Inco-ordination of oesophageal motor activity

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Physiology of the oesophagus

The oesophagus is essentially a motor organ and oesophageal disease often results in disordered motor function. Under resting conditions the gullet is normally sealed at its upper and lower extremities by sphincters which relax in a co-ordinated manner before the oncoming bolus. The oesophageal peristaltic wave travels at 2-3 cm/sec and is initiated either by swallowing or by reflux of gastric contents into the gullet (primary and secondary peristalsis).

The neural control of the oesophageal phase of the swallowing reflex is still not fully understood. The pharyngo-oesophageal sphincter is formed by the lowermost portion of the cricopharyngeus muscle which consists of striated muscle. In the cat under resting conditions a stream of impulses can be recorded from the vagal fibres supplying this sphincter, and relaxation of the sphincter is accompanied by a temporary interruption of this stream (Andrew, 1956). Vagotomy disrupts the swallowing reflex, the extent depending upon the level at which the nerve is sectioned, but sympathectomy appears to have little effect on oesophageal peristalsis. At intramural level, the Guy's group (Trounce *et al.*, 1957) have demonstrated cholinergic receptors (mediating muscle contraction) and two types of adrenergic receptors (mediating contraction and relaxation, respectively). Using isolated muscle from guinea-pig oesophagus Bailey (1965) has demonstrated α -excitatory and β -inhibitory adrenergic receptors in both the circular and longitudinal coats. β -blockade with pronethanol abolished the relaxation response to sympathomimetic amines and α -blockade with piperoxan abolished the contractile response.

The circular muscle coat of the lowermost portion of the human oesophagus differs from the longitudinal coat and from the circular coat above, in that the adrenergic receptors mediating relaxation predominate and post-ganglionic nerve fibre stimulation with nicotine causes relaxation rather than contraction (Ellis *et al.*, 1960b). These findings emphasize the difference in function between the circular coat in the lower oesophagus from which the oesophago-gastric sphincter is

formed and the remainder of the oesophageal musculature.

Achalasia

The term achalasia was coined by Hurst because failure of relaxation appeared a better description than the word cardiospasm which had been used by von Mickulicz. However, neither is entirely satisfactory since they fail to emphasize that this is a widespread disorder of oesophageal function rather than one limited to the oesophago-gastric function alone. The functional disturbance is characterized by disruption of the swallowing reflex in all except the uppermost portion of the gullet. Co-ordinated peristalsis is replaced by irregular contractions of the gullet and the oesophago-gastric sphincter fails to relax although its tone fluctuates and if the oesophagus is full it may contract tightly.

There is much to indicate that in achalasia the ganglion cells of the intramural plexus of the oesophagus have degenerated. After autonomic denervation of a viscus the response to the appropriate chemotransmitter is increased (Cannon's Law) and Kramer & Ingelfinger (1951) have demonstrated that the achalasic oesophagus contracts violently in response to acetyl β -methylcholine suggesting that cholinergic innervation has been lost. That the ganglion cells or post-ganglionic nerve fibres have degenerated in achalasia is confirmed by the work of Ellis and his colleagues (1960a) who demonstrated that isolated muscle strips from the oesophagus in achalasia did not respond to nicotine (which stimulates the post-ganglionic nerve fibre) although the response to acetylcholine was still present, indicating that the muscle fibres had retained their contractile power. The oesophageal motor disturbance in achalasia closely resembles that found in mega-oesophagus caused by Chagas' disease. This condition is endemic in South America and results from infection with *Trypanosoma cruzi*, destruction of which releases a neurotoxin causing degeneration of ganglion cells in the intramural plexus of the gut including the oesophagus.

The cause of the degeneration of ganglion cells

in achalasia remains obscure; similar changes are not found in any other part of the body and it seems likely that this may be due to a specific toxin as yet undiscovered.

Clinical features

The symptoms of achalasia may commence at almost any time of life, although they most commonly start in the fourth decade, and there is no sex difference.

Dysphagia, often intermittent at its onset and going back for several years, is the most common presenting feature. In the early stages there is usually little weight loss but as the condition progresses this becomes more prominent.

Anterior chest pain is common especially in the early stages and consists of a cramp-like sensation lasting for several minutes at a time.

Regurgitation of sour-tasting oesophageal contents into the mouth sometimes occurs early in the course of the illness. Occasionally aspiration of oesophageal contents into the respiratory tree causes recurrent bouts of pneumonia progressing to pulmonary fibrosis with the patient complaining of dysphagia. Lipoid pneumonia may ensue especially if liquid paraffin is used as an aperient or as a lubricant on bougies.

Arthropathy is a rare complication of advanced achalasia with gross oesophageal stasis, relief of which commonly improves the joint condition.

Carcinoma of the oesophagus may complicate mega-oesophagus in which oesophageal stasis and chronic oesophagitis are of longstanding. It seems probable that adequate treatment of the achalasia will, by controlling oesophagitis, help to reduce the incidence of this complication. But the severely damaged mucosa does not undergo complete healing after cardiomyotomy and I have known oesophageal carcinoma occur 2 years after this operation had successfully relieved dysphagia.

The clinical course of achalasia

Although dysphagia and pain often occur early in the disease these may subsequently remit for several years (compensated phase) before returning and progressing to cause weight loss and constitutional upset.

Radiological changes

It is important to remember that in the early phase of achalasia, even when dysphagia is troublesome, oesophageal dilatation may be absent. At this stage the diagnosis is commonly overlooked and if the inco-ordinated contractions are seen these are attributed to oesophageal spasm (Fig. 1). Absence of the gastric air-

bubble may point to this diagnosis but as the oesophagus dilates to become first cucumber-shaped and later sigmoid with a constantly contracted lower segment, there can no longer be any difficulty.

FIG. 1. Radiographs of the oesophagus in a woman of 26 with achalasia. That on the left was taken after dysphagia had been present for 6 months and shows no dilatation although in-co-ordination of peristalsis was detected manometrically. The radiograph on the right was taken 2 years later when dysphagia was more disabling and shows oesophageal dilatation with contraction of the lower segment, typical of achalasia.

Diagnosis

Carcinoma at the oesophago-gastric junction may cause diagnostic difficulty particularly if this arises on the gastric side of the cardia where it may grow underneath the intact oesophageal epithelium and thus be easily missed at endoscopy. Hence the diagnosis of achalasia should never be made lightly in an elderly person with a short history of dysphagia and oesophageal motor studies may be required to confirm the presence of widespread motor inco-ordination.

Treatment

Spontaneous cure of achalasia is extremely rare and because of the risk of aspiration pneu-

monia and even of carcinoma where the disease is of long standing, treatment should not be delayed.

Dilatation. The circular muscle fibres at the oesophago-gastric junction must be torn for dilation to have any lasting effect and for this reason bouginage has no place in the treatment of achalasia. Dilatation has been used more frequently in North America than in this country; the technique has been described by Nansen (1962) who advocates a radio-opaque Mosher bag and full radiological control throughout the procedure. Local rather than general anaesthesia is used since only if pain is experienced can the operator be certain that the procedure has been successful and Nansen believes that the relaxation of the cardia caused by general anaesthesia may preclude successful dilatation. The bag is inflated to a pressure of 15 lb/in² which is maintained for 15 sec which usually distends the cardia to a diameter of 3–4 cm. Two or three dilatations at the same sitting are usually applied. Results comparable with those of cardiomyotomy have been obtained at certain centres and the morbidity and mortality of the procedure is low.

Cardiomyotomy is still the treatment of choice provided the patient's general condition is suitable. Good results have been reported by Ellis (1962) who classified the outcome as good in 75%, better in 18% and worse in 7% and by Lawrence & Shoesmith (1959) who found 53% cured and 45% better. Not infrequently the hiatus has become stretched and a reverse type of hernia may develop with the lower portion of the distended oesophagus passing down into the abdomen. The advantage of operative treatment of achalasia is that the hiatus can be tightened and thus help to diminish the incidence of post-operative symptoms of gastro-oesophageal reflux.

Oesophageal spasm

Oesophageal spasm is a term commonly used to describe non-progressive inco-ordinated contractions which may be confined to a localised part of the gullet or may extend over most of its length. The names diffuse spasm, diffuse muscle hypertrophy, corkscrew oesophagus and tertiary contractions have been applied to this condition. Although confusion with achalasia in its early stages may occur, oesophageal spasm is a separate entity and ganglion cells do not disappear from the oesophageal wall nor is there increased sensitivity to acetyl β -methylcholine (Creamer, Donaghue & Code, 1958).

It has long been known that oesophageal spasm is common in patients with hiatus hernia. It seems

highly improbable that the hernia is simply a consequence of oesophageal shortening caused by muscle contraction and there is now much evidence to support the view that oesophageal spasm is commonly caused by acid reflux (Bennett, 1966).

(1) Oesophageal spasm can often be produced by perfusion of the normal oesophagus with N/10 hydrochloric acid. Oesophageal spasm and pain can be reproduced in patients by this means.

(2) Overnight intra-oesophageal pH recordings reveal evidence of reflux of acid in many patients with oesophageal spasm.

(3) Endoscopy and oesophageal mucosal biopsy usually show evidence of oesophagitis in these patients.

Clinical features

Oesophageal spasm may be quite symptomless as any radiologist will testify. In other patients oesophageal spasm is associated with cramping retrosternal pain or with heartburn. Dysphagia is much less prominent in the majority of patients with oesophageal spasm and if it predominates care must be taken to exclude early achalasia. The symptoms are usually episodic and may be precipitated by emotional stress.

Radiological diagnosis of oesophageal spasm

Irregular non-progressive oesophageal contractions point to the diagnosis of oesophageal spasm and the crenated outline of the gullet should not be mistaken for the filling defects caused by oesophageal varices. Unlike achalasia, oesophageal spasm is a variable phenomenon and is often absent at the time of radiological examination. Acidified contrast media may serve to provoke this disorder. If spasm is of long standing, pulsion diverticula may be present and the oesophageal muscle coat may hypertrophy and so produce a double oesophageal outline after swallowing barium, the distance between the outer shadow and the edge of the barium representing the thickness of the gullet wall (Johnstone, 1960).

Manometry is useful in the diagnosis of oesophageal spasm perhaps because of the irritant effect of a polythene tube introduced into the oesophagus. Code aptly describes the motor response to a swallow as being too soon, too much and too long compared with normal peristalsis. There is no significant difference in the pattern of motility between those with symptoms and those without (Roth & Fleshler, 1965).

The treatment of oesophageal spasm is usually that of the causative gastro-oesophageal reflux and hiatus hernia. Antispasmodics are of little value and by relaxing the oesophago-gastric

sphincter may actually aggravate reflux and pain. Extended oesophageal myotomy has been used with disappointing results and this operation has no rational basis. Vagotomy is equally ineffective in controlling symptoms.

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