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Two Cases of Achalasia (Mega-Oesophagus), Secondary to Encephalitis

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Achalasia is a generalized disorder of oesophageal motility. The condition has been described by various terms, some of which, such as "Cardiospasm", are misleading; the term achalasia, indicating a failure relaxation, was introduced by Hurts in 1914. In recent years, manometric and radiologic studies have shown that the essential features of the condition are the inability of the lower end of the oesophagus to relax, and the inability to swallow associated with the disorder of the normal peristaltic waves in the body of the oesophagus. In a normal subject, the lower end of the oesophagus relaxes about 1.5 - 2.5 sec. after swallowing begins. This relaxation has been shown, experimentally, to depend upon the integrity of both the vagus and the myenteric ganglia.[1 2 3 4] In achalasia, the motor disturbance is due to a lesion of the myenteric nerve plexus and possibly also to a lesion of the vagus of unknown cause.[1]

We detail here two cases of achalasia seen at our hospital within an interval of 2 months.

Cases

Case 1. An 8 year-old-boy came to our hospital complaining of difficulty in swallowing, vomiting, constipation, cough, difficulty in speaking and failure in growth and development. He had had encephalitis when he was one year old (we have observed this point in his

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medical record), after which his complaints started and gradually became more severe. Physical, neurological, and X-ray examination revealed mental retardation, growth failure (under 3%), exaggerated tendon reflexes and positive Babinski on both sides, flouroscopic examination of the oesophagus revealed nonpropulsive peristalsis and a lack of relaxation of the vestibule. X-ray examination showed (see Figure 1) megaoesophagus (dilated and elongated), conical narrowing of the lower end of the oesophagus, and absence of air in the stomach. He was successfully dilated with the use of pneumatic balloons, and since dilatation, he has been doing well.

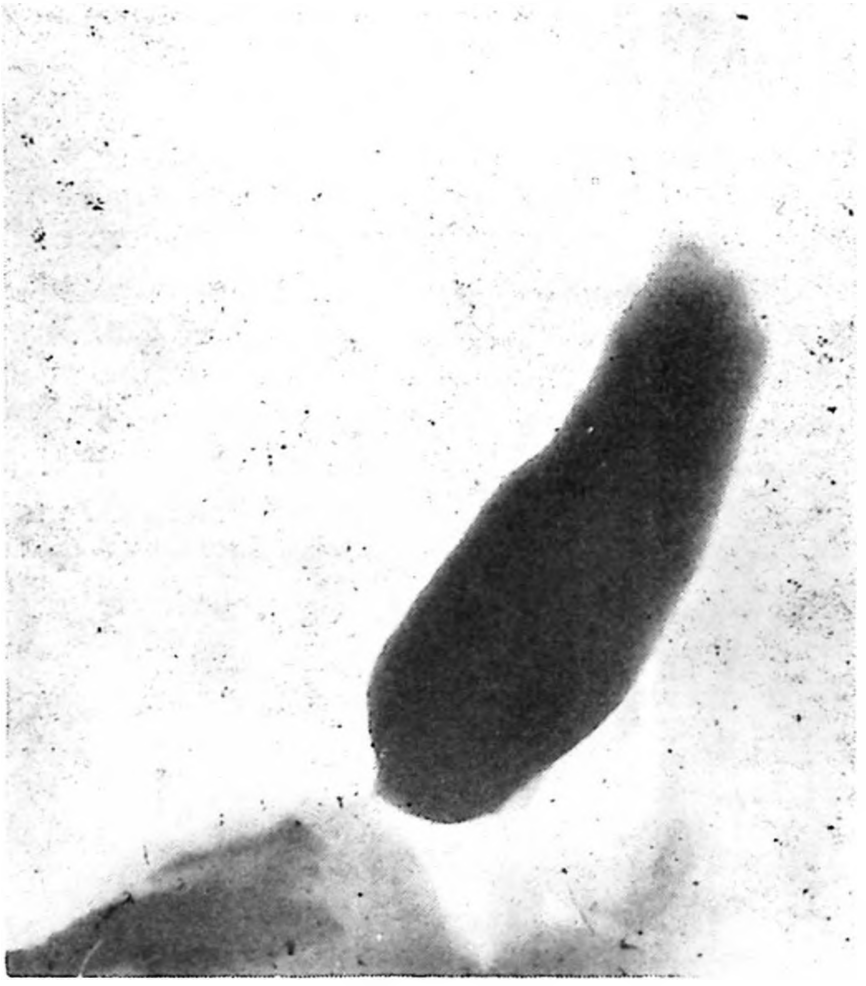

Figure 1

Case 2. A six year-old-boy was admitted to our hospital complaining of difficulty in swallowing, vomiting, failure in growth and development, difficulty in walking and talking. He was well until 4 months of age, when he had encephalitis (He was unconscious, had seizures for 10 days; neurological examination and laboratory findings had shown encephalitis.). He could hold his head up at 6 months of age, sat at 3 years of age and started imperfect walking at the age of 5. He could not speak yet, excepting a few simple words.

Vomiting had begun after he had encephalitis and gradually became severe. Physical examination revealed mental retardation, growth failure, imperfect walking, slight spasticity in all extremities, exaggerated tendon reflexes, positive Babinski on both sides and scrotal hernia on the right. Flouroscopic examination revealed disorganized motility and a failure of relaxation of the vestibule. X-Ray findings (see Figure 2) were mega-oesophagus and conical narrowing of the vestibule and no air in the stomach. Oesophagoscopy was performed and revealed dilated oesophagus, narrowing vestibule, oesophagitis and an ulcer on the wall of the oesophago-gastric junction. A narrow dilator was passed through the vestibule into the stomach without difficulty but a larger one could not pass. Biopsy: No plexus is detected on the oesophageal wall from the specimen which we sent. The patient had the Heller operation on 9/9/1969. Operation confirmed our preoperative diagnosis.

Figure 2

As you see from Table I, both patients had encephalitis in the past, both were vomiting and had difficulty in swallowing before treatment, both were mentally retarded, underweight, slightly spastic

TABLE I

SUMMARIZING TWO CASES

Case	History	Neurological Symptoms				Symptoms of Achalasia				
	Past history	Mental retardation	Spasticity	Tendon reflexes	Babinski sign	Vomiting	Difficulty in swallowing	Growth failure	X-Ray α flouroscopic findings	Oesophagoscopic findings
1	Encephalitis	+	Slight	Exaggerated	+	+	+	+ under 3 %	Typical	Typical
2	Encephalitis	+	Slight	Exaggerated	+	+	+	+ under 3 %	Typical	Typical

with exaggerated tendon reflexes and positive Babinski signs. X-Ray, flouroscopic and oesophagoscopic findings were typical for Achalasia.

Discussion

Achalasia in childhood and infancy appears to be a rare condition (4 percent - 5 percent) in oesophageal surgical cases.[5 6]

The cause of this condition still remains unknown in etiology. According to some authors it could be a congenital disorder[5 7] or originally acquired as a cause of C.N.S. pathology,[8] such as encephalitis, hydrocephaly etc. Etzel reported 626 cases of achalasia due to nutritional deficiency (Vit. B_1) associated with mega-ureter and mega-colon. In Chagas disease, mega-colon and mega-oesophagus are seen. In children, emotional factors may precipitate the symptoms of Achalasia.

Although the cause of achalasia is unknown, pathogenesis of the motor disability of the entire oesophagus is clear. The derangement of motor behavior in Achalasia is due to degeneration and reduction in the number of ganglion cells at all levels of the oesophagus. The loss of ganglion cells may be a secondary phenomenon rather than the primary cause of the disease.[4]

Avery Jones[1] has mentioned an experimental study according to which reduction of the number of nerve cells in the dorsal motor nuclei of vagus produced similar changes in the Auerbach plexus of the oesophagus. This study suggested that the degeneration of the myenteric plexus is secondary to a lesion of the vagus at the level of the brain-stem.

Roth[3] has reported an experimental study of the high bilateral cervical section of the vagus. Low transaction of the oesophagus abolished the apparent reflex relaxation of the cardia produced by distention of the body of the normal oesophagus in dogs. Relaxation of the vestibule thus appears to depend upon intact parasympathetic innovations.

Josepp G. Chusid[2] included difficulty in swallowing and regurgitation among the symptoms and signs suggesting tenth nerve damage and he also mentioned that oesophageal, cardiac or pyloric spasm not due to local causes may be of vagal origin.

Meisner[9] has divided the pathogenesis of achalasia into two groups, achalasia is a congenital disorder, or it may be an acquired disorder secondary to C.N.S. pathology.

Summary

In the light of our two cases and the review of the medical literature in the pathogenesis of achalasia, a lesion of the brainstem or of the dorsal nucleus of the vagus and its preganglionic fiber may be a cause of motor disturbance of the entire oesophagus, as much as a lesion of the Auerbach's plexus itself.

Therefore we believe that our two cases of achalasia secondary to encephalitis confirm this observation.

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