Not... achalasia

Achalasia is a rare disease; disorders which imitate it are even less common, with the exception of Chagas’s disease, which should not be overlooked easily if the patient is questioned about travel to South America. Achalasia may be imitated, however, by a neoplastic tumour in or close to the lower oesophagus—a syndrome first described over 60 years ago and long known as a diagnostic trap for the unwary clinician or radiologist. The term “secondary achalasia,” sometimes used, is misleading as the mechanism may be different from the presumed neural abnormalities which cause true achalasia. If eponyms were fashionable it might be called Howarth’s syndrome.

Most cases are due to adenocarcinoma of the cardia,1-4 for when a carcinoma encircles and narrows the oesophageal outlet the radiological appearances are like those of achalasia, since such a constriction rapidly abolishes oesophageal peristalsis, which is replaced by “spasm”—as shown by Kelley in man6 and by our unpublished studies in monkeys. In some case reports, however, the adenocarcinoma did not completely encircle the cardia, and this was usually the case when other malignant tumours were responsible; these other lesions included bronchial carcinoma,6 reticulum cell sarcoma,7 gastric lymphoma,8 and pancreatic carcinoma.8 In all such cases the primary or metastatic growths were close to the distal oesophagus. When the histological appearances have been reported Auerbach’s plexus has been infiltrated by tumour,9-11 sometimes localised to a short segment close to the gastro-oesophageal junction.12 This adds some weight to the belief that true achalasia may also begin with neural abnormalities localised to the cardia, the motor changes in the body of the oesophagus being secondary.

Benjamin et al reported two patients with widespread lymphoma (including spread to the central nervous system) with dysphagia due to oesophageal motor abnormalities, though not simulating achalasia.13 They speculated that the abnormality in motility might originate in the central nervous system but produced no anatomical proof that the oesophagus itself was not directly affected.

Awareness of this unusual abnormality is important because it is so easy to believe that a patient has true achalasia—the rarity of that condition bemusing the unwary diagnostian. Tucker et al reported that patients with “secondary achalasia” tend to be over 50, with appreciable loss of weight and dysphagia for less than a year1; but this is not always so,7 and such a history occurs quite often in true achalasia.14 These features should, however, always put the clinician on his guard. The radiologist may find a smoothly tapered narrowing of the cardia, sometimes with dilatation of the body of the oesophagus; and, though with care he may see distortion of the fundus or rigidity of the narrow segment,8 especially if cine radiographs are studied, the appearance may be indistinguishable from true achalasia, even on review.

Endoscopy is obligatory in any patient with dysphagia, and this is just as true when the radiological diagnosis of achalasia seems obvious. If there is dilatation and retention of food daily washouts of the oesophagus and a diet of clear fluid only may be necessary for two or three days if an adequate view is to be obtained. In achalasia the cardia opens to firm pressure by the endoscope, the mucosa is smooth, and the gastric fundus is normal when an inversion view is obtained. Yet Tucker et al showed that carcinoma of the stomach may be overlooked, even when specimens are taken for histological and cytological examination,4 and extrinsic carcinomas may be even harder to detect. It might be thought that intraluminal manometry—the touchstone of the oesophagologist—would be the most accurate arbiter, but in reported cases the findings (aperistalsis and a high pressure, poorly relaxing sphincter) have been identical with those in true achalasia. It is more clinically gratifying than it is physiologically surprising that oesophageal function may return to normal when the neoplasm is removed or treated by radiotherapy or chemotherapy, and the patient will be given useful symptomatic improvement even when the lesion is itself incurable.5 7 8 13

Until recently almost all achalasia was treated surgically in Britain, so that differentiation of true achalasia from a tumour of the cardia was less important since the true state of affairs would inevitably be discovered at operation. Now that achalasia is increasingly being treated by forceful pneumatic dilatation, accuracy of diagnosis has become of greater importance. If there is doubt, then operative treatment should probably be recommended: if dilatation is inadvertently attempted a tumour at the cardia may be manifested by its lack of distensibility, the bag failing to reach its proper inflated outline under full pressure when viewed radiologically.

An uncommon mimic of a rare disease will not tease a clinician with any frequency, but those who deal with the oesophagus should be alive to the problem and make certain that their diagnostic approach ensures the minimum possibility of error.

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The heart in the Guillain-Barré syndrome

The Guillain-Barré syndrome, or Landry's ascending paralysis, is an acute immunologically mediated predominantly motor neuropathy. It is characterised by a raised protein concentration without an increase in the cell count in the cerebrospinal fluid. The disease progresses for between two and four weeks, during which the flaccid motor paralysis tends to ascend, hence Landry's eponym. If the chest muscles are affected it may be necessary for the patient to be ventilated. In some cases disease goes on to affect the cranial nerves.

The autonomic nervous system is responsible for moment to moment regulation of heart rate, the force of myocardial contraction, and the capacitance of the blood vessels and their resistance to forward flow. The cardiac nerves therefore control cardiac output, arterial pressure, and perfusion of the tissues. The sympathetic and parasympathetic preganglionic cells receive both excitatory and inhibitory impulses from the cardiovascular centres in the medulla and from spinal neurones.

Autonomic dysfunction may occur in the syndrome, particularly in those who show weakness of the respiratory muscles. Vasomotor control may thus be disturbed, sometimes with hypertension (perhaps because the carotid sinus is affected) and sometimes with postural hypotension. Abnormal sweating and pupillary disturbances may occur. Patients with the Guillain-Barré syndrome may die suddenly, and since necropsies have not shown a cause and arrhythmias are a recognised complication of the disease, death has been attributed to abnormalities of the cardiac nerves. Both bradycardia and paroxysmal tachycardia may occur and the patient may need to have a demand pacemaker inserted. In a survey of patients treated at the Mayo Clinic the syndrome was fatal in about 20% of children whose trunk and respiratory musculature was affected. The authors of that paper suggested that cardiac monitoring should be instituted whenever assisted ventilation is needed (as would be wise in any acutely ill patient requiring mechanical ventilation).

Some deaths might be avoided if we had a test to predict potentially dangerous impairment of the cardiac autonomic system. A reduction of the normal beat to beat variation in heart rate may occur in the Guillain-Barré syndrome and is thought to indicate that the vagus nerve is affected. Persson and Solders have described a study of six patients with the syndrome in whom the electrocardiogram was recorded during two minutes of normal breathing and, when possible, during two minutes of deep breathing, after which consecutive R-R intervals were plotted against time and the variation in R-R interval in relation to the mean R-R interval calculated both for normal and for deep breathing. The recordings were repeated serially and compared with control recordings. All six patients showed similar results, with a gradual decrease in beat to beat variation, which became maximal two to four weeks after the onset of symptoms. This pathological regularity of the heart was most pronounced in those with the most severe muscle paralysis. Vagal paresis was suggested by finding an almost fixed sinus tachycardia of over 100 beats a minute. Three patients later showed periods of fluctuation in blood pressure or bradycardia, or both. Improvement in R-R variation paralleled clinical recovery. The most abnormal findings were in two patients who needed mechanical ventilation, while control studies in two other patients on ventilators but with no neurological disease showed no abnormality.

Although variations in R-R interval tend to decrease with increasing heart rate, and sinus arrhythmia is usually associated with sinus bradycardia, the Swedish workers did not believe that the tachycardia by itself was responsible for the decrease in R-R variation and the simple observation of tachycardia alone would not suffice as an indication of impaired vagal function because the differences in relative R-R interval variation were greater in the serial studies than changes in the recorded heart rate. Possibly, therefore, serial measurement of the R-R interval in patients with severe disease affecting the trunk and respiratory musculature may identify those patients at risk of developing fatal arrhythmias.

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Correction

Assessment of pituitary function

We regret that the 13th line of the 8th paragraph of Dr T D R Hockaday's leader on "Assessment of pituitary function" (10 December 1983, p 1738) was incorrect. It should have read "... growth hormone releasing hormone deficiency) on single shot."