Is Secondary Achalasia A Reality? Primary Versus Secondary Achalasia Role of Barium Swallow

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This study was conducted in the Department of Radiology Mayo Hospital, Lahore from January 2002 to December 2002. 30 Patients with findings on barium swallow of Achalasia were taken. Post procedure follow up was carried out either in the shape of endoscopic biopsy or post operative biopsy. The results showed that 25 out of 30 had Primary Achalasia and 5 had Secondary Achalasia. The results were compared with age of the patients and it was found that Secondary Achalasia, which was due to carcinoma of the lower esophagus or fundus of stomach, was virtually non existent before the age of 30. These were 5 patients all above 30 years of age.
Key words. Primary Achalasia, Secondary Achalasia, Barium Swallow, Endoscopic Biopsy

Department of Radiology of Mayo Hospital is a very busy unit and it is very easy to find patients of dysphagia in the fluoroscopic room. In relatively younger age group the causes of dysphagia get narrowed down and Achlasia is a very common disorder in these patients. The need to do this sort of study arose due to the fact that previously reported Primary Achalasia was tried to be dilated and it was found to be due to a malignant tumor. Barium swallow is the primary investigation in these cases and shows characteristic appearances.

Patients and method

30 patients referred from the ENT, Medicine and Surgical ward were taken which had narrowing of the lower end of esophagus below the diaphragm. All of them had been performed barium swallow. The age group ranged from 15-65 years of age. Study was conducted in the Department of Radiology Mayo Hospital from January 2002 to December 2002. It was a descriptive study in which convenient sampling was done. The X ray machine used was Toshiba 500mA fluoroscope with image intensifier. Barium Swallow was done with double contrast technique in erect posture. Post procedure follow up was carried out in the form of either Endoscopic biopsy of surgical exploration.

Results

Out of 30 patients 25 had Primary Achalasia and 5 had Secondary Achalasia. The cause in all 5 of secondary Achalasia was malignant infiltration of the walls of lower end of esophagus and the lower esophageal sphincter.

Table 1. Percentage of the number of patients

	n=	%age
Primary Achalasia	25	83
Secondary Achalasia	5	17

Table 2. Age of the Patient and Achalasia

Age of Patient	Primary Achalasia	Secondary Acalasia
Below 30 years	10	None
Above 30 years	15	5

The results were also compared in the age group form and it was found that secondary Achalasia was virtually non existent before the age of 30 years as shown in Table 2.

Discussion

Achalasia is an esophageal motor disorder characterized by increased lower esophageal sphincter (LES) pressure, diminished-to-absent peristalsis in the distal portion of the esophagus composed of smooth muscle, and lack of a coordinated LES relaxation in response to swallowing[1,2,3].

Primary achalasia is the most common subtype and is associated with loss of ganglion cells in the esophageal myenteric plexus. These important inhibitory neurons induce LES relaxation and coordinate proximal-to-distal peristaltic contraction of the esophagus[1,4,5].

Secondary achalasia is relatively uncommon. This condition exists when a process other than intrinsic disease of the esophageal myenteric plexus is the etiology. Examples of maladies causing secondary achalasia include certain malignancies, diabetes mellitus, and Chagas disease[3,5,6,7].

Sir Thomas Willis first described achalasia in 1674. Willis successfully treated a patient by dilating the LES with a cork-tipped whalebone. Not until 1929 did Hurt and Rake first realize that the primary pathophysiology resulting in achalasia was a failure in LES relaxation[1].

The exact etiology of achalasia is not known. The most widely accepted current theories implicate autoimmune disorders, infectious diseases, or both. The last decade has witnessed much progress in the understanding of the cellular and molecular derangements in achalasia[1,4].

Degeneration of the esophageal myenteric plexus of Auerbach is the primary histologic finding. However, with early achalasia, a mixed inflammatory infiltrate of T cells, mast cells, and eosinophils is found in association with myenteric neural fibrosis and with a selective loss of inhibitory postganglionic neurons from the Auerbach plexus. In these patients with early achalasia, neurons of the myenteric plexus are relatively well preserved[2].

The inhibitory neurons produce nitric oxide (NO) and vasoactive intestinal peptide (VIP). NO and VIP are inhibitory neurotransmitters responsible for relaxation of the LES and for coordinated esophageal peristalsis. The loss of inhibitory neurons allows unopposed excitatory stimulation by postganglionic cholinergic neurons of the Auerbach plexus, which leads to a failure in LES relaxation and, eventually, to aperistalsis of the distal esophagus due to loss of the esophageal body latency gradient. Essentially, this means that this portion of the esophagus is unable to relax and subsequently generate a proper, sequential peristaltic wave[5]. The radiologic examination of choice in the diagnosis of achalasia is a barium swallow study performed under fluoroscopic guidance.

A diagnosis of achalasia supported by the results of radiologic studies must always be confirmed by performing upper gastrointestinal endoscopy and esophageal manometry. These tests allow the direct evaluation and inspection of the esophageal mucosa and an objective measurement of esophageal contractility.

Endoscopy, supplemented by biopsy when necessary, helps in excluding gastroesophageal malignancies, fungal or bacterial infections, and other disease processes that can mimic achalasia. Features of achalasia depicted at barium study under fluoroscopic guidance include the following:

Failure of peristalsis to clear the esophagus of barium with the patient in the recumbent position. Antegrade and retrograde motion of barium in the esophagus secondary to uncoordinated, nonpropulsive, tertiary contractions .Pooling or stasis of barium in the esophagus when the esophagus has become atonic or noncontractile (which occurs late in the course of disease) .LES relaxation that is incomplete and not coordinated with esophageal contraction. Dilation of the esophageal body which is typically maximal in the distal esophagus. Tapering of the barium column at the unrelaxed LES, resulting in the bird beak sign .Associated epiphrenic diverticula (possible finding)[7,8,9].

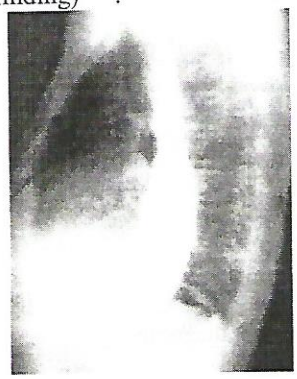

Fig 1. Turned out to be Secondary Achalasia

Fig 2 Classical Primary Achalasia

A pseudoachalasia from carcinoma involving the cardia and gastroesophageal junction may be difficult to differentiate from achalasia. Motor abnormalities in the esophagus result from tumor infiltration of the esophageal wall and resultant nerve damage. If mucosal irregularity or mass effect is present at the tapered gastroesophageal junction, pseudoachalasia should be considered. The use of amyl nitrite can often be helpful, as the LES relaxes in achalasia but remains fixed in pseudoachalasia[9,10].

Conclusion

All Achalasia are not Primary Achalasia. There is a definite entity known as Secondary Achalasia which has a major group comprising of Malignancies. And Primary Achalasia is more common in younger age group rather that older ones.

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