

Transition from symptomatic diffuse spasm to cardiospasm¹

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EDITORIAL COMMENT This paper provides further evidence that these two conditions may be related disorders. The transition from diffuse spasm to cardiospasm is recorded in one patient.

In previous communications (Kramer, Fleshler, and McNally, 1963; Kramer, Fleshler, McNally, and Harris, to be published) we suggested that symptomatic diffuse spasm and cardiospasm are related disorders. This conclusion was based upon the finding that the oesophagus of patients with symptomatic diffuse spasm responds in a hypersensitive fashion to Mecholyl, a phenomenon which has been accepted as being characteristic of cardiospasm (Kramer and Ingelfinger, 1951; Hightower, Olsen, and Moersch, 1954).

The purpose of this report is to present further evidence supporting this suggestion by describing the changes in symptoms and oesophageal motility that occurred in a patient during eight years of observation. When he was first seen in 1956 his oesophageal radiographs, balloon-kymographic, and manometric recordings were typical of diffuse spasm. In 1963 and 1964 these examinations revealed the characteristic radiographic and motility pattern of cardiospasm.

CASE HISTORY

F.C., a man aged 70, noted the onset of difficulty in swallowing at age 61 in 1955. He was then admitted to the Boston Veterans Administration Hospital. In 1956 his symptoms were of food sticking at the level of the lower end of the sternum. All foods would produce this sensation but solid foods were particular offenders. Pain on swallowing as well as retrosternal discomfort without swallowing were often felt. He also described a feeling as if his swallowing tube would open and close. He frequently was awakened at 2 to 3 a.m. with high epigastric distress which he described as a fullness or a 'gassy' sensation. At no time was there any regurgitation, nausea, or vomiting. His weight was stable.

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Over the next six or so years his symptom did not change particularly, except that he appeared to have heartburn and a burning epigastric distress. He was oesophagoscoped in 1962 at the Boston Veterans Administration Hospital and a mild oesophagitis was visualized; a regimen of elevation of the head of the bed and frequent antacid ingestion was instituted.

In 1963 dysphagia became worse so that soft foods seemed to become lodged in the oesophagus and he would have to drink at least 8 ounces of water or take 'heavy solid' substances to force the food through into the stomach. If the food did not become dislodged he would become nauseated and would vomit or belch. Belching would often bring up undigested food. Several times during the period he was awakened at night with food in his mouth or nose. He lost 8 lb. in weight.

Because of symptoms and x-ray findings, a forceful dilatation with a Browne-McHardy dilator was performed. Since that time (September, 1963), his swallowing has improved greatly, food now passing into the stomach with little difficulty. Belching, nausea, vomiting, and regurgitation have disappeared completely. Epigastric burning is still a frequent complaint.

MOTILITY STUDIES

Oesophageal motility was investigated by means of radiography following a barium swallow, balloon-kymography, and manometry. Standard methods were utilized as previously described (Kramer and Ingelfinger, 1949; Fleshler, Hendrix, Kramer, and Ingelfinger, 1958). During this long period of observation eight balloon-kymographic and seven manometric studies were performed; only those which are particularly pertinent will be discussed.

RADIOGRAPHY Examination of the oesophagus in 1956 with the patient in the upright position revealed that this organ was somewhat dilated and emptying was slightly delayed. The sphincteric segment opened readily. When in the right anterior oblique prone

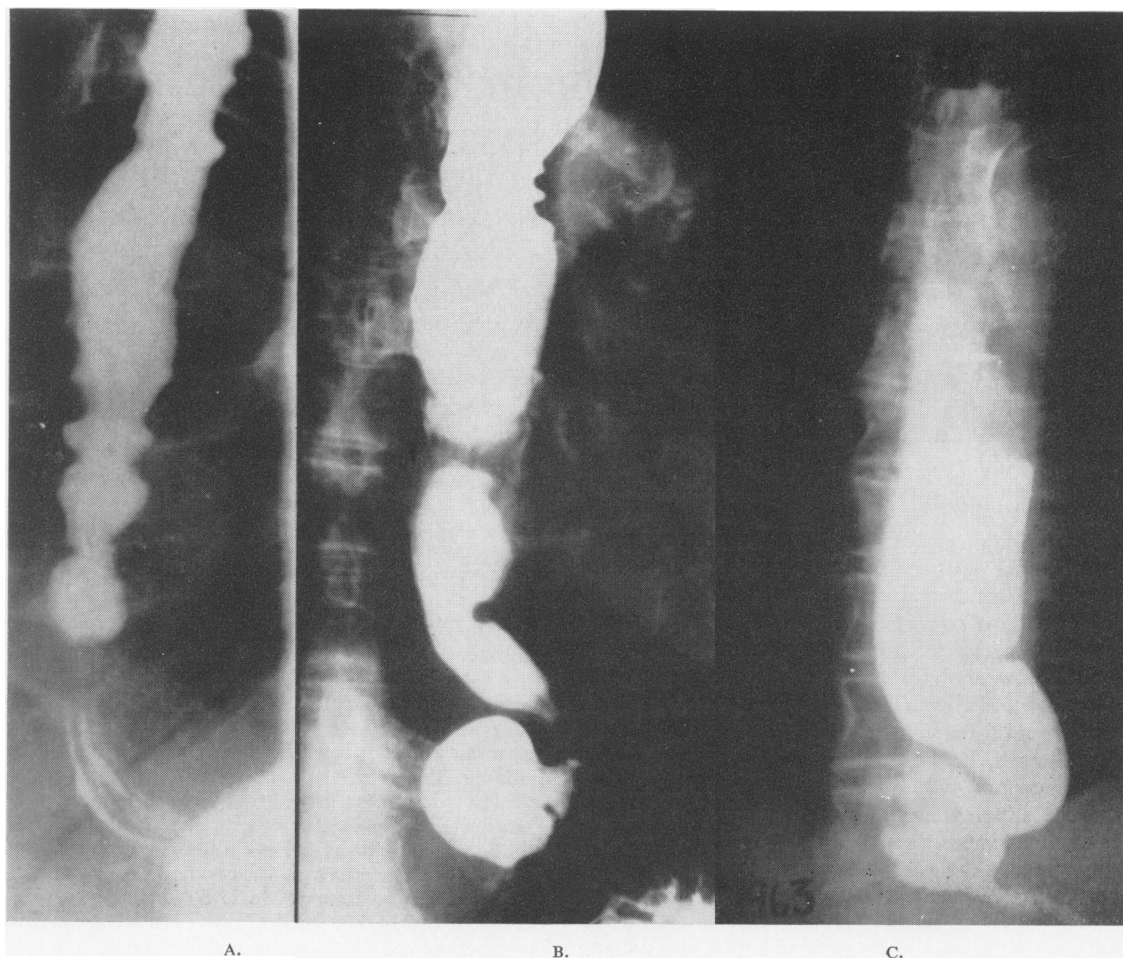


FIG. 1A. Radiographs taken in 1956 show segmental contractions. The sphincteric area is open widely. The diagnosis of symptomatic diffuse spasm was made at this time.

FIG. 1B. Segmental contractions were markedly decreased in an oesophagram taken in 1962. The upper one-half of the oesophagus was somewhat dilated. A large epiphrenic diverticulum can be seen in the lower oesophagus.

FIG. 1C. In 1963 the entire oesophagus was more dilated and contractions were seen only occasionally. In the terminal portion of the oesophagus a narrowed tapered area is seen. The appearance is that of cardiospasm.

position, primary peristalsis was seen to proceed as far as the arch of the aorta and then the lower two-thirds of the oesophagus contracted as a whole, showing localized segmental contraction (Fig. 1A).

A change in the appearance of the gullet was observed in 1962; the upper half was now dilated and there was an epiphrenic diverticulum (Fig. 1B). A more radical alteration in appearance was noted in 1963 (Fig. 1C); the entire oesophagus was now markedly dilated and there was a fluid level. Above the epiphrenic diverticulum, the gullet had a sigmoid appearance, while below it ended in a narrowed tapered segment. A small quantity of barium had

been given and after an hour only a minimal amount was seen to have entered the stomach.

Re-examination in the upright position in 1964, *i.e.*, after the dilatation, revealed that the distal narrowed area now was opened widely and emptying occurred readily. In the prone position, the oesophagus was still markedly dilated, occasional localized segmental contractions were observed; no emptying occurred during the one-half hour of observation.

BALLOON - KYMOGRAPHY A balloon - kymographic tracing in 1956 (Fig. 2A) showed tone of 13 ml., and

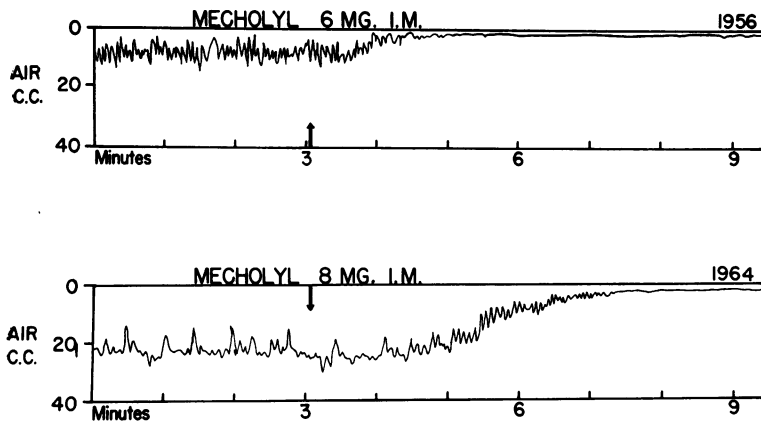


FIG. 2. Balloon-kymographic tracings obtained in 1956(A) and 1964(B). Both demonstrate lumen-obliterating contractions in response to Mecholyl. In the lower tracing the response to 8 mg. is shown because the graph obtained with 6 mg. was technically inferior and difficult to reproduce. The 6 mg. response was similar to that with the 8 mg. dose.

a regular wave pattern of 15 contractions per minute (normal average 8 per minute) (Kramer and Ingelfinger, 1949). Three and 6 mg. of Mecholyl intramuscularly produced a lumen-obliterating contraction expelling all air from the balloon, a response which has been defined as demonstrating hypersensitivity to Mecholyl (Kramer and Ingelfinger, 1951); at the same time the patient stated he experienced retrosternal pain identical to that he felt on swallowing.

A repeat study in 1964 (Fig. 2B) showed that the tone now was lower, being 25 ml., the wave pattern

was irregular so that the number of contractions per minute could not be counted. The response to Mecholyl was unchanged, *i.e.*, the same lumen-obliterating contraction occurred. The wave pattern, tone, and Mecholyl effect were identical to those seen in cardiospasm.

MANOMETRY Repeated manometric recordings in 1956, 1957, 1959, and 1960 revealed the characteristic motility pattern of diffuse spasm, *i.e.*, repetitive, simultaneous, non-peristaltic contractions involving a long segment of the oesophagus although

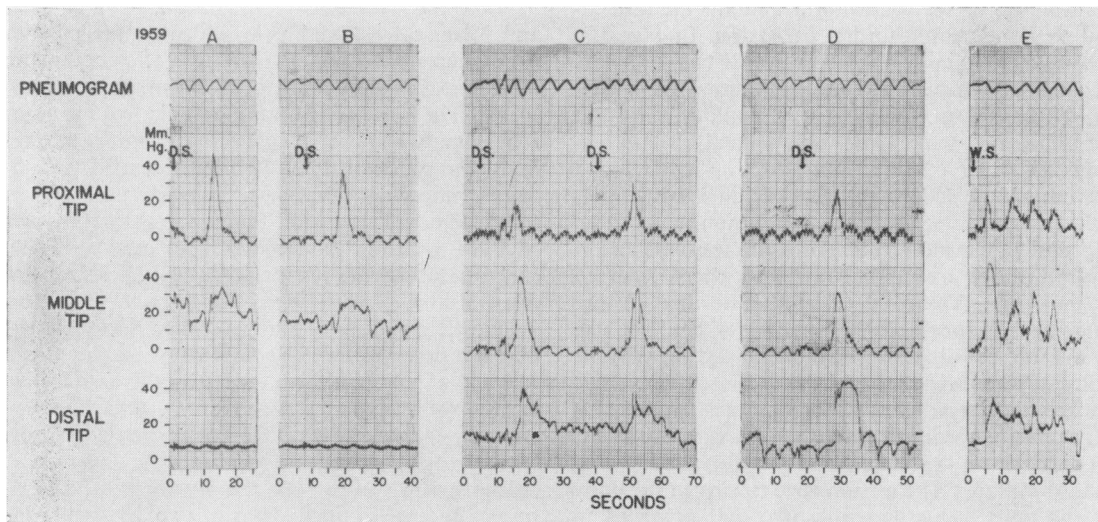


FIG. 3. Manometric tracings obtained in 1959 during one study; the catheter tips are 6 cm. apart. In panels A and B the middle tip is in the sphincteric area and the distal tip is in the stomach. A normal sphincteric opening pattern is seen following a dry swallow. In C, D, and E all the catheter tips are in the oesophagus. C shows that the contractions are sequential thereby demonstrating normal peristalsis. In D the waves obtained via the proximal and middle tip occur simultaneously. In panel E, a wet swallow produced simultaneous repetitive contractions typical of diffuse spasm.

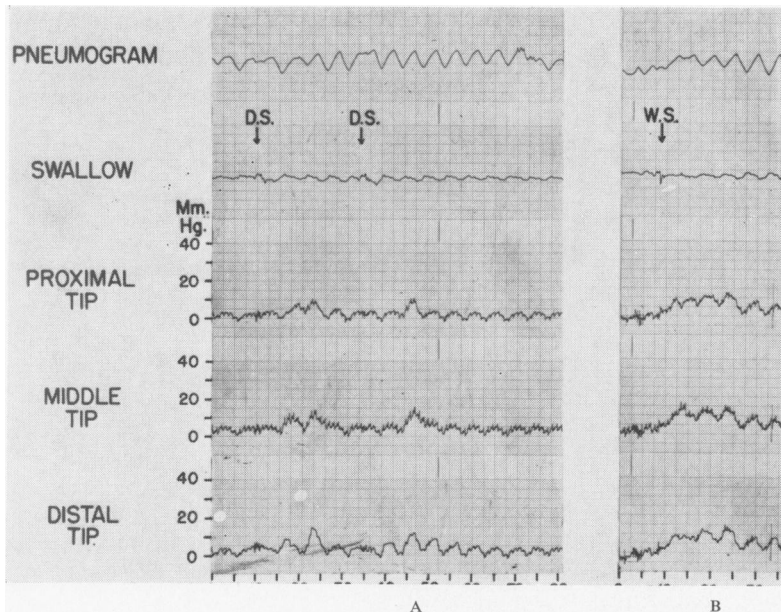


FIG. 4. Manometric tracings obtained in 1964. The distal tip is 48 cm. from the external nares just after it passed through the diaphragm on a centimetre by centimetre pull-through of the catheter ensemble. No high pressure zone could be detected. The distal tip should, however, be in the sphincteric area. Only feeble simultaneous contractions were seen in the body of the oesophagus following a dry in A and a wet swallow in B.

a normal peristaltic wave was rarely seen. All the tracings shown in Fig. 3 were obtained from one study in 1959. In Figs. 3A and 3B the recordings via the middle tip demonstrate a high resting pressure indicating a closed sphincter; on swallowing an 'opening' pattern is seen. All catheter tips are in the distal one-half of the oesophagus in Fig. 3C; the contraction waves following a swallow are sequential thus indicating normal peristalsis. In Fig. 3D, the waves obtained via the proximal two catheters occur simultaneously after a dry swallow, the pattern of diffuse spasm. Following a swallow of water, simultaneous, repetitive, non-peristaltic contractions are observed (Fig. 3E).

Manometric recordings obtained in 1964 (Fig. 4) now demonstrated a marked alteration in motility. No zone of elevated resting pressure was detected in the sphincteric area and no opening pattern occurred at any time. The absence of the elevated resting pressure and opening pattern, however, could be due to the forceful dilatation in the previous year. In the body of the oesophagus only feeble simultaneous contractions were seen on dry or wet swallows; a slight rise in basal pressure occurred with the latter. At no time was normal peristalsis observed following many swallows. The manometric tracing in the body of the oesophagus was now like that observed in advanced cardiospasm.

DISCUSSION

Although Barrett (1964) stated that he had observed cases of diffuse spasm which later looked like cardio-

spasm, we have not found documentation of this transformation by motility studies. Before this change can be accepted as being demonstrated, two questions must be answered: 1 Did our patient originally have diffuse spasm or did he represent an atypical case of cardiospasm from the beginning? 2 Does he presently have cardiospasm?

While his symptoms of dysphagia, odynophagia, and lower retrosternal pain which awakened him from sleep were more characteristic of symptomatic diffuse spasm they could occur in atypical cardiospasm. The oesophageal radiographic findings after a barium swallow of localized, segmental contractions, the minimal dilatation, and only slightly impaired emptying were more suggestive of diffuse spasm than cardiospasm. Balloon-kymography was of little value in differential diagnosis since the control tracings in normal elderly individuals are often bizarre. The response to Mecholyl was of no diagnostic significance since we have demonstrated that the oesophagus in symptomatic diffuse spasm as well as in cardiospasm reacts in a hypersensitive fashion to Mecholyl (Kramer *et al.*, 1963; Kramer and Ingelfinger, 1951). The manometric records, however, were more consistent with the diagnosis of diffuse spasm and exclude the possibility of cardiospasm, because of the findings of a sphincteric opening and occasional normal peristalsis.

Does this patient now have cardiospasm? The substernal pressure on eating, which could be relieved by washing his food through with water, and the regurgitation are symptoms of cardiospasm. The findings of a dilated oesophagus with a fluid

level, the marked impairment of emptying, the absence of peristalsis on swallowing, the presence of a narrowed tapered area are the typical x-ray changes of cardiospasm. Excellent relief of symptoms by a forceful dilatation also is typical of cardiospasm. Balloon-kymography and manometry support the diagnosis: the former because of the oesophageal hypersensitivity to Mecholyl, the latter because of the feeble non-peristaltic contractions on swallowing. Unfortunately sphincteric opening could not be evaluated because of the previous forceful dilatation in 1963. Thus symptoms, the radiographic, balloon-kymographic, and manometric evidence strongly support the present diagnosis of cardiospasm.

SUMMARY

A patient is presented whose symptoms, oesophageal radiographs, balloon-kymographic, and manometric studies in 1956 were diagnostic of diffuse spasm. In 1963-64 the clinical features, oesophageal radiographs, balloon-kymographic, and manometric

records were quite characteristic of cardiospasm. This progressive change from a picture of diffuse spasm to one of cardiospasm provides further evidence that these two disorders are related.

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