Successful pneumatic dilatation of achalasia cardia in a preschool child

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Achalasia is a motility disorder characterized by obstruction at the gastro-oesophageal junction, made worse by lack of peristaltic wave in oesophagus, and failure of inferior sphincter to relax in response to swallowing¹. It is commonly seen in middle-aged females but is uncommon in childhood².

Case report

A 3 year and 10 month old boy was admitted to the university paediatric unit, North Colombo Teaching Hospital, Ragama in March 2001 due to persistent vomiting since age of 9 months. He vomited all that he ate and drank. Nasal regurgitation was noted on many occasions. Vomitus was never bilious or blood stained. His appetite remained voracious, despite these complaints. The child had not had recurrent respiratory infections. He did not have a history of accidental ingestion of corrosives.

He was an only child, a product of a non-consanguineous marriage. He was born at term, with a birth weight of 1.04 kg. His developmental milestones were normal. His weight and height were well below the 3rd centile for his age. He was marasmic, severely dehydrated and showed abnormally aggressive behaviour at presentation. He was not pale; however angular stomatitis was noted. Examination of the abdomen and other systems did not reveal any abnormality.

The complete blood count, blood urea and electrolytes, liver function tests and urine culture did not reveal any abnormality. Ultrasound scan of the abdomen was normal. Upper GI endoscopy did not reveal any specific abnormality and upper GI motility studies could not be performed because of his behaviour. Barium swallow and follow through showed abnormal motility and gross dilatation of oesophagus, with the characteristic tapering "breaking" at the gastro-oesophageal junction, which clinched the diagnosis (Figures 1 and 2).

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The child underwent balloon dilatation of lower oesophagus using a CRG balloon under general anaesthesia. He made a dramatic symptomatic improvement following the first dilatation. His weight started to catch up. He required a second dilatation 3 months later and another 6 months later. Subsequent to that he made an almost complete recovery and his behaviour became normal and was well settled in his pre-school.

Discussion

The incidence of achalasia in childhood is extremely low, fewer than 5% of cases occurring in those under 15 years of age. A few cases had been reported in this country. A survey of paediatric cases by Thomas et al. showed a wide distribution of the age of onset of symptoms with only 18% developing symptoms in infancy. A central abnormality was implicated as opposed to the degeneration of ganglion cells shown in older subjects.

In infantile achalasia the symptoms mimic those of gastro-oesophageal reflux. The vomitus characteristically contains un-curdled milk. Respiratory symptoms predominate in infants and may overshadow the vomiting. Other symptoms include failure to gain weight, blepharospasm and dysphagia. Cases have been reported among siblings and there is an association with adrenal insufficiency and alacrima.

The treatment of achalasia is either dilatation of lower oesophageal sphincter or Heller cardio-myotomy. Pneumatic dilatation is the preferred method for adults and has become increasingly popular as first line management in children as well. This is the first reported case of achalasia in this country who was successfully treated with pneumatic dilatation.

References
