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Diaphragmatic flutter

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Summary

A case is reported of diaphragmatic flutter with an unusually long duration of symptoms. The patient described demonstrates many of the classical features of this disorder.

Introduction

Diaphragmatic flutter is a rarely diagnosed disorder (Rigatto and DeMedeiros, 1962) and, because of the frequently bizarre symptomatology, diagnosis may be delayed, and referral for psychiatric assessment is common. The true cause of the symptoms is often only apparent when an attack is witnessed, or during X-ray screening, and it seems likely that some cases go undetected.

Case report

A Saudi Arabian female, first presented at the age of 15 years in 1957 with attacks of pain in the upper abdomen and lower chest, radiating round the lower ribs on the left side and down the left arm. The onset of pain was briefly preceded by marked epigastric pulsations and loud, rhythmical, splashing sounds. These features lasted for 2–15 min, whilst the pain persisted for up to a further 30 min. The frequency of attacks was very variable, sometimes occurring ten to fifteen times a day. Periods of freedom seldom exceeded 2 weeks. The attacks did not occur during sleep. Before correct diagnosis, she

underwent appendectomy, intercostal injections of local anaesthetics, numerous drug therapies and psychiatric referral. A laparotomy and pyloroplasty were performed in 1967 and conversion to a gastro-jejunostomy a week later for persistent vomiting. In 1968 diaphragmatic flutter involving the left hemidiaphragm was diagnosed by X-ray screening during a visit to London. Temporary relief from attacks was obtained by infiltration of the left phrenic nerve in the neck with local anaesthetic, and surgical crushing of the nerve was then carried out. This produced relief from symptoms for 3 months. This procedure was repeated, producing a further 1 month's freedom from attacks. Her symptoms then recurred, together with intermittent vomiting and continued up to her return to this country and referral to Professor L. A. Turnberg in July 1975.

On admission she was well nourished, and physical examination and biochemical investigations were normal. Many attacks of diaphragmatic flutter were witnessed during her stay in hospital. Each attack began with marked epigastric pulsations, vigorous enough to cause the whole bed to shake, and a bizarre rhythmical splashing sound which was audible from a distance of several yards, and loud enough to awaken nearby patients. She complained of severe pain and was obviously dyspnoeic, and a venous blood carbon dioxide content of 15 mmol/l was found on one occasion. A regular tachycardia of 120/min was noted during attacks and the blood pressure was unchanged. The epigastric pulsations

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occurred at a rate of approximately 60/min, and phonocardiography revealed that they were irregular and bore no constant relation to the cardiac cycle (Fig 1a, b). Auscultation of the heart during an attack revealed only the transmitted splashing sound which obscured all other findings. There was no evidence to suggest that the patient could initiate the attacks at will, nor were there any physical manoeuvres which would consistently provoke or terminate them.

ECG between attacks was normal, but during attacks T wave inversion occurred in leads III and V1-V4 inclusive (Fig. 2a, b). There was a sinus tachycardia of 120/min and the QRS axis was unchanged. Chest X-ray was normal and barium meal examination showed a gastro-jejunostomy which functioned normally.

X-ray screening between attacks showed normal diaphragmatic respiratory movement, but during attacks the left hemidiaphragm was seen to contract

very vigorously at approximately 60/min, with an amplitude of several inches. Barium swallowed during an attack revealed that the stomach was swung forcibly downwards and towards the midline with each contraction of the diaphragm, producing splashing sounds. The normal respiratory movements of the left hemidiaphragm were abolished by these contractions, although the excursions of the right hemidiaphragm were normal in amplitude but increased in frequency.

Treatment

Following the apparently successful use of diphenylhydantoin in this disorder (Phillips and Eldridge, 1973), this agent was given in a dose of 100 mg twice daily with chlorpromazine for persistent vomiting which was considered to be psychogenic in origin. Within 48 hr the patient complained of pain over the dorsum of both hands and examination revealed tender, non-erythematous swelling on

FIG. 1. (a) Phonocardiogram between attacks; (b) phonocardiogram during an attack. Note: (i) sinus tachycardia; (ii) transmitted abdominal splashing sounds which are irregular and not related to cardiac cycle.

FIG. 2. (a) ECG lead V4, between attacks; (b) ECG lead V4, during an attack.

the hands, forearms and in one heel pad. Carbamazepine 200 mg daily was then substituted for diphenylhydantoin. This regime had to be rapidly terminated, however, following the appearance of an acute cutaneous vasculitis associated with fever, a raised ESR and joint pains which ultimately necessitated treatment with systemic corticosteroids. LE cell preparations and anti-nuclear factor tests were negative. The attacks of diaphragmatic flutter continued unabated and surgical therapy was proposed. Since crushing of the phrenic nerve had previously produced only temporary relief from symptoms, excision of a portion of the left phrenic nerve in the neck was performed by Professor M. H. Irving. The attacks of diaphragmatic flutter immediately ceased and postoperative screening revealed paralysis of the left hemidiaphragm. The patient's symptoms have not recurred some 3 months after surgery.

Discussion

Diaphragmatic flutter is a rarely diagnosed condition, only some fifty cases having been reported in the literature, and the present case demonstrates many of the classical features which have been reviewed by Rigatto and DeMedeiros (1962). This rarity may be more apparent than real, however, Graber and Sinclair-Smith (1965) having observed five cases in a period of 18 months, and it seems likely that the bizarre symptomatology often prominent in this disorder may lead to misdiagnosis. Antony Van Leeuwenhoek (1723a, b), the Dutch inventor of the microscope, first recorded diaphragmatic flutter, from which he suffered, in 1723. He refuted his physician's diagnosis of palpitations of the heart by noting that his pulse was not disturbed during an attack and correctly concluded that the diaphragm was the source of his symptoms. Misdiagnosis appears to be common, and in the present case 11 years elapsed before the correct diagnosis

was reached. The duration of symptoms, some 18 years, also appears to be one of the longest recorded. The most prominent symptom is usually pain, and Van Leeuwenhoek recorded that during an attack he 'felt death to be most certain'. The pain has been likened to, and even confused with, the pain of angina pectoris (Porter, 1936; Whitehead, Burnett and Lagen, 1939) or myocardial infarction (Goodman, 1941; Moore and Schoff, 1947) which it may resemble not only in intensity, but also in distribution, radiation down the left arm being noted in several cases including the present one (Porter, 1936; Whitehead *et al.*, 1939). In the present patient radiation round the chest wall to the back at the level of the attachment of the diaphragm was also prominent.

Epigastric pulsations and abdominal splashing sounds are perhaps the best clue to diagnosis but are seldom as marked as in the case reported here where they were of startling intensity. The prominence of these features is presumably related to the force and amplitude of the abnormal diaphragmatic contractions. Tachypnoea not infrequently occurs (Rigatto and DeMedeiros, 1962) and normal diaphragmatic respiratory movement is occasionally abolished (Skillern, 1931; Dowman, 1927; Gamble, Pepper and Muller, 1925; Bird, 1927) as in this case, although more often the flutter is merely superimposed on normal respiratory movements. The contractions of the diaphragm are characteristically irregular and unrelated to the cardiac cycle except in states of severe metabolic alkalosis (Rigatto and DeMedeiros, 1962; Soderstrom, 1945). The rate is often greater than in this case, and can be up to 400/min (Rigatto and DeMedeiros, 1962).

ECG abnormalities have occasionally been noted, either as rhythmical interference with the base line level (Phillips and Eldridge, 1973) which is presumably a movement artefact, or as T wave inversion with ST segment abnormalities (Porter, 1936;

Soderstrom, 1945). In the present patient the QRS axis was unaffected by the flutter, but T wave inversion developed in leads III and V1-V4, together with a sinus tachycardia. P waves were unchanged. The mechanism of these changes is unclear but since the cardiac rate was much faster than the flutter rate, a purely movement artefact seems unlikely.

The aetiology of diaphragmatic flutter is in many cases obscure. Its occurrence during the course of encephalitic illness is well documented (Skillern, 1931; Dowman, 1927; Gamble *et al.*, 1925), and in some cases it has been associated with local, often intrathoracic, lesions such as cervical rib (Hunt 1909), fractured xiphoid process (Bird, 1927), rheumatic heart disease (Smith, 1932), and pleurisy and enlarged mediastinal lymph nodes (Rigatto and DeMedeiros, 1962). However, in the majority of reported cases, no such factors were noted, and several features such as the failure of attacks to occur during sleep, precipitation by emotion or excitement and 'neurotic tendencies' have led to the suggestion that psychogenic factors may be important. However, consistently effective precipitating and relieving factors are uncommon and long periods of misdiagnosis and referral to innumerable doctors may well play a part in producing this impression.

The demonstration by Rigatto and Correa (1968) and Phillips and Eldridge (1973) of widespread electromyographic abnormalities affecting not only the diaphragm but also the scalene, intercostal and abdominal muscles in this condition has placed our understanding of its aetiology on a firmer footing. Phillips and Eldridge have suggested that diaphragmatic flutter is one feature of a more widespread myoclonic disorder involving all the respiratory muscles, and their patient also exhibited abnormal post-hyperventilation apnoea, indicating a disordered neural respiratory control mechanism. Unfortunately, EMG studies were not practicable in the present patient and since in both the cases in which these abnormalities were reported, the diaphragmatic involvement was bilateral, it is unclear whether similar findings are to be expected in cases of unilateral flutter, although it is known that progression to bilateral involvement can occur (Smith, 1932).

Drug therapy in diaphragmatic flutter has been disappointing until the recent apparently successful use of diphenylhydantoin by Phillips and Eldridge (1973). Its effect in the present patient could not be ascertained because of a hypersensitivity reaction. Crushing of the phrenic nerve may produce only temporary relief from symptoms, and as in the present case, excision of the nerve in the neck may be

the only effective manoeuvre. However, this may be inadvisable, particularly in the elderly in whom chronic lung disease may be present. Newer drugs, such as sodium valproate (Epilim) which raises brain concentrations of the naturally occurring inhibitory transmitter γ -aminobutyric acid, and dantrolene (Dantrium) which relaxes skeletal muscle, possibly by a direct effect on muscle fibres, may be worth a therapeutic trial in this condition.

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