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INTRAMURAL TRACHEOBRONCHIAL REMNANTS: A MISDIAGNOSED CAUSE OF CONGENITAL ESOPHAGEAL STENOSIS

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ABSTRACT

In cases of congenital esophageal stenosis due to tracheobronchial remnants, symptoms of partial esophageal obstruction appear at the time of weaning or during early childhood. In the absence of esophagitis, esophagram combined with cinefluoroscopy demonstrates fixed stenosis of distal esophagus and appears to be diagnostic. Dilatation of rigid stenosis is invariably unrewarding and surgical resection is mandatory. Pathologically, the presence of cartilage or respiratory seromucinous glands in the esophageal wall are pathognomonic features. Other congenital anomalies may be encountered in some cases (30%).

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INTRODUCTION

Congenital esophageal stenosis is a rare cause of esophageal obstruction, causing regurgitation or dysphagia during infancy, which is in most instances erroneously confused with other common causes of infantile esophageal obstruction. Three types of congenital esophageal stenosis have been described:[1] (1) segmental stenosis, (2) membranous webs, and(3) intramural rests of tracheobronchial remnants. The latter is the least common type and results in rigid stenosis because of encircling cartilaginous rings. We present here the details of such a case and carefully analyze 29 similar cases reviewed from the literature,[2-9] to identify the diagnostic features of this entity.

CASE REPORT

A 17-month-old female was admitted to this hospital on 15th March, 1988 with the chief complaint of regurgitation after meals since the age of six months when semisolid foods were added to her formula. On examination she was underweight and anemic. No other congenital anomalies or significant physical findings were noticed. Meticulous work up of her complaint by several physicians during this long period by frequent barium studies and endoscopic examination could not establish any definitive diagnosis. Endoscopic evaluation demonstrated a rigid stenosis at the distal end of the esophagus. No evidence of reflux esophagitis or hiatal hernia was found. Endoscopic esophageal biopsy revealed normal mucosa without any evidence of inflammation or fibrosis, although it was difficult to take biopsy from the actual stenotic portion. Barium swallow demonstrated an abrupt narrowing at the distal end of the esophagus with proximal dilatation (Fig. 1). Fluoroscopy demonstrated fixed stenosis and no peristaltic movements were noticed on swallowing. As previous attempts of dilatation yielded no improvement,the patient was referred for surgery.

The esophagus was explored through a left anteromedial thoracic incision and a firm stenosis in the distal end of the esophagus just above the diaphragm was found. Stenosis was too rigid to be dilated by a Foley catheter, therefore the stenotic portion of the esophagus was resected and an esophago-gastric anastomosis was performed in one layer. Postoperative course was uneventful. Later on an esophagram

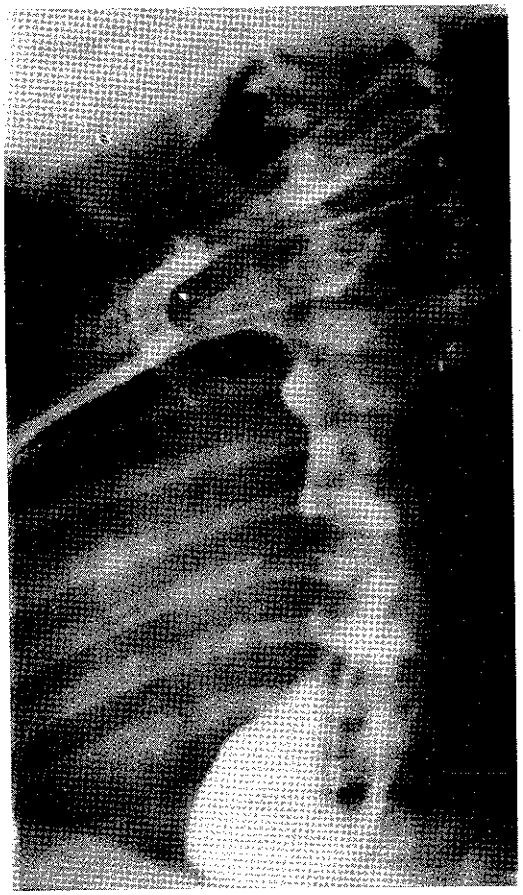

Fig.1. Esophagram shows esophageal dilatation proximal to an abrupt esophageal stenosis at distal end.

revealed a satisfactorily patent anastomosis. The child did well during the past nine months after operation. She was taking normal diet without any difficulty and gained weight.

Pathologic examination

The grossly stenotic part of the esophagus was firm with an external diameter of 8 mm. On transverse section internal diameter was 3 mm. Macroscopically, the mucosa was found to be normal and a firm area was felt in the esophageal wall but it was difficult to identify cartilage. Microscopically, in the stenotic zone, however, the architecture of the esophageal wall was grossly disorganized but mucosa and lamina propria were unremarkable. The submucosa and inner muscular layer contained a plate of hyaline cartilage extending over one third the circumference of the wall. Many seromucinous glands were scattered in the submucosa and deep into disorganized muscular layers (Fig. 2). Some small cysts lined by columnar epithelium, sometimes pseudostratified and ciliated and surrounded by a lymphocytic cuff, were associated with the glands. Above and below the lesion the esophagus was normal without any evidence of fibrosis.

DISCUSSION

Careful review of the literature revealed that despite the rarity of tracheobronchial remnants in the esophageal wall, it appears to be one of the most

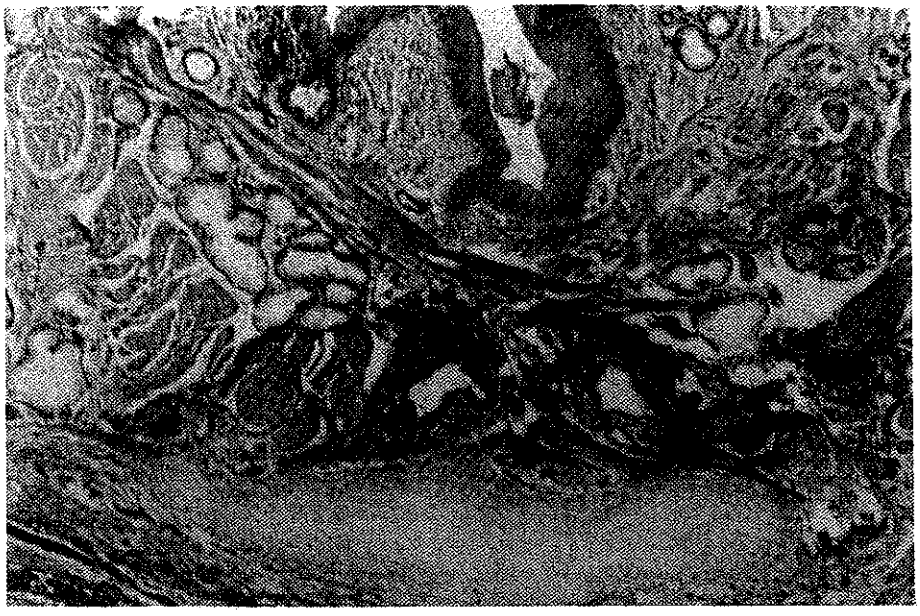

Fig.2. Transverse section of stenosed zone of esophagus contained cartilage, seromucinous glands and small cyst-like structures surrounded by lymphocytes (H & E) × 60.

Table I.

Ref.	Age/Sex	Age at onset of symptoms	Site of stenosis	Other anomalies	Major histologic components: cartilage	glands	resp epith. + lymphoid tissue	Operative procedures
2.	19yrs/F	?	Distal end	---	+	+	+	Postmortem diagnosis.
3.	43 yrs/M	28 yrs	?	---	–	–	+	Extramuralexcistion
4.	52 yrs/M	3 yrs	distal end	---	+	+	+	Resection
5.	57 yrs/F	childhood	lower third	---	–	+	+	Resection
6.	49 yrs/F	Birth	lower third	---	–	+	+	Resection
7.	10 mo/F	4 mo	distal end	---	+	+	–	Resection
8.	8 mo/F	4 mo	lower end	---	+	–	+	Resection
	14 mo/F	6 mo	distal end	---	+	+	+	Extramucosal excision
9.	17 mo/M	7 mo	distal end	---	+	+	+	Resection
	4 yrs/M	6 mo	distal end	---	+	+	+	Resection
	5 yrs/M	1 yr	distal end	---	+	+	–	Resection
10.	1 yr/F	6 mo	lower third	---	–	+	+	Resection
11.	1 day/F	Birth	upper third	Esophageal atresia	+	+	–	Cervicaloesophagostomy
12.	8 yrs/F	7 mo	distal end	Multiple*	+	–	+	Resection
13.	2 yrs/F	6 mo	lower third	Multiple**	+	+	–	Resection with coloninterposition
	10 mo/M	5 mo	lower third	Multiple***	+	+	–	Postmortem diagnosis
14.	4 yrs/M	18 mo	lower third	---	+	–	–	Resection
	20 mo/M	9 mo	distal end	---	+	+	+	Resection
	19 mo/M	4 mo	distal end	---	+	+	+	Resection
	4 yrs/M	8 mo	distal end	---	+	+	?	Resection
	13 mo/F	7 mo	distal end	---	+	+	?	Myomectomy
15.	7 mo/?	7 mo	distal end	---	+	+	+	Resection
	8 yrs/F	Birth	lower third	Down's syndrome	+	+	+	Resection
16.	20 mo/F	6 mo	distal end	---	+	+	+	Resection with Nissen
17.	6 mo/M	2 mo	distal end	Hypospadias	+	+	+	fundoplication
18.	6 mo/M	6 mo	lower third	---	+	+	+	Resection with
	6 yrs/F	6 yrs	lower third	Multiple****	+	+	+	pyloroplasty
19.	15 mo/F	14 mo	distal end	Anovestibular fistula	+	+	?	Resection
	20 mo/M	Birth	distal end	Microphthalmoswith iris coloboma	+	+	+	Resection
								Resection
Present case	17 mo/F	6 mo	disal end	---	+	+	+	Resection
								Resection

*Small ventricular septal defect, pulmonic valvular stenosis and physiologic two chambered right ventricle.

**Esophageal atresia and tracheo-esophageal fistula

***Tracheo-esophageal fistula, esophageal artesia and rectal agensis

**** Esophageal atresia, tracheo-esophageal fistula, duplication of duodenum and pancreatic cyst.

common causes of congenital rigid stenosis of the distal part of the esophagus during early childhood. Most cases have been reported from Japan,[19] however they were often misdiagnosed and resulted in delayed or mismanagement despite their distinct clinicopathologic features. Most cases presented during the pediatric age group (83.4%) (less than 6 months 3.3%, 6-24 months 56.6%,2-8 years 23.3% and more than 8 years 16.6%). Interestingly in Japanese cases males comprised 80% of cases, while in cases from other parts of the world 74% of patients were female. The most common presenting symptoms were dysphagia (83%), regurgitation (61%), or both, usually beginning at the time of weaning (53%), otherwise during early childhood (96%). Noticeably in cases presenting during adulthood the cartilage was absent from lesions (Table I) with few exceptions.[2,4] Eighty three percent of children were found to be underweight and malnourished because of frequent vomiting and dysphagia. Pulmonary infections were noticed in 23.3% of cases at the time of presentation.[8,10,13-15] Other congenital anomalies were encountered in 30% of cases (Table I). Nishana et al[19] reported associated anomalies in 17.3% of cases and stated that the evidence of associated anomalies in congenital esophageal stenosis due to tracheobronchial remnants was significantly lower than that in esophageal atresia. Tracheoesophageal fistula, esophageal atresia, Down's syndrome and anovestibular fistula were the most frequently noticed anomalies associated with tracheobronchial rests in the esophagus.[19]

The esophagram invariably demonstrates characteristic narrowing of the distal portion of the esophagus with proximal dilatation. In some cases

linear tracks of barium representing ducts of tracheobronchial glands that extend from the stenotic area were noticed.[4,12,15] In contrast to achalasia, cinefluoroscopy revealed fixed stenosis.[10,15] Endoscopic findings were available only in 13 cases and all patients demonstrated firm and rigid stenosis without any evidence of reflux esophagitis or hiatal hernia. Dilatation was attempted in 15 cases without rewarding results and proved fatal in one case.[13] The most common erroneous diagnoses were achalasia,[7-9,14,16,17] and inflammatory strictures.[12,13,18] While other reports did not mention any specific clinical diagnosis, in none of the reported cases was tracheobronchial rest suspected the cause of stenosis.

The grossly stenotic part was harder than a fibrotic stricture, measuring 1-5 cm in length and with an internal diameter of up to 3 mm. No mucosal ulceration was found. Microscopically, the lesion characteristically comprised of seromucinous glands (86.6%) and small linear or crescentic plates of hyaline cartilage (86.6%) either encircling the whole circumference of the esophagus or part of it, as seen in the tracheobronchial tree. Usually, cyst-like structures lined by respiratory epithelium with lymphoid mantle (66.6%) and sometimes connected to the esophageal lumen through ducts were also present. In cases without cartilage, respiratory seromucinous glands were the convincing evidence to consider them as tracheobronchial remnants and resulted in less severe stenosis with delayed presentation [2,5,6] Ibrahim, et al[18] considered lymphoid aggregates as part of a developmental anomaly rather than inflammation.[11] In many cases patches of stratified ciliated columner epithelium were found in the esophageal mucosa, which may present normally at birth [20] Reports with detailed microscopic descriptions of lesions including ours stressed on disorganization of the musculature of the stenotic esophagus, where tracheobronchial remnants were present as one of the major abnormalities.[15,18] This may contribute to fixed stenosis demonstrated by cinefluoroscopy.

Differential diagnosis between congenital and acquired stenosis as well as achalasia of the cardia is of great importance as the treatment has to be arranged accordingly. The existance of other deformities, especially stenosis and atresia of the gastrointestinal tract, suggest the presence of congenital esophageal stenosis. An esophagram combined with cinefluoroscopic evaluation of esophagus was the most useful diagnostic measure to differentiate it from achalasia.

Stenosis due to tracheobronchial remnants does not dilate with swallowing and remains fixed,[10] while in achalasia absence of stripping wave and uncoordinated esophageal contractions may be demonstrated.[21] Endoscopic examination and biopsy confirms the absence of esophagitis and hiatal hernia and rules out inflammatory strictures. Unlike achalasia, esophageal webs, and fibromuscular stenosis, stenosis due to tracheobronchial remnants was resistant to dilatation; and invariably unable to relieve the obstruction, Sneed et al[16] recommended surgical resection of the stenotic segment coupled with an antireflux procedure, if the gastroesophageal junction was removed. Overall, the prognosis was excellent if treated in proper time.

According to the most popular theory, its
ment is related to the defective separation of
embryonic respiratory tube from the primitive
during early embryonic stage, resulting in
tion of tracheobronchial precursor cells in the wall
the esophagus and located in the distal part of
esophagus because of the differential growth between
the esophageal and respiratory tubes.[7-9,11,18,19]

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