clinical manifestations of the irritable colon syndrome. Kirsner and Palmer (1958) viewed the disorder of colonic function as a reflection of an imbalance of the autonomic nervous system, on a psychogenic basis. Emotional stress may be attended by contraction of the longitudinal and circular muscle of the gut (Grace, Wolf and Wolff, 1951). Manometric studies have demonstrated altered colonic activity in states of emotional conflict; excessive motor activity was evoked by aggressive or hostile sensations whereas reduced activity was evident in depressive reactions (Almy, Abbott and Hinkle, 1950). An exaggerated colonic response follows an injection of prostigmine in patients with the irritable colon syndrome, even in the symptom-free phase, possible indicative of a state of parasympathetic over-activity as an accompaniment of emotional stress (Chaudry and Truelove, 1961). The psychological factors participating in the genesis of the irritable bowel syndrome were analysed by Chaudhary and Truelove (1961). In women, more frequently involved than men, the psychological stress was principally related to their families, whereas in men it was largely related to their careers. A distinctive personality pattern in the splenic flexure syndrome has been suggested by Palmer (1963) distinguishable from other forms of the irritable colon syndrome and resembling that encountered in peptic ulcer subjects. He observed excessive self-confidence, hostility and general tension without overt nervousness; often there was a degree of cynicism with resentment of the medical explanation of the symptomatology. The diagnostic difficulties were manifested in the range of diagnoses preferred by the referring practitioners, and in the number of investigations which had been conducted. There was a considerable incidence also of negative exploratory laparotomies. The simulation of cardiac disease is of great significance to which reference has already been made. As Fishberg (1954) indicates, while there is a decreasing tendency to mistake cardiac disease for indigestion, there is a growing tendency to err in the opposite direction.

Treatment is directed largely to the reassurance of the patient of the absence of serious organic disease. Psychotherapeutic measures may be necessary in the individual case. Antispasmodics, sedatives and antidepressants have yielded somewhat varied but generally disappointing results. Asafetida, prescribed in a dose of 0.3 g. with each meal, was found successful in 50% of cases by Palmer, Deutsch and Scott (1955) although they were unable to offer a satisfactory pharmacological explanation for this response.

Summary

The clinical features of the splenic flexure syndrome are described and the underlying causative mechanism discussed. The importance of distinguishing it from cardiac and other forms of abdominal disease and its benign nature are stressed.

REFERENCES


FAMILIAL INCIDENCE OF DIFFUSE INTERSTITIAL PULMONARY FIBROSIS

R. H. Ellis, M.D., M.R.C.P.

from the Gloucestershire Royal Hospital

First described by Hamman and Rich (1944) as an acute progressive condition, diffuse interstitial pulmonary fibrosis is now considered to be more commonly a chronic disease.

The clinical, radiographic, physiological and histological features are fully appreciated by physicians interested in chest disease, and increasing numbers of cases are being diagnosed.

In a recent description of the largest series of cases so far published in this country, Livingstone,
Lewis, Reid and Jefferson (1964) found no familial incidence of the disease, and a search of the British medical literature for such cases, has proved negative.

In America, however, adult twin sisters suffering from the disease were reported by Schechter (1953), who also described two brothers from another family. Rubin and Lubliner (1958) reviewed the literature five years later, and collected 54 cases of the disease, of whom 15 were their own, discovering six siblings amongst them. In a further review, Donohue, Laski, Uchida and Munn (1959) found 12 siblings among 87 cases, of whom two were mother and daughter. In 53 out of 73 of the cases, no family history was recorded, and these authors felt that a higher familial incidence would have been found had this been done. They also described five children from three families with an unusual form of interstitial pneumonitis, and in these three families, five adults were subsequently found to have diffuse interstitial fibrosis of the lungs.

Case Reports

On Examination: colour good, no clubbing, fine crepitations especially well heard over lower lobes. X-ray: diffuse mottling and translucencies maximal at bases and periphery of lungs. Admitted to hospital.


Necropsy: Cor pulmonale, purulent tracheobronchitis, diffuse fibrosis and cystic changes throughout both lungs, especially marked in posterior and basal parts. Histology: diffuse fine interstitial fibrosis leading to replacement of lung tissue, emphysematous cysts and bronchiolectasis.

On Examination: marked finger clubbing, fine crepitations heard over both lower lobes. X-ray: mottling involving mainly the periphery of the lung fields. Admitted to Hospital.

TABLE 1

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>59</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>Absent</td>
</tr>
<tr>
<td>Finger Clubbing</td>
<td>Present</td>
</tr>
<tr>
<td>Crepitations</td>
<td>Coarse mottling &amp; transluences of both lungs</td>
</tr>
<tr>
<td>X-ray</td>
<td>Present</td>
</tr>
<tr>
<td>Polycythaemia</td>
<td>Present</td>
</tr>
<tr>
<td>Hyperglobulinæmia</td>
<td>Normal</td>
</tr>
<tr>
<td>ECG</td>
<td>Right ventricular strain and ischaemia</td>
</tr>
<tr>
<td>Ventilation Tests</td>
<td>Reduced inspiratory capacity without airways obstruction</td>
</tr>
<tr>
<td>Diffusion Tests</td>
<td>Not done</td>
</tr>
<tr>
<td>Corticosteroid therapy</td>
<td>No effect</td>
</tr>
<tr>
<td>Histology</td>
<td>Interstitial fibrosis epithelialised cysts and bronchiolectasis</td>
</tr>
</tbody>
</table>

Discussion

The two brothers presented differing pictures of the disease. Case 1, the elder, came from a lower income group. His symptoms of breathlessness and dry cough had been present for a short time only. His condition deteriorated rapidly and he died of cor pulmonale after only 18 months. There was no change in the extent of radiographic abnormality, which took the form of mottling and transluences, and which was already extensive by the time that he was first seen.

His brother enjoyed a better standard of living, and his breathlessness and cough had been present for more than two years when he first appeared in the Chest Clinic. His radiographic mottling was confined to the bases and peripheral parts of the lung fields. In spite of this, and unlike his brother, he had marked finger clubbing, and his exercise tolerance is steadily diminishing.

Both men presented with the typical symptoms of exertional breathlessness without wheeze and a dry cough, and crepitations were especially audible over the lower halves of their lungs. Both had polycythaemia without obvious cyanosis when first seen, though this developed terminally in Case 1 with the onset of cor pulmonale. Neither had evidence of airways obstruction on ventilation tests, and neither obtained any real benefit from corticosteroid therapy.

The histological findings after autopsy in Case 1 confirm the diagnosis, and are grade 4-5 in degree, as described by Livingstone and others (1964). Epithelialised air spaces, considered by Spencer (1962) to be a feature of the disease, are evident. According to Livingstone and others (1964), the diagnosis can be made, in the absence of histological proof, upon the clinical and radiographic signs in the presence of a diffusion defect. The diagnosis in Case 2 is based upon these criteria in the absence of any evidence pointing to a different condition.

Summary

Diffuse interstitial pulmonary fibrosis occurring in two brothers is described.

The course of the disease was different in the two cases. The elder brother had a short history of breathlessness and much more extensive radiographic changes. He pursued a rapid downhill course and died within 18 months of cor pulmonale and myocardial ischaemia without radiographic deterioration. The diagnosis was confirmed by histological evidence obtained at autopsy.

The younger brother had a longer history, less radiographic abnormality, and remains reasonably well, though his exercise tolerance is slowly diminishing. The diagnosis in this case rests upon the clinical and radiographic findings, in the presence of a diffusion defect.

The main interest of these two cases lies in the fact that familial incidence of diffuse interstitial pulmonary fibrosis has not hitherto been described in this country.

My thanks are due to Dr. K. F. W. Hinson, Brompton Hospital, and Dr. C. T. Vincent, Gloucestershire Royal Hospital, for pathological advice in Case 1, and to Dr. L. H. Capel, London Chest Hospital for his help with the respiratory physiology in Case 2.

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Familial Incidence of Diffuse Interstitial Pulmonary Fibrosis

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