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Pediatric Esophageal Achalasia: Navigating Rare Challenges in Young Patients

Nino Tamarashvili[1], Beso Zenaishvili[1], Rusudan Gudjabidze[1], Lela Laghidze[1], Nino Vardosanidze[2]

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ABSTRACT

This case report describes a 9-year-old girl diagnosed with esophageal achalasia after a 5-year delay in treatment, presenting initially with aspiration pneumonia and respiratory failure. Esophageal achalasia is a rare neurodegenerative disorder predominantly affecting adults, with pediatric cases comprising less than 2% of diagnoses. In our case of pediatric esophageal achalasia complicated by aspiration pneumonia and respiratory failure. Initially managed for fever and respiratory symptoms with outpatient antibiotics, her condition deteriorated rapidly, necessitating hospitalization. Clinical examination revealed respiratory failure, persistent regurgitation, and severe malnutrition. A diagnostic workup confirmed aspiration pneumonia, underscoring the diagnostic challenges in pediatric achalasia. The treatment plan involved fluid resuscitation, antibiotics, and surgical intervention post-stabilization. This case highlights the critical importance of early recognition and intervention in pediatric esophageal achalasia to mitigate severe complications. Awareness among clinicians is essential, as delayed treatment can lead to profound morbidity in young patients. Further studies are warranted to enhance diagnostic strategies and optimize outcomes in this rare pediatric population.

Keywords: Aspiration pneumonia; Esophageal achalasia; Respiratory failure.

INTRODUCTION

Esophageal achalasia occurs because the lower esophageal sphincter fails to relax; predominantly recognized in adults, a prevalence of 10 per 100000 presents a unique and challenging scenario when diagnosed in pediatric patients.[1,2] We present a compelling case of a 9-year-old girl admitted with a complex constellation of symptoms stemming from untreated esophageal achalasia.[2,3] Initially managed as an outpatient for fever and respiratory symptoms, her condition worsened, and she had to be hospitalized. Clinical examination revealed significant findings, including fever, cough, signs of respiratory distress, persistent regurgitation, inability to swallow food and liquids, and severe malnutrition.[2,4] Diagnostic investigations confirmed aspiration pneumonia, underscoring the diagnostic intricacies in pediatric achalasia cases. Following stabilization, comprehensive treatment involving fluid resuscitation, targeted antibiotics, antifungal medications, and subsequent surgical intervention was implemented.[5] This case underscores the critical importance of early identification and intervention in pediatric esophageal achalasia to mitigate severe complications.

CASE

A 9-year-old female patient was admitted to the hospital, presenting with a 7-day history of sore throat, cough, and high fever that did not respond to antipyretic medications. She had received outpatient treatment with azithromycin for three days prior to admission. Despite medical management, her condition deteriorated, characterized by lethargy, increased cough frequency, persistent fever, and difficulty in swallowing liquids and solids. The patient's medical history revealed a diagnosis of achalasia at four years of age, which had not been treated. Upon admission, she exhibited severe malnutrition (weighing 19 kg, BMI – 11.2 kg/m2, indicating severe and prolonged malnutrition) and constipation due to inadequate nutrient intake. Early symptoms included hyperthermia, respiratory failure, and low oxygen saturation levels (SpO2 – 84-86% without oxygen supplementation). Severe vomiting ensued, leading to an inability to consume fluids and solid food, resulting in severe dehydration.

Her laboratory tests revealed anemia (Hemoglobin – 107 g/l, Hematocrit – 33.3%, Mean Corpuscular Volume – 64.3), a negative rapid test for COVID-19, chest X-ray showed infiltration in the right lower lobe and fibrous lung parenchyma, and markedly increased C-reactive protein levels (122.3 mg/l). Abdominal ultrasound showed pseudolithiasis, possibly related to treatment with ceftriaxone. (Onset of pseudobiliary stones varies from 2 to 22 days after starting treatment with ceftriaxone) Sputum sample results indicated Staphylococcus aureus and fungal infection; because of this, antifungal medication was added to the treatment. Due to the persistent fever and low oxygen saturation, we decided to perform a chest CT scan. This decision was based on the patient's deteriorating general condition, worse than what could be assessed through auscultation and radiologic changes seen on X-ray. Additionally, upon receiving the results of the antibiotic susceptibility testing (antibiogram), we opted to switch from ceftriaxone to piperacillin-tazobactam.

Taking into account the patient's medical history, along with laboratory and radiographic analyses, the diagnosis was confirmed as aspiration pneumonia as a complication of esophageal achalasia.

Chest CT revealed infiltrations, especially in the lower right lobe, with bronchodilation on the same side and mild pleural effusion. An esophageal x-ray revealed an enlarged diameter of the esophagus, and emptying of the esophagus from contrast was delayed up to 15 minutes. As the patient was already unable to swallow fluids or solids and had forceful vomiting, their general condition worsened, and severe dehydration appeared; the team placed a nasogastric tube for proper feeding. A cardiologist excluded any severe cardiac abnormality. Over the past five years, she experienced persistent vomiting, regurgitation, and intermittent sub-febrile temperature.

The final diagnosis was esophageal achalasia grade III. After ten days of treatment, expectations included stabilization of respiratory function, resolution of pneumonia with appropriate antibiotic therapy, and improvement in nutritional status through high-calorie intake. After stabilization, surgical treatment was performed without complications.

DISCUSSION

The case of a 9-year-old girl presenting with aspiration pneumonia and respiratory failure secondary to esophageal achalasia underscores several critical aspects of this rare condition in pediatric patients.[6] Esophageal achalasia, characterized by impaired esophageal peristalsis and inadequate relaxation of the lower esophageal sphincter, is a notably uncommon diagnosis in children, further emphasizing the significance of early recognition and intervention.[7] Esophageal achalasia in children poses unique challenges due to its atypical presentation and delayed diagnosis, as evidenced by this case, where symptoms began at the age of 4. However, definitive treatment was not initiated until age 9.[1] This delay resulted in severe complications such as malnutrition, recurrent aspiration episodes, and, ultimately, respiratory failure, highlighting the potentially grave consequences of untreated achalasia in pediatric patients.[5] Clinicians should maintain a high index of suspicion for achalasia in children presenting with chronic symptoms of dysphagia, regurgitation, and respiratory distress, particularly in the context of failed outpatient treatments and persistent symptoms. Early diagnostic interventions, including esophageal imaging studies and manometry, are crucial to confirm the diagnosis and initiate appropriate therapeutic strategies promptly.[8] Treatment options for esophageal achalasia in pediatric patients typically involve a multidisciplinary approach, including medical management, pneumatic dilation, surgical myotomy, or peroral endoscopic myotomy (POEM), tailored based on the patient's age, symptom severity, and anatomical considerations such as esophageal diameter.[6] In this case, surgical intervention was warranted after stabilization, aiming to alleviate esophageal dysmotility and prevent further complications. Despite the efficacy of surgical interventions in improving symptoms and quality of life, challenges such as post-procedure complications and the potential for treatment efficacy to diminish over time necessitate long-term follow-up and consideration of alternative therapeutic modalities.[7] This underscores the importance of ongoing monitoring and individualized management strategies in pediatric patients diagnosed with esophageal achalasia.[5] Furthermore, the rarity of esophageal achalasia in children, accounting for less than 2% of cases under the age of 16, highlights the need for increased awareness among healthcare providers to expedite diagnosis and intervention. Early recognition and treatment not only mitigate the risk of severe complications like aspiration pneumonia but also improve long-term outcomes and quality of life for pediatric patients affected by this challenging condition.[1]

CONCLUSIONS

In conclusion, this case highlights the importance of early recognition and management of esophageal achalasia in pediatric patients to prevent severe complications such as aspiration pneumonia and respiratory failure. Esophageal achalasia not only affects body weight but also impacts mental health and social life. Clinicians should maintain a high index of suspicion for achalasia in children presenting with symptoms suggestive of esophageal dysmotility and promptly initiate appropriate diagnostic and therapeutic interventions to optimize patient outcomes.

Early recognition and prompt management of esophageal achalasia in pediatric patients are crucial to prevent severe complications like severe malnutrition, aspiration pneumonia, respiratory failure, and impaired social life. Clinicians should maintain a high index of suspicion in children presenting with chronic dysphagia and regurgitation, ensuring timely diagnosis and tailored treatment. Continued research and awareness are essential for optimizing outcomes and improving the quality of life for affected young patients.

AUTHOR AFFILIATIONS

[1]Geo Hospitals Tbilisi Multiprofile Medical Center, Tbilisi, Georgia;
[2] Petre Shotadze Tbilisi Medical Academy, Tbilisi, Georgia.

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