Achalasia cardia in a child with severe malnutrition

A case report

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INTRODUCTION

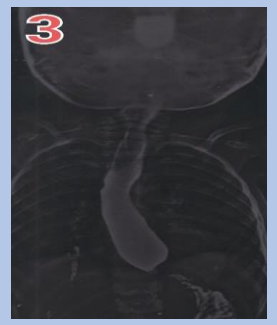
Achalasia cardia is an esophageal motility disorder due to the failure of the lower esophageal sphincter to relax. It is extremely rare in the pediatric population with an incidence of 0.11 per 100,000 children. In children, it is usually seen in association with other conditions like adrenal glucocorticoid deficiency and down's syndrome. Common symptoms include recurrent vomiting, dysphagia, regurgitation and weight loss. It's exact etiology is still unclear. It is considered to be due to degeneration of the myenteric plexus that innervates the lower esophageal sphincter leading to difficulty with swallowing and absence of peristalsis.

CASE DESCRIPTION

We present the case of a 3-year-old with history of recurrent vomiting and poor weight gain. Her symptoms have been progressive for over 2years. The child was initially treated at multiple local hospitals for Gastroesophageal reflux disease with no improvement. At presentation she was severely malnourished and weighed 13lbs(fig.1). Barium study showed tapering of distal esophagus, representing the classic bird-beak sign(fig. 3,4,5), and the diagnosis of achalasia cardia was confirmed. She was treated surgically by Heller's cardio-myotomy and fundal wrap(fig.6,7,8,9). Her post-op recovery was uneventful. Her weight at 1month follow-up visit was 20lbs and at 3 months it was 33lbs(fig.2).

Fig.1: Initial visit; 20lbs

Fig.2: 3months post-op; 33lbs



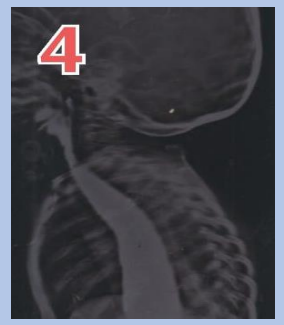
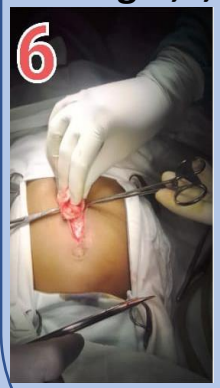
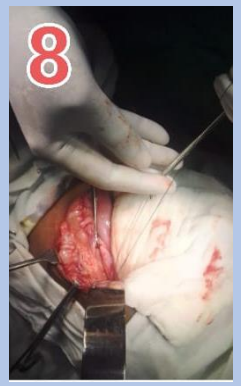


Fig.3,4,5: Barium swallow: Bird-beak sign





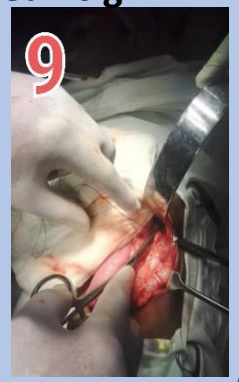


Fig.6,7,8,9: Heller's Cardio-myotomy with fundal wrap

DISCUSSION

Achalasia cardia is a rare presentation in children but if left untreated it can lead to severe complications. Due to the non-specific symptoms it is often misdiagnosed as gastroesophageal reflux disease leading to delayed diagnosis, severe malnutrition and failure to thrive. Diagnosis is through barium swallow study and manometry. It is followed by endoscopic biopsy in adults to rule out malignancy. However, biopsy is not a consideration in children. Manometry studies are also generally avoided in children due to technical difficulties. Medical management has often shown recurrence of symptoms, particularly in children, and is not considered as an effective choice of treatment. Surgery is the definitive treatment and includes Heller's esophago-myotomy with or without anti-reflux procedures. Prompt diagnosis and treatment usually has excellent prognosis.