Clinical, Manometric, and Pathological Studies in Diffuse Oesophageal Spasm

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Osgood (1889) is generally credited with the introduction of the term “diffuse spasm” of the oesophagus. However, though Moesch and Camp (1934) recorded a classical description of the clinical and radiological features of the disease it attracted little interest until the introduction of oesophageal motility studies in the 1950s. Since then a characteristic motility pattern has been described (Creamer et al., 1958; Roth and Fleshler, 1964), and an operation which relieves the symptoms of the severely affected patients has been devised (Ellis et al., 1960).

However, there remains some confusion in the terminology because “diffuse spasm” has been applied not only to the clinical entity as reported here but also to various radiological and physiological findings which, though characteristic of diffuse spasm, may also be seen under certain circumstances in patients with carcinoma (A. P. Skyring and R. M. H. Kater, personal communication), during acid infusions of esophagitis (Siegel and Hendrix, 1963), and in asymptomatic nonagenarians (Soergel et al., 1964). Some radiologists have used the term interchangeably with rippling, curling, corkscrew, and elevator oesophagus regardless of the pathological process underlying the motility disturbance.

There is no doubt that motility and radiological changes similar to those found in diffuse spasm may be seen in patients with organic disease of the oesophagus, but these may be excluded by oesophagoscopy, cytology, acid perfusion, and mechollyl tests. There remains a group of patients with a clinical history of pain and dysphagia in whom peristalsis is replaced by synchronous high pressure and repetitive activity in the absence of organic disease. This group represents the clinical entity of diffuse spasm. We have had the opportunity to study such a group of patients, and we report here the clinical, radiological, motility, and pathological changes, and the results of surgery, in severely affected patients.

Materials and Methods

A group of 21 patients was selected from the patients referred for investigation of oesophageal symptoms by motility studies. All had a history of intermittent attacks of pain or dysphagia in the absence of a demonstrable organic lesion, and in all cases the normal peristaltic activity after swallowing was replaced by synchronous contractions which were often repetitive and of high pressure.

All patients were interviewed when manometry was performed and were followed for periods of up to four years by means of interviews and a written questionnaire.

Motility studies were performed, using the direct-writing Sanborn Polygraph, which was connected to oesophageal catheters via pressure transducers. Recordings were taken simultaneously at two or three points 5 cm. apart at 1 cm. intervals throughout the length of the oesophagus.

Results

Clinical Features

Twenty-one patients (11 males and 10 females) met the clinical and manometric criteria for diagnosis. The age and sex distributions are shown in Table I.

<table>
<thead>
<tr>
<th>Age Group</th>
<th>No. of Males</th>
<th>No. of Females</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 40 years</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>40-60</td>
<td>7</td>
<td>3</td>
</tr>
<tr>
<td>&gt; 60</td>
<td>6</td>
<td>4</td>
</tr>
</tbody>
</table>

All patients gave a history of intermittent attacks of dysphagia which were often precipitated by emotional distress. Pain was associated with dysphagia in about 60% of cases, and was usually described as a gripping sensation behind the lower end of the sternum which on occasions radiated to the left chest and left arm. The dysphagia was described as a sensation of food sticking, usually in the lower oesophagus, but occasionally in the mid or upper third. One patient presented with a history of episodes of choking for two weeks. Dysphagia was usually relieved by repeated swallowing, by regurgitation of the offending food, or by eructation. On some occasions dry retching relieved the dysphagia, but in severely affected patients spontaneous or induced vomiting was the only method of producing relief. In the majority of cases the dysphagia and associated pain were relieved within a few seconds or minutes. However, two patients were admitted to hospital with complete bolus obstruction which had been present for 8 to 12 hours. One of these, a young woman, required oesophagoscopy and removal of a piece of meat before her symptoms were relieved. Barium studies in this patient showed no organic obstruction. A motility disturbance was confirmed by manometry.

The pain associated with dysphagia was usually of moderate severity and, as stated, was relieved when the offending food had been removed. In some patients who experienced pain and dysphagia the pain would persist for periods up to one hour after the obstructive symptoms had been relieved. In addition, pain, unrelated to dysphagia, occurred during episodes of emotional distress and lasted for periods ranging from a few minutes to several hours. Severe dysphagia would occur almost invariably if patients were to eat a meal during such attacks.

* Brit. med. J., 1967, 2, 527-530
Loss of weight occurred in patients who had frequent severe attacks of pain and dysphagia, associated with vomiting (Table II). Two patients had lost more than 2 stone (12.7 kg.) in weight when they were submitted to surgery.

In all cases the symptoms, once established, recur with constant severity for periods of up to four years. The duration of symptoms at the time of presentation was less than three years in all but one patient (Table III). In six of the seven patients who were severely incapacitated and required surgery the history was less than two years. The majority of patients volunteered the information that emotional distress often precipitated an attack. Some were able to control symptoms by avoiding large, rapidly eaten meals during periods of tension. One patient had been asymptomatic for two years by avoiding food which in his experience caused severe pain and dysphagia.

Radiology

Barium swallows were performed in all patients. Sixteen were available for review at the time of this study. Though a motility disorder of some type was present in 12 patients only four showed "classical diffuse spasm" in which there was spasm with succussion and diverticulum formation and evidence of thickened oesophageal muscle (Fig. 1). In a further eight patients an oesophageal dysrhythmia was present which in these studies had been reported as showing "rippling, curling, or tertiary activity." The severity of the radiological change did not show a close correlation with the clinical, manometric, or pathological findings (Fig. 2).

Endoscopy

Oesophagoscopy was performed in 13 patients but in no instance contributed to the diagnosis. However, by this means it was possible to exclude the presence of oesophagitis or organic stricture or carcinoma.

Manometry

Analysis of the motility patterns has enabled better definition of the abnormalities which may be expected in patients with severe diffuse spasm. Disturbances of motility occurred mainly in the mid and lower oesophagus, but in some patients abnormal patterns were present in the upper third of the oesophagus (Table IV).

**TABLE II.—Clinical Features**

<table>
<thead>
<tr>
<th>Feature</th>
<th>Males</th>
<th>Females</th>
<th>Males</th>
<th>Females</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dysphagia</td>
<td>100%</td>
<td>90%</td>
<td>41%</td>
<td>40%</td>
</tr>
<tr>
<td>Pain</td>
<td>64%</td>
<td>60%</td>
<td>48%</td>
<td>50%</td>
</tr>
<tr>
<td>Weight loss</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vomiting</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

In all cases the symptoms, once established, recur with constant severity for periods of up to four years. The duration of symptoms at the time of presentation was less than three years in all but one patient (Table III). In six of the seven patients who were severely incapacitated and required surgery the history was less than two years. The majority of patients volunteered the information that emotional distress often precipitated an attack. Some were able to control symptoms by avoiding large, rapidly eaten meals during periods of tension. One patient had been asymptomatic for two years by avoiding food which in his experience caused severe pain and dysphagia.

**TABLE III.—Duration of Symptoms in 17 Patients in Years**

<table>
<thead>
<tr>
<th>No. of patients</th>
<th>&lt;1</th>
<th>1-2</th>
<th>&lt;3</th>
<th>30 (severe 1 month)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males</td>
<td>8</td>
<td>4</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Females</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>1</td>
</tr>
</tbody>
</table>

In six of the seven subjects to surgery duration was <2 years.

**TABLE IV.—Distribution of Motility Disturbances in 21 Patients**

<table>
<thead>
<tr>
<th>Site in oesophagus</th>
<th>Synchronous</th>
<th>Repetitive</th>
<th>Prolonged High Pressure</th>
<th>Pressure &gt; 40 mm. Hg</th>
<th>Spon-Taneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males</td>
<td>11</td>
<td>8</td>
<td>10</td>
<td>11</td>
<td>8</td>
</tr>
<tr>
<td>Females</td>
<td>10</td>
<td>8</td>
<td>12</td>
<td>10</td>
<td>7</td>
</tr>
<tr>
<td>Total</td>
<td>21</td>
<td>16</td>
<td>20</td>
<td>21</td>
<td>11</td>
</tr>
</tbody>
</table>

Synchronous activity (Fig. 3) was present after every swallow recorded in the lower oesophagus in all but one patient, and was present in the upper third after more than 80% of swallows in 16 patients.

Repetitive activity (Fig. 4) with two to five contractions after swallowing and lasting for periods up to 30 seconds was present in 20 patients and occurred in the upper third in 10.

Contractions lasting longer than six seconds were present in the upper oesophagus in 11 patients and in the lower two-thirds in all patients. The duration of contractions ranged from 4 to 20 seconds (Fig. 5).

In the lower two-thirds of the oesophagus pressures after swallowing and during repetitive and spontaneous activity ranged from 20 to 120 mm. Hg and were often in excess of 60 mm.
mm. Hg. In the upper third pressures greater than 40 mm. Hg were shown in 10 cases and in three cases pressures greater than 110 mm. Hg were recorded.

There was an overall rise of the mean resting pressure of the oesophagus in this group of patients, with a range of 5 to 25 mm. Hg. A resting pressure greater than 10 mm. Hg was observed in the lower oesophagus in nine patients and in six of these the high pressures extended into the upper third (Fig. 6). In three patients the lower oesophageal sphincter had a resting pressure ranging from 15 to 20 mm. Hg but relaxed normally on swallowing. In all other patients the lower sphincter was normal.

Spontaneous activity manifested by high pressure and synchronous and repetitive activity in the absence of swallowing (Fig. 6) was present in eight severely affected patients and was seen in the upper oesophagus in four of these.

![Fig. 4](image_url) - Motility recording from the upper oesophagus in a 70-year-old man with symptoms of intermittent dysphagia and pain for the previous two years. There was no change in the symptoms after three-years follow-up.

![Fig. 5](image_url) - Motility study of a 65-year-old man complaining of intermittent dysphagia, showing prolonged high-pressure waves in the mid-oesophagus after swallowing.

![Fig. 6](image_url) - Motility study of a severely affected young man showing spontaneous, high-pressure, synchronous, and repetitive activity and a high oesophageal resting pressure.

**Treatment**

In seven patients symptoms were severe enough to warrant oesophagectomy. The duration of symptoms was less than two years in all but one case. This patient had experienced an isolated episode of dysphagia 30 years earlier but had been asymptomatic until three months before operation. In this surgical group attacks of pain and dysphagia developed fairly rapidly over about three months and then increased in severity and frequency until the time of presentation, when all patients were severely incapacitated, unable to work, and afraid to eat, and were experiencing severe pain which was only occasionally relieved by vomiting. Loss of weight was severe in all, and two had lost more than 2 stone (12.7 kg.).

Motility studies in these seven patients showed severe abnormalities. Synchronous activity involved the entire length of the oesophagus in all but one patient. Spontaneous activity was found in all patients, and occurred in the upper oesophagus in two. Repetitive and prolonged activity was frequent and severe throughout the length of the oesophagus. There was a rise in the mean resting pressure up to 25 mm. Hg. Pressures after swallowing were very high, and in two cases exceeded 110 mm. Hg.

Of the seven patients submitted to surgery three were men and four were women; four patients were under 40 years, their ages being 29, 34, 35, and 36, and three were over 60. All patients were relieved of their pain and dysphagia, and became essentially asymptomatic. They gained weight and led normal lives up to three years after operation. There were no post-operative complications.

At operation there was thickening of the oesophageal muscle extending from the cardia up to the arch of the aorta and to a less extent beyond this. In some cases the muscle was 2 cm. thick. Histological examination revealed the presence of chronic inflammatory change with an eosinophilic infiltration; ganglion cells were present in all four of the patients from whom biopsy specimens were taken.

A comparison of the premyotomy and postmyotomy studies in the patients submitted to surgery showed that, though synchronous activity persisted, the pressures recorded after swallowing were lower, and repetitive and spontaneous activity no longer occurred after surgery.

Thirteen out of 14 patients who had been managed conservatively were followed up for periods of up to four years. Antacids were of no value and symptoms had remained unchanged after the initial motility studies. The majority of patients were able to lead normal lives, though emotional distress continued to precipitate symptoms.

Motility studies were repeated in some of the patients managed conservatively, and it was found that these studies were almost identical with those recorded at the time of the initial presentation.

**Discussion**

The symptoms of pain and dysphagia of which the selected group reported here complain may reasonably be ascribed to the physiological and pathological abnormalities described. The age distribution in this group supports the argument that this is a disease entity rather than an extension of the normal physiological change seen with advancing age. Further support is proved by the operative finding of muscle hypertrophy and chronic inflammation in all patients, including the young. The presence of normal ganglion cells distinguishes the disease from achalasia.

The presence of synchronous high pressure and repetitive activity in the upper third of the oesophagus in this group suggests that the disease may be more extensive than previously recognized. Other series emphasize the presence of abnormal contractions in the lower two-thirds (Creamer et al., 1958;
Normal Confinement: A Domiciliary and Hospital Study


In view of the current interest in various patterns of obstetric care a special survey has been made of mothers whose antenatal care and delivery were free from complications. This paper describes the survey results and is divided into three parts, the aspects analysed being certain clinical details, results of early discharge from hospital, and maternal preference for place of confinement.

The material consists of a series of consecutive normal confinements during the winter of 1964–5 at the Barratt Maternity Home, Northampton, and domiciliary confinements in the area of Northamptonshire normally looking to that home for the provision of obstetric services. There were 233 confinements at home and 270 in hospital, recorded by hospital or domiciliary midwives, and the mothers were subsequently interviewed 28 days after confinement, when a questionary dealing with parturition and with antenatal and postnatal experiences was completed. It is worth emphasizing that these interviews were undertaken by health visitors who, in all but a few cases, did not act as midwives, thus reducing the possibility of biased answers which might have been given to the midwives associated with the confinements.

Antenatal and postnatal care was supplied by general practitioners and domiciliary midwives for patients confined at home, and they also shared with hospital personnel in the provision of such care for those with normal pregnancies who had their babies in hospital.

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