Achalasia: symptoms and radiology

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Summary

A radiographic and symptomatic study of 280 patients with achalasia before and after treatment is presented including diagnostic features of the symptom pattern and radiographic findings.

THE essential disturbance in achalasia is a failure of the cardiac sphincter to relax adequately when the patient swallows. This causes the oesophageal tube to be closed to both liquids and solids by an elastic process that can be forced open a varying amount by building up pressure in the oesophagus, in addition there is damage to some of the postganglionic neurones in the wall of the oesophagus so that not all the impulses sequentially emitted from the medulla get through to the oesophageal muscle and the peristaltic wave is partially or wholly disorganized. Spasm is a word to be avoided because it has no useful meaning; diffuse spasm is a form of achalasia, and corkscrew oesophagus is either a form of achalasia or an achalasia-like picture probably caused by senile changes in the postganglionic neurones of the oesophageal wall. Achalasia does not always cause significant dysphagia, and it does not always produce an enormously dilated oesophagus. It is rarely a progressive disorder; the damage to the postganglionic neurones seems to happen over a short period of time and changes in the size of the oesophagus and the symptoms seem to depend on the severity and persistence of the obstruction. If the obstruction is relieved early enough the oesophagus may return a long way but never the whole way towards a normal shape and activity.

Symptoms

The oesophageal tube is closed to liquids and to solids and dysphagia for both characteristically develop at the same time. Often the difficulty is first evident with lumpy food, or with gassy drinks. Occasionally solids may be more difficult than liquids for a while and sometimes the patient develops the habit of gulping water to force food into the stomach. They are almost always aware however, that if they drink quickly they are building

up pressure within the oesophagus and drinking is not normal.

If much of the peristaltic wave persists or the wall contracts violently the patient may experience pain sometimes after swallowing solids not unlike the bolus obstruction syndrome of stricture. More commonly there is no distinct impact sensation nor any pain when food sticks, but a vague awareness that it is not moving on. Because the closed segment can be forced, many patients can eat most of a meal if they cut it and chew it well enough and eat it very slowly in contradistinction to carcinoma, where the dysphagia for solids is absolute if there is any difficulty with liquids.

A curious and characteristic symptom in achalasia is the spontaneous pain which occurs in about 75% of patients. The pain may be mild or agonizingly severe and very frightening, is usually called indigestion or heartburn or spasm, and may occur many times a day or at intervals of days, weeks or months. It occurs at any time of the day or night, lasts a few minutes to several hours, and may extend over the full oesophageal distribution of gums, ears, arms, fingers and chest. It is not provoked by eating or drinking or by any position, has nothing to do with reflux of gastric contents onto the oesophageal mucosa, and does not appear to be produced by muscular contraction in the oesophagus. It is usually relieved by drinking cold water, is not influenced by 'antispasmodic' or anticholinergic drugs, is not cured by cardiomyotomy but is usually less frequent and less severe, and frequently precedes dysphagia by months or years. Its mechanism is a mystery. Four types of pain or discomfort can be clearly distinguished by the patient who experiences all of them; the other three are heartburn due to reflux, pain due to distension when drinking fast, and pain due to impaction of food at the sphincter.

Patients commonly regurgitate (not vomit, although they may so call it) much saliva and over a period of minutes may produce a cupful or more indicating that the oesophagus has a large capacity. Food tends to sink in the bag of saliva and is regurgitated later, milk tends to float. The regurgitate may taste of nothing in particular, or may be fermented, or foul, but does not taste of vomit.

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Radiology

If the oesophagus is examined correctly there should be no difficulty in diagnosing achalasia. Any disorder which prevents the free flow of barium into the stomach when the patient is drinking erect, together with a disorganization of the peristaltic wave viewed in the 10° head down position belongs to the syndrome of achalasia whatever the size of the lumen and the degree of activity of the wall. Obstruction to liquid by cancer makes the wall of the distended oesophagus appear to behave abnormally and the persisting peristaltic nature of the contraction may be difficult to see. If the oesophagus is emptied and a single mouthful of barium is swallowed against gravity, the behaviour of the wall can be distinguished. Supine, head down, the achalasic

wall shows irregular disorganized spontaneous contractions which distinguish it from the dilated oesophagus of systemic sclerosis which has a peristaltic type of contraction if any contraction is visible, and is characterized by the absence of effective sphincter contraction, so that when drinking erect the barium flows easily into the stomach. The lesion is loss of muscle rather than loss of neurones. The tapering 'rat-tail' end of the barium column writhes and squirms in achalasia, constantly changing the shape of its shoulders, and sometimes the lower 1 or 2 cm of the oesophagus is separated off from the tube above by an irregular segmented contraction which may be mistaken for a small hiatal hernia.