as 5 months or as late as early adulthood. Based on clinical criteria, we have diagnosed the index case, however genetic work up is not done due to economic constraints with parental refusal.

The child underwent esophagogastroduodenoscopy (EGD scopy), upper GI series, so as an assessment of esophageal motor function is essential in the diagnosis of achalasia. Barium esophagram and esophagogastroduodenoscopy (EGD) are complementary tests to manometry in the diagnosis and management of achalasia. However, neither EGD nor barium esophagram alone is sensitive enough to make the diagnosis of achalasia with certainty. The gold standard esophageal manometry which helped in the diagnosis of Achalasia cardia as LES pressure was 37 mmHg as resting pressure is required. In this case, the child had presented with hypoglycemic episodes with fluid refractory dopamine resistant shock due to adrenal insufficiency. Adrenal insufficiency is also an early manifestation and manifests as severe hypoglycemic or hypotensive attacks during childhood which may lead to sudden death. So, the documentation of normal electrolytes indicated normal mineralocorticoid production although we have not measured plasma aldosterone levels. The only other cause of primary adrenal insufficiency with preservation of mineralocorticoid production is familial glucocorticoid deficiency. Although, most of the patients with Allgrove syndrome have preserved mineralocorticoid production, it may be impaired in 15% of patients. Dry eye with irritation was demonstrated by ophthalmological evaluation including Schirmer’s test. The child has undergone repeat upper GI studies(Gastrografin study) after surgery which showed resolution of Achalasia and passage of dye into the stomach and small intestine.

On follow up after 3 months, Child has gained weight and asymptomatic and having more than average scholistic performance.

**Conclusion**
Allgrove’s syndrome is although a rare disorder. It’s catastrophic complications can be prevented with adequate cortisol and specific measures such as cardiac pneumatic dilatation or myotomy along with other supportive management. The prognosis for health and quality of life can be significantly improved by early diagnosis and treatment.

**Conflict of Interest :** Nil  
**Source of Funding :** Nil  

**References**


How to cite this article:

How to cite this URL: