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CHILDHOOD ACHALASIA*

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Achalasia is a motility disorder of the esophagus characterized by absence of normal peristalsis and failure of relaxation of the lower esophageal sphincter. Among the most common clinical findings are vomiting and weight loss. Roentgenographically, a narrowed gastroesophageal junction and dilated esophagus are observed. This condition is uncommon in children. Here a seven-year old female patient with achalasia is reported. *Key words: achalasia, childhood.*

Achalasia is a motor abnormality of the esophagus characterized by inability of the lower esophageal sphincter (LES) to relax[1]. The condition affects primarily adolescents and adults; children under the age of four years comprise less than five percent of patients[2-4]. This seven-year-old female patient is presented because achalasia is very infrequently observed in childhood.

Case Report

A seven-year-old girl was admitted to our hospital with persistent vomiting just after eating, sometimes in the form of saliva, in the previous few months. She was also suffering from weight loss. She had no history of corrosive agent ingestion. She displayed no hematemesis or melena.

Physical examination on admission revealed an exhausted child with bilateral ronflan rales in the respiratory system. The neurological examination was normal. Her weight and height were in the 50th percentile.

Complete blood count, urine and stool analyses were normal. Biochemical analysis of the blood was also normal. Water's x-ray showed right maxillary sinusitis, and the chest x-ray showed bilateral infiltrations in the paracardial area.

The esophagus x-ray showed a narrowed gastroesophageal junction and absence of the propulsive peristaltic wave. There was also a massive esophageal dilatation (Fig. 1, 2). No stricture was noted, and hiatal hernia was not observed. There was no evidence of gastroesophageal reflux on endoscopy.

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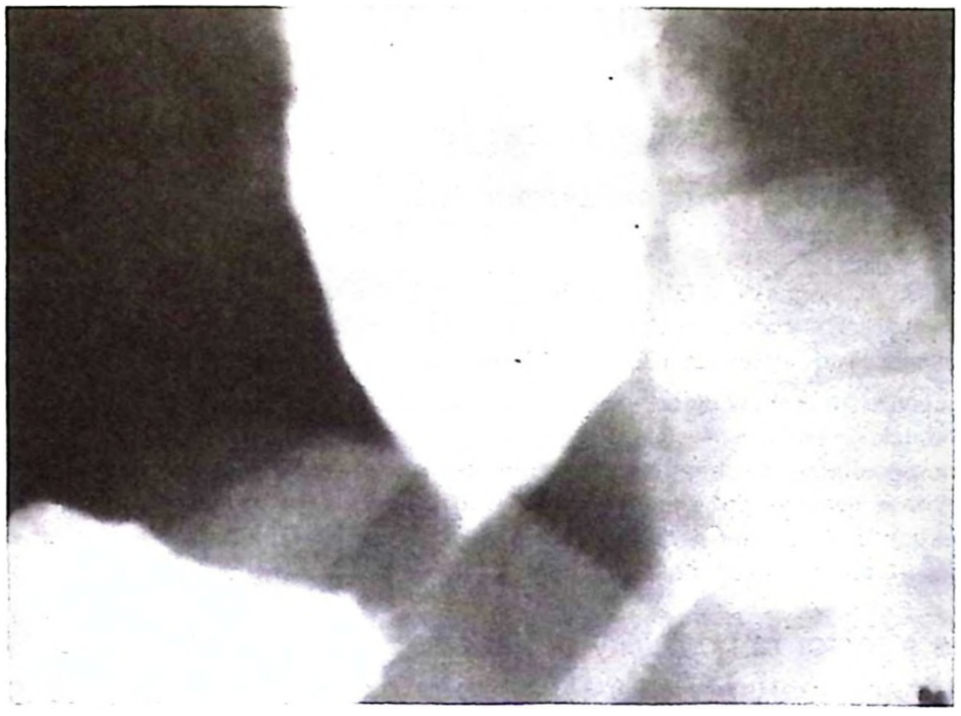

Fig. 1: The typical conical narrowing of the lower end of the esophagus.

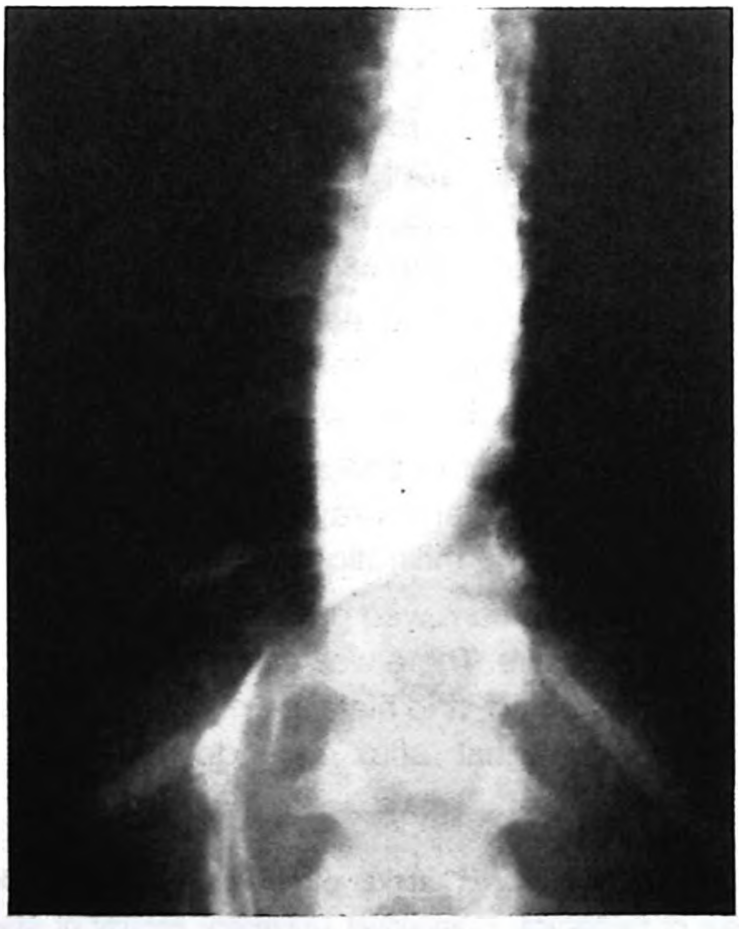

Fig. 2: A barium swallow demonstrating the typical radiological appearance of the esophagus in achalasia.

During her hospitalization, antibiotics were administered for pneumonia and sinusitis. On the seventh day of her hospitalization, the pneumonia resolved and she was referred to the Department of Pediatric Surgery.

Cardiomyotomy combined with a floppy Nissen fundoplication was performed, dramatically relieving the patient's symptoms.

Discussion

Achalasia is usually characterised by loss of peristaltic contractions in the body of the esophagus, and the LES often has increased pressure. As the disease progresses, the esophagus becomes dilated[1], as in our patient.

The incidence of achalasia is estimated to be 0.5-1 per 100,000, with less than five percent of cases presenting in childhood[2,5]. The etiology of the condition is unknown, but selective impairment of the non-adrenergic non-cholinergic inhibitory nerves near the lower esophageal sphincter appears to play a role[6-8]. Ganglion cells are absent from the myenteric plexus in some patients. total absence of ganglion cells in the distal esophagus has been reported in more than 90 percent of patients with achalasia, and denervation of the proximal stomach in half[9].

Familial glucocorticoid deficiency associated with achalasia of the cardia and deficient tear production were described in 1978 by Allogrove et al[10]. This state has been termed "3 A syndrome". Since then there have been a number of reports, including a series of 35 children with achalasia of the cardia by Nihoul-Fekete et al[11]. They determined alacrima and cortisol deficiency in only one child, indicating that most children with achalasia have normal adrenal functions.

It has been reported that if achalasia is associated with absent tear secretion or any other neurological features, cortisol secretion should be tested. Our patient's neurological examination was normal, and there was no evidence of alacrima[12]; consequently, 3 A syndrome was ruled out. Two cases of achalasia secondary to encephalitis have been reported in Turkey[13].

Usually the onset of symptoms is gradual. The diagnosis should be considered in any child with difficulty in swallowing. Often dysphagia of solids occurs before that of liquids, and symptoms such as weight loss, vomiting, and aspiration pneumonia may not be present in the early stages of the disease[9]. Our patient suffered from vomiting at first, and had complaints of weight loss and aspiration pneumonia.

The diagnosis is usually made roentgenographically[2]. The use of barium is diagnostic in all patients, demonstrating the typical features of achalasia: dilated body of the esophagus, absence of normal peristalsis and the typical narrowing at the esophagogastric junction[1,2]. As seen in Figs. 1 and 2, all of the evidence was present in our patient's radiography.

Treatment of this disorder consists of disrupting the failure to relax by the LES[1,2]. Drugs such as nitrates or calcium antagonists, particularly nifedipine, cause a significant decrease in sphincter pressure and may produce immediate relief of symptoms. However, the effect is most often short-lived and rarely avoids the need for surgery[14,15].

In adults, pneumatic dilatation is the recommended initial treatment, while in children this technique has been less successful and surgery is the preferred option[11,16]. Our patient, whose general condition was good, was subjected to cardiomyotomy combined with a floppy Nissen fundoplication.

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