CARDIOSPASM IN TWO BLOOD RELATIONS

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We have found no reference in the literature to a family incidence of cardiospasm, and the discovery of two cases of this condition in blood relatives is therefore of interest.

Case 1

A man aged 40 was admitted to hospital several days after the onset of an acute respiratory infection. Fever, cough, sputum, and right-sided pleuritic pain were associated with clinical evidence of consolidation at the right lung base, and he was thought to have a simple right lower lobar pneumonia. X-ray examination of his chest confirmed the presence of consolidation as well as that of some fluid, but it also demonstrated gross dilatation of the oesophagus, the appearance of which was typical of cardiospasm.

Further questioning of the patient then revealed that for over twenty years he had had difficulty in swallowing, and had often felt his food "sticking in his throat." His symptoms began in early adolescence, and he soon found that he could usually overcome the obstruction in his gullet by drinking large quantities of water both during and after meals. His dysphagia was always worse when he was worried or nervous or in a hurry, and frequently he was forced to regurgitate his food. The consequent chronic malnutrition had led to considerable emaciation. His dysphagia was first investigated when he was serving in the Army during the second world war. There had been a strong rumour that his unit was to be sent to the Middle East, and his comrades, knowing that he had to drink large quantities of water with all his meals, suggested to him that he would fare badly in the desert. Realizing the truth of this, he consulted his medical officer, and subsequent investigations demonstrated cardiospasm. No treatment was offered, but he was not posted to the Middle East. His affliction was so much a part of his daily routine that it did not occur to him to mention it to us when he was admitted to hospital on the present occasion, but it was certainly relevant to his presenting complaint, since his chest infection took the form of an aspiration pneumonia.

A barium swallow (Fig. 1) confirmed the diagnosis of cardiospasm already made from the straight x-ray film of the chest and from the history. An attempt was made to empty the oesophagus before the barium examination, and among the food secretions which were withdrawn were found the remains of vegetables which the patient had eaten some thirty-six hours before—an indication of the severity of the "spasm." Inhalation of octyl nitrite relieved the "spasm" momentarily during the initial x-ray screening, but, despite further inhalation, barium was still present in the oesophagus many hours later. There was no sign of oesophageal peristalsis during screening, and a study of the oesophageal motility by the balloon-kymograph technique described by Kramer and Ingelfinger (1949) showed extreme depression of motility and almost complete absence of the so-called secondary peristaltic waves in the middle and lower thirds of the oesophagus.

In view of the particularly severe symptoms a modified Heller operation was performed after the chest infection

had been overcome. The dysphagia was completely relieved following the operation, and when last seen seven months later the patient was still symptom-free and had put on 2 st. (12.7 kg.) in weight. The symptomatic improvement was paralleled by the improvement in the radiological appearances. The chest film showed no evidence of an oesophageal shadow, and a barium examination revealed a considerable diminution in the size of the oesophagus, with little or no obstruction at the cardia. Oesophageal motility, however, was still minimal, as demonstrated both on the x-ray screen and on the tracing obtained by the balloon-kymograph technique.

Case 2

Mrs. X, the maternal aunt of the patient in Case 1, visited her nephew while he was in hospital, and for the first time the two discovered that they suffered from the same complaint. Her symptoms were not nearly so severe as those of her nephew, but, like his, they began when she was a young girl. During her early adult life her dysphagia was a source of considerable embarrassment to her, but in later

FIG. 1.—Case 1.

FIG. 2.—Case 2.

years she became more placid and swallowing was easier. A plain film of the chest showed evidence of cardiospasm, and this was confirmed by a barium swallow (Fig. 2). The "spasm" was not so marked as in Case 1 and there was quite obvious vigorous peristalsis in the lower oesophagus, confirmed by balloon-kymograph studies. Octyl nitrite had no effect on the "spasm."

Comment

Mrs. X was one of a family of three brothers and three sisters, and Case 1 was the son of her elder sister. So far as could be discovered, no other member of the family suffered from this condition, but unfortunately the inquiry could not be very exhaustive, as three of Mrs. X's siblings had emigrated to America and could not be traced.

One of the common complications of cardiospasm is aspiration pneumonia, and repeated attacks may lead to bronchiectasis. It is not unusual, therefore, for the condition to be diagnosed first from a radiograph of the chest.

Individually the two cases described are fairly typical, but the familial incidence is most unusual. It is possible that this association is merely accidental, although the odds against such a coincidence must be enormous.

We should like to express our thanks to Professor D. M. Dunlop and to Dr. W. S. Shearer for permission to publish these two cases.

REFERENCE

Kramer, P., and Ingelfinger, F. J. (1949). *Amer. J. Med.*, 7, 168.