

Spontaneous pneumothorax in young subjects

A clinical and pathological study

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Spontaneous pneumothorax may complicate lung disease which is clinically or radiologically apparent in patients suffering from chronic bronchitis, asthma, tuberculosis, bronchiectasis, and, less commonly, bronchial cancer, silicosis, pulmonary infarction, and other more rare disorders. These patients are usually in the older age group, and the commonest predisposing factor is chronic bronchitis. Pneumothorax occurring in these diseases is well recognized and needs no further elaboration.

Spontaneous pneumothorax may also occur in apparently healthy people with no demonstrable pulmonary lesion. The subjects are often young, usually male, and have been in good health prior to their first episode. They are often athletic and tend to be of tall, thin physique. A group of 20 cases which falls into this latter category forms the basis of this study. They were all treated by wedge resection of apical lung disease. The clinical and histological findings are presented and the literature is briefly reviewed.

Approximately 120 patients were admitted to the Thoracic Surgical Unit of the Wakari Hospital with spontaneous pneumothorax between the years 1962 and 1968. Most of these patients had clinical and radiological evidence of established bilateral pulmonary disease such as bronchitis and emphysema, bronchiectasis or tuberculosis and have not been included in this study.

Fifty of the patients stimulated particular interest because they gave no antecedent history of chest disease and they form the basis of this study. More than half of this group responded to therapy by intercostal tube drainage. In six the air leak persisted despite drainage; 14 patients were readmitted because of recurrent pneumothorax. These 20 patients, two of whom suffered bilateral pneumothoraces, were subjected to thoracotomy and wedge resection. The clinical and pathological data from this group have been reviewed.

CLINICAL DATA

Relevant clinical findings are summarized in the Table.

The indication for resection was recurrence in 14 patients and persistent leak in six. The pneumothorax was bilateral in two patients. Each lung was affected with approximately equal frequency.

Predilection for the male sex is noted, only three of the patients being females. Sixteen were between the age of 16 and 30 (range 16 to 47). Two of the older patients were females.

Measurements of body stature were not recorded but most of the patients tended to be tall and thin. A history of athletic activity was frequent.

OPERATIVE FINDINGS

At operation the characteristic findings were as follows:

At the apex of the lung there was a small area of fibrosis, usually no larger than 3×2 cm, surmounted by a thin-walled bullous cyst or cysts. Commonly there were several cysts, and these measured from about 0.2 cm in diameter to 1 cm or more (Fig. 1). Only occasionally was a small pinhole leak apparent at the apex of the cyst. In the case of leaks that failed to seal within 48 hours of drainage, a tear was sometimes found in a large cyst that had been responsible for considerable air leak and failure to heal. The remainder of the lung appeared normal. Adhesions were uncommon.

PATHOLOGY

The histological material from the specimens in the series has been reviewed and a similar pattern of abnormalities was observed throughout.

TABLE
SUMMARY OF CLINICAL FEATURES

Case	Age/Sex	Side Affected	No. of Previous Episodes	Clinical Presentation	Histological Findings	Follow-up
1	18 M	Right	1	One previous episode of spontaneous pneumothorax. Recurred 10 mth later. Wedge resection for recurrence	Extensive fibrosis and active chronic inflammation with linear collapse and lymphocytic infiltration and oedema. Prominent emphysema with large cysts, some with surrounding chronic inflammation. Numerous intra-alveolar macrophages. Mesothelial and alveolar cell proliferation. Dilated bronchi filled with mucus. Vessels showed endarteritis	18 mth No recurrence
2	25 M	Left	3	Three previous episodes of spontaneous pneumothorax. Wedge resection for further recurrence	Widespread fibrosis with active patchy chronic inflammation. Emphysema with fibrous-walled subpleural cysts. Abundant pigmented macrophages. Distended bronchi containing mucus. Marked endarteritis obliterans	7 yr No recurrence
3	27 M	Left	1	Previous pneumothorax 2½ yr earlier treated by intercostal tube drainage. Wedge resection for recurrence	Extensive linear fibrosis and collapse incorporating distorted bronchioles. Emphysema not seen. Abundant pigmented macrophages. Superficial cysts	3 yr No recurrence
4	21 M	Left	1	Previous episode of spontaneous pneumothorax 5 wk earlier. Further episode, treated by intercostal tube drainage with continuing air leak for 2 days. Wedge resection for persistent air leak	Patchy fibrosis. One scar contained polymorphs surrounded by chronic inflammation. Focal emphysema. Pigmented macrophages and phagocytes in relation to cholesterol. Alveoli have prominent cellular lining. Cysts lined by simple fibrous tissue	18 mth No recurrence
5	24 M	Right	-	First episode of pneumothorax. Treated by intercostal tube drainage. Air leak continued for 6 days, when wedge resection was done for persistent air leak	Widespread fibrosis and chronic inflammation. Patchy emphysema with fibrous walled cysts. Pigmented macrophages. Collapsed distorted bronchi in scars. Vessels show endarteritis	7 yr No recurrence
6	19 M	{Left Right	2	Left side: Two previous episodes of left spontaneous pneumothorax and one on right. Wedge resection on left for further recurrence	Severe focal fibrosis with intense chronic inflammation. Emphysema and many fibrous-walled cysts. Mesothelial, bronchiolar and alveolar cell proliferation.	3 yr No recurrence
				Right side: One year later presented after 6 episodes of right-sided spontaneous pneumothorax, 4 of which had occurred within the past 2 mth. Wedge resection for recurrence	Abundant pigmented macrophages. Many bronchioles which contain macrophages and pigment. Vessels show endarteritis	2 yr No recurrence
7	19 M	Left	7	Seven previous episodes of spontaneous pneumothorax over previous 4 yr. Wedge resection for recurrence	Focal fibrosis marked with patchy chronic inflammation. Emphysema and fibrous-walled cysts. Abundant pigmented macrophages with focal cholesterol deposition. Alveolar cell proliferation. Dilated bronchioles containing macrophages	5 yr No recurrence
8	24 F	Right	-	Tension pneumothorax treated by intercostal tube drainage. Air leak continued for 6 days. Wedge resection for persistent air leak	Large subpleural scar with underlying emphysema. Solitary fibrous-walled cyst with pigmented macrophages in its wall	7 yr No recurrence
9	16 M	Left	2	Two episodes of spontaneous pneumothorax over previous 7 mth. Wedge resection for further episode	Fibrosis involving almost entire specimen. Emphysema not seen. Focal chronic inflammation in scars. Cysts only on surface	8 yr No recurrence
10	20 M	Right	2	Two previous episodes of spontaneous pneumothorax over preceding 2 mth treated by intercostal tube drainage. On second occasion camphor oil was instilled into pleural cavity. Third episode treated by wedge resection	Multifocal scarring with florid chronic inflammation and lipoid granulomata (history of camphor oil instilled into pleural cavity). Severe focal emphysema. Fibrous-walled cysts. Active alveolar cell proliferation. Collapsed and distorted bronchioles containing secretion. Severe endarteritis of small vessels	3 yr No recurrence
11	31 M	Right	1	One previous episode of spontaneous pneumothorax 5 mth earlier. On second occasion treated by intercostal tube drainage. Continued air leak over 2-wk period. Wedge resection for persistent air leak	Subpleural and intrapulmonary fibrosis. Alveolar cell hyperplasia in scarred areas. Single cyst lined by fibrous tissue with haemosiderin and cholesterol deposition suggesting old haemorrhage	4 yr No recurrence

(cont.)

Table continued

Case	Age/Sex	Side Affected	No. of Previous Episodes	Clinical Presentation	Histological Findings	Follow-up
12	20 M	Right	3	Two episodes of spontaneous pneumothorax over previous 2 mth. Third episode treated by wedge resection	Widespread focal fibrosis with patchy chronic inflammation. Emphysema with fibrous-walled cysts, some lined by cellular layer. Alveolar walls lined by prominent cells. Dilated bronchi containing macrophages	5 yr No recurrence
13	47 M	Right	-	First episode of spontaneous pneumothorax treated by intercostal tube drainage. Continued air leak for 3 days. Wedge resection for persistent air leak	Extensive fibrosis with active chronic inflammation and lymphoid hyperplasia. Emphysema with cysts lined by prominent cellular layer. Abundant pigmented macrophages. Alveolar cell hyperplasia in scars. Small thick-walled bronchi. Vessels show marked endarteritis	7 yr No recurrence
14	21 M	Left	1	Second spontaneous pneumothorax within 5 wk. Treated by wedge resection	Extensive focal fibrosis with a solitary cavity surrounded by macrophages. Widespread emphysema. Numerous pigmented macrophages. Alveolar cell proliferation. Severe endarteritis	4 yr No recurrence
15	37 F	Right	1	Spontaneous pneumothorax 5 yr earlier. Second episode treated by wedge resection	Intrapulmonary fibrosis with intense chronic inflammation. Emphysema with large fibrous-walled cysts. Abundant pigmented macrophages. Mesothelial, bronchiolar, and alveolar cell proliferation. Bronchioles contain mucus and pigmented macrophages. Endarteritis of small vessels	3 yr No recurrence 3 mth later pneumothorax on contralateral side
16	33 F	Left	1	Second episode of spontaneous pneumothorax within 2 mth. Treated by wedge resection	Large areas of fibrosis with adjacent emphysema. Pigmented macrophages in scars. Chronic bronchiolitis with mucus plugs. Endarteritis of small vessels. Cysts lined by simple fibrous tissue	5 yr No recurrence
17	26 M	Left	2	Two episodes of spontaneous pneumothorax over previous 3½ yr. Third spontaneous pneumothorax treated by wedge resection	Patchy fibrosis and active chronic inflammation with focal emphysema. Abundant pigmented macrophages. Cysts lined by flat cellular layer	6 yr No recurrence
18	19 M	Left	1	Three episodes of spontaneous pneumothorax over previous year. Further recurrence treated by wedge resection	Patchy fibrosis and emphysema, pigmented macrophages, small fibrosed bronchi. Cysts lined by prominent cellular layer	5 yr No recurrence
19	26 M	Left	3	Three episodes of spontaneous pneumothorax over previous year. Further recurrence treated by wedge resection	Patchy fibrosis and emphysema. Alveolar and bronchiolar cell proliferation, pigmented macrophages, distended bronchi with chronic inflammation. Cysts lined by fibrous tissue	5 yr No recurrence
20	18 M	Left Right	4-5 5-6	Left side: Asthma since age 13. Five episodes of spontaneous pneumothorax on left side and one on right over previous 2 yr. Further recurrence on left side treated by wedge resection Right side: Admitted 18 mth later after 4 episodes of spontaneous pneumothorax on right side. Wedge resection for recurrence	Dense vascular subpleural scar with patchy chronic inflammation. Emphysema and subpleural fibrous-walled cysts. Distended bronchi with cartilage in walls. Vessels show endarteritis in one biopsy only	7 yr No recurrence 5 yr No recurrence

In each case the portion of lung was disorganized by fibrosis, collapse, and cyst formation, all variable in degree. There was no evidence of adhesions to the chest wall.

MICROSCOPIC FINDINGS Histologically the changes were non-specific. Although common features were present, these varied in prominence from case to case, suggesting that the process was in different stages of progression at the time of presentation.

The changes consisted of emphysema and cyst formation, atelectasis, fibrosis, chronic inflammation, pigment deposition, bronchiolar, alveolar cell, and mesothelial proliferation, bronchial lesions, and vascular changes.

Emphysema and cyst formation Emphysema was constant and focally distributed, being compensatory in some areas and destructive in others (Fig. 2). Cysts were usually lined by fibrous tissue (Fig. 3)

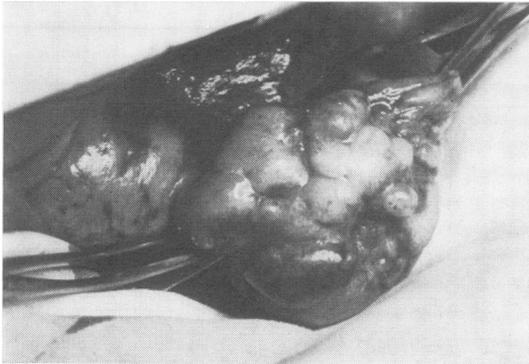


FIG. 1. Appearance at operation showing cysts and scarred lung tissue.

but a prominent mesothelial layer was present in some instances (Fig. 4). Cysts were usually subpleural but occasionally were seen in scars.

Atelectasis This was a constant feature, being distributed either in linear fashion or in ill-defined circular

areas. Atelectatic foci were often invaded by fibrous tissue and were in many instances related to adjacent compensatory emphysema.

Fibrosis This process varied in nature and distribution. Dense subpleural scars were common (Fig. 5) but in addition the lung tissue itself was fibrosed with replacement of architecture (Fig. 6). In other areas fibrous tissue caused thickening of alveolar walls, especially in foci of atelectasis.

Chronic inflammation Active infiltration of dense scar tissue was seen in some cases but this was not the general rule. When present, the cells consisted mainly of lymphocytes and plasma cells, polymorphs being inconspicuous (Fig. 6). Intra-alveolar macrophages were seen frequently. In case 10 there was florid inflammation with foamy macrophages. This patient had been treated elsewhere by camphor instillation.

Pigment deposition This was inconstant and, when present, was found in macrophages, both in scar tissue and also lying free in air spaces. Both haemosiderin and carbon were identified (Fig. 7).

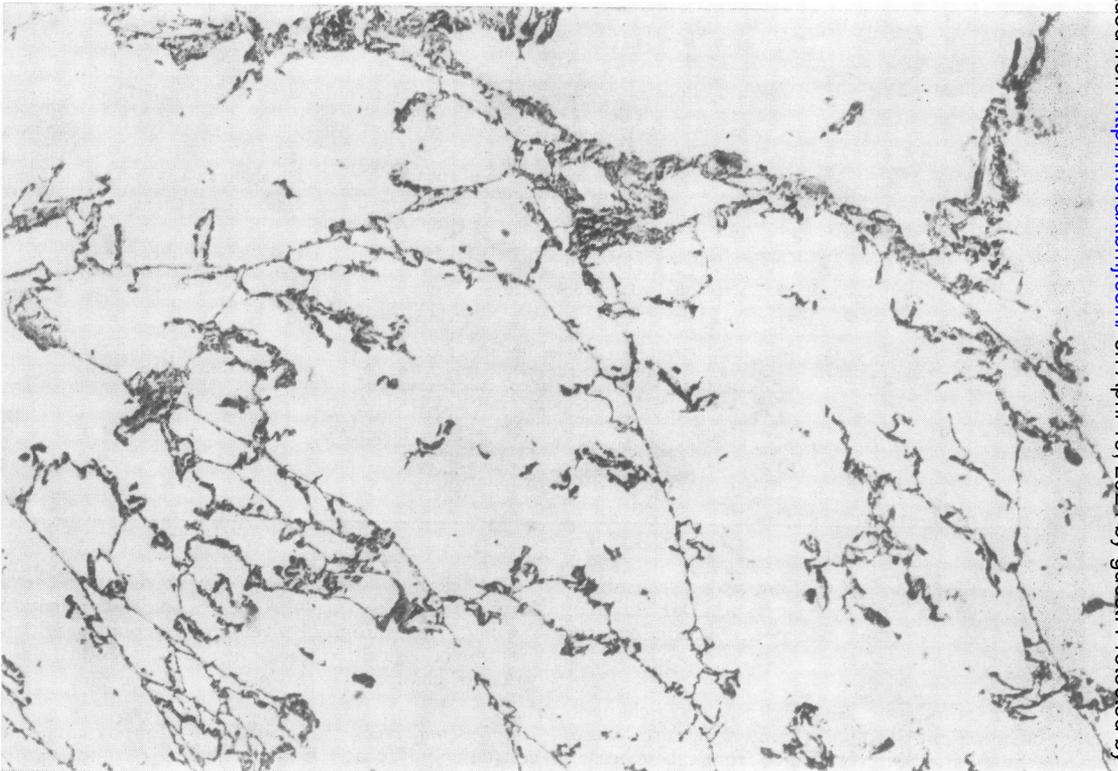


FIG. 2. Photomicrograph showing compensatory emphysema. H. and E. $\times 44$.

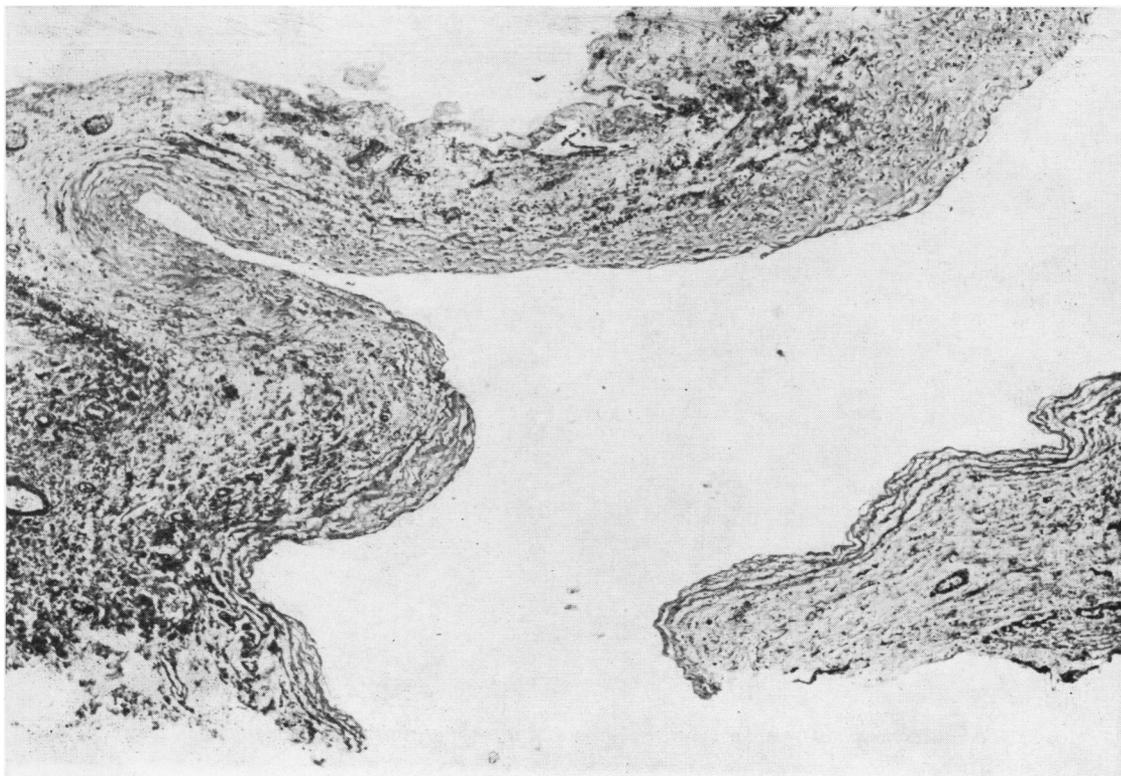


FIG. 3. *Photomicrograph of subpleural cyst lined by fibrous tissue. H. and E. × 48.*

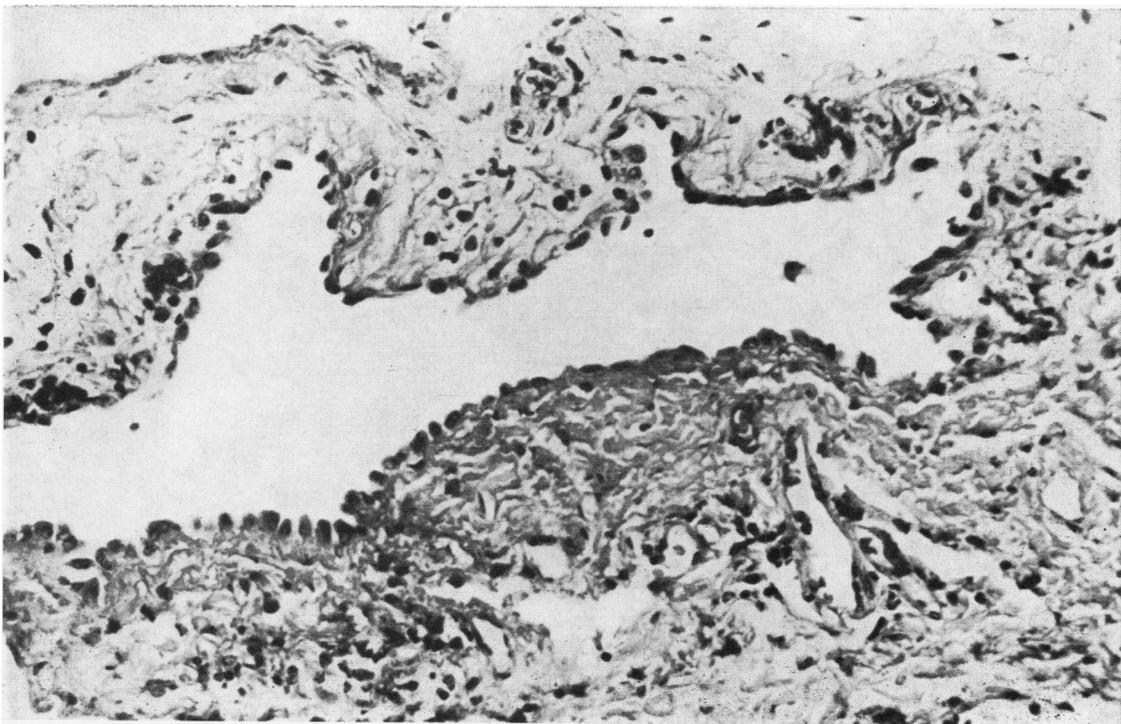


FIG. 4. *Photomicrograph of cyst lined by prominent mesothelium. H. and E. × 226.*

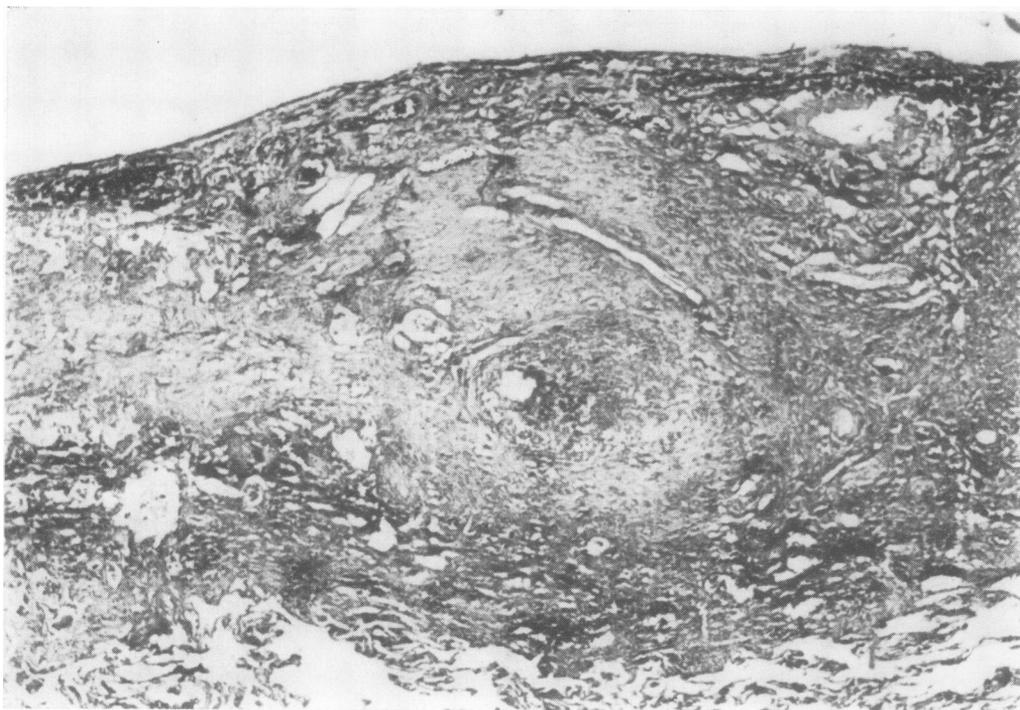


FIG. 5. *Photomicrograph of vascular subpleural scar with a focus of residual inflammation. H. and E. × 50.*

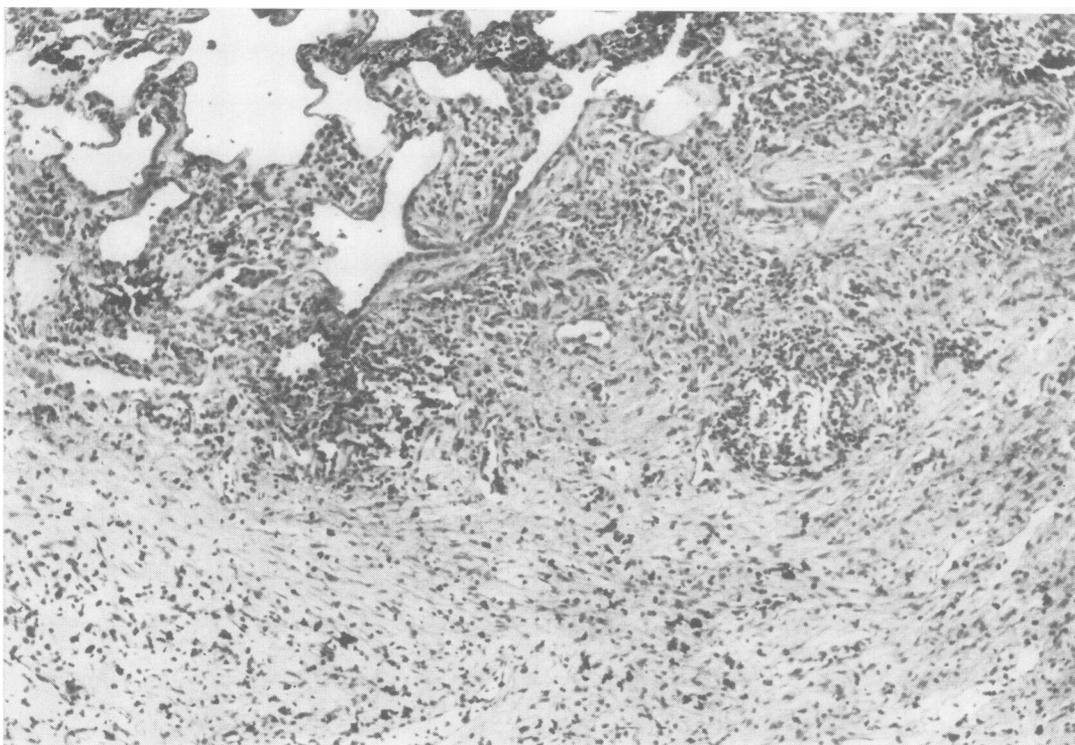


FIG. 6. *Photomicrograph of focus of cellular scar tissue with residual inflammation. There is fetalization of an adjacent alveolar lining. H. and E. × 115.*

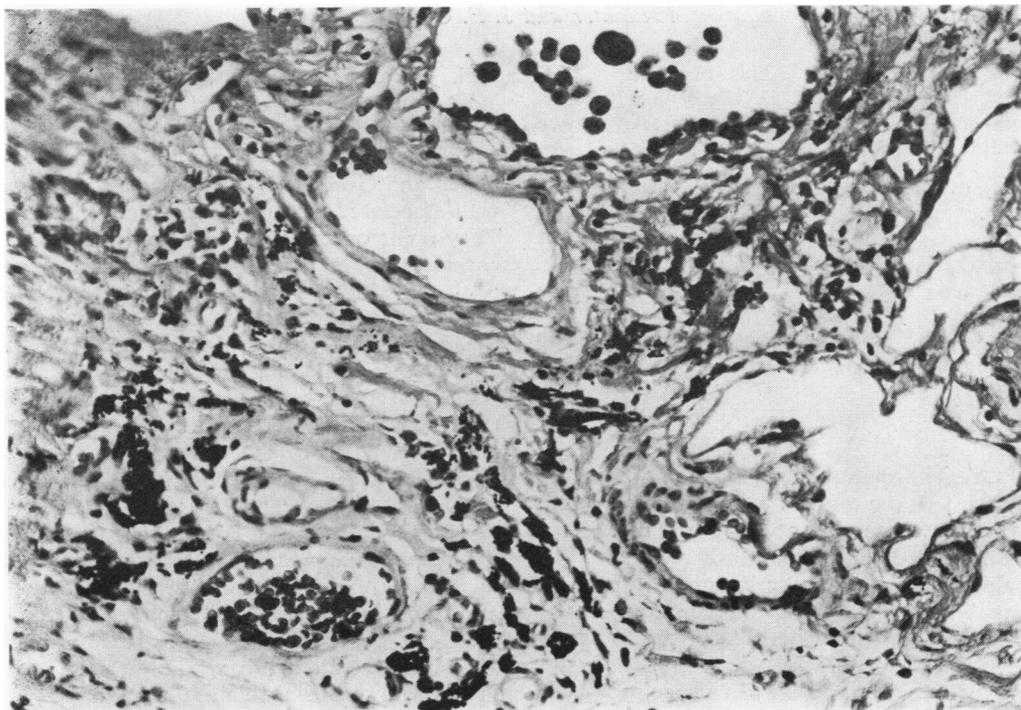


FIG. 7. *Photomicrograph of pigmented macrophages in alveolar space. Pigment is also deposited in adjacent fibrous tissue. H. and E. × 256.*



FIG. 8. *Photomicrograph of small bronchi distended with secretion. There is peribronchial inflammation. H. and E. × 115.*

Alveolar cell proliferation This occurred in disorganized scarred areas (Fig. 6). The cellular reaction was considered to be related to nearby scarring and inflammation.

Bronchial lesions When bronchi were found in the biopsies, they were on occasion filled with eosinophilic secretion as though obstructed by scar tissue (Fig. 8). Some showed peribronchial fibrosis and mild inflammation of their walls.

Vascular lesions No specific inflammatory or occlusive lesions were seen but endarteritis obliterans was common.

DISCUSSION

AETIOLOGY The special site of the damage in each case suggested the possibility of a tuberculous aetiology, but there was no histological evidence for this in any of the cases.

Localized congenital cystic lung was entertained as a possible explanation but the constant relation of cysts to scar tissue made this difficult to assess.

Post-inhalational damage was excluded by the site and by the absence of any evidence of inhaled material apart from carbon.

Localized trauma to the lung suggested itself in view of the youth of the patients and the history of athletic activity, but there was no real evidence of organizing haematomata. The finding of haemosiderin pigment was inconstant and insufficient to explain the findings on a basis of haemorrhage.

On histological grounds alone the changes are best explained on a basis of post-inflammatory disorganization, the inflammation being most likely due to non-specific infection. The curious localization of the lesions and their occurrence predominantly in young males are obscure features which suggest some local inherent predisposition.

Many of the previous reports relating to primary spontaneous pneumothorax in young subjects have not included detailed accounts of the histological changes (DuBose, Price, and Guilfoil, 1953; Bernhard, Malcolm, Berry, and Wylie, 1962; Carpathios and Bogedain, 1963; Leading article, 1965; Lynn, 1965; Mills and Baisch, 1965; Levy, 1966; Shields and Oilschlager, 1966).

Most authors have favoured a congenital origin for the cysts or have postulated inflammation and fibrosis with cyst formation. An earlier hypothesis by Brock (1948), that air may leak through an imperfect visceral pleura, has not been reiterated in recent studies, all of which record the presence of bullae.

The hyposthenic habitus of the patients is of interest and it has been suggested (Withers, Fish-

back, Kiehl, and Hannon, 1964) that in tall thin individuals there is a rapid growth rate relative to pulmonary vasculature accounting for relative ischaemia and bleb formation during growth. Killen and Gobel (1968) describe bleb formation with associated fibrosis and chronic inflammation. They point out that the aetiology is unknown but consider that a congenital fault may be present or that the lesion may be secondary to local stress injury or degenerative change. Similarly, it was suggested in the *British Medical Journal* (Leading article, 1968) that the lesion might result from a congenital fault or an inflammatory scar. Hyde (1963) drew attention to the long narrow chest and related this to the development of blebs as a congenital anomaly. The same author (1962) had noted the fact that the patients were 20–30 lb (9–13 kg) underweight. Aust (1961) presumed that the blebs were congenital in nature. Thomas (1959) divided his cases into those with congenital cysts and those with acquired blebs or bullae. He indicated that loss of elasticity, fibrosis, and adhesions resulted from pulmonary vascular insufficiency, causing bleb formation.

None of the above authors has incriminated tuberculosis in the aetiology of primary pneumothorax, and this disease was not in evidence in any of our cases.

INCIDENCE The true incidence of apical lung cysts in the population is not known, recognition depending on the complication of rupture. Radiological study prior to the development of pneumothorax is usually negative (Baronofsky *et al.*, 1957; Bernhard *et al.*, 1962; Reid, Stevenson and McSwan, 1963; Leading article, 1968). Our own experience confirms the frequent absence of radiological changes.

According to a leading article in the *British Medical Journal* (1968) the incidence of the complication of pneumothorax rose from 0.25 per thousand in the 1950s to 0.4 per thousand in the 1960s. This is probably due to better recognition of the condition. Several large series have been reported from military sources and these all stress the importance of the condition in young apparently fit men (Thomas, 1959; Withers *et al.*, 1964; Mills and Baisch, 1965).

The frequency of involvement of both sides at different times in the same patient is worthy of note, and suggests that bullous disease may be present in an intact state on the contralateral side in a significant proportion of patients with apparently unilateral disease. Pneumothorax occurred bilaterally in two of our patients (10%) and this

may rise to a higher figure as our patients are followed up for longer periods. Baronofsky *et al.* (1957) were so impressed with the bilateral nature of the bullous disease that they performed thoracotomy on both sides in each of their 26 patients. Bullae were found on both sides in all but one of their patients. Details of the pathological findings in their series were not given.

THERAPY Wedge resection of diseased lung tissue as used in this series of patients reflects the localized apical nature of the disease process which caused the pneumothorax. Failure to understand the focal affection of lung tissue has in the past led to irrational therapy such as pleural cavity obliteration by the instillation of irritating fluids, a procedure liable to fail. When leakage persists for longer than 48 hours, or when the pneumothorax recurs after simple intercostal tube drainage, thoracotomy seems to be the only logical approach to permanent cure. So far, in our series, the results have been good with no recurrences. Baronofsky *et al.* (1957) recommended resection while Withers *et al.* (1964) found that the technique compared favourably with other methods. Reid *et al.* (1963) reported similarly when they compared the results of intercostal tube drainage, kaolin pleurodesing agent, and segmental resection.

CONCLUSIONS

1. A series of 20 patients with recurrent or persistent spontaneous pneumothorax is presented from the clinical and pathological standpoints. All the patients were treated by wedge resection of diseased lung tissue.
2. The pathological features have been reviewed and an attempt has been made to explain the aetiology.
3. The histological lesions are non-specific but suggest post-inflammatory fibrosis and cyst formation. An underlying congenital abnormality related to the anatomy of the thorax in tall thin athletic individuals appears to be an attractive hypothesis. Defects in blood supply and aeration in these circumstances may be important factors.
4. Treatment by wedge resection seems to be a rational approach to the underlying pathology.

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