Desquamative Interstitial Pneumonia-Like Reaction Accompanying Pulmonary Lesions*


Six patients with specific pulmonary diseases had pulmonary tissue surgically resected for diagnostic purposes. All six cases were characterized by space-occupying lesions surrounded by a peculiar reaction of the pulmonary parenchyma indistinguishable from desquamative interstitial pneumonia. If small biopsies from these areas had been taken, a diagnosis of desquamative interstitial pneumonia would have been made, and the underlying process would have gone undetected. These cases corroborate even further the concept that desquamative interstitial pneumonia is a pattern of pulmonary reaction, rather than a well-defined disease entity. Furthermore, it seems advisable to make the diagnosis of desquamative interstitial pneumonia only when other conditions have been carefully ruled out by thorough clinical and pathologic investigations.

Desquamative interstitial pneumonia was first described by Liebow and associates1 in 1965 as a clinicopathologic entity with the following features: a diffuse, bilateral interstitial infiltrate with a ground-glass appearance on the chest x-ray films; a uniform microscopic appearance with large numbers of mononuclear cells within the alveolar lumen, in the absence of necrosis or severe thickening of alveolar walls; and a relatively benign clinical course abbreviated by prompt response to steroid therapy. To date, about 120 cases have appeared in the literature, including some variants from the original description, such as (1) several cases that did not respond to steroid therapy,2 (2) some cases that progressed to interstitial fibrosis,3,4 (3) a case that seemingly evolved into pulmonary alveolar proteinosis,5 (4) a case with asbestos bodies in the material from biopsy,6 (5) a localized form of the disease restricted to only one pulmonary segment,7 and (6) a case that apparently followed long-term therapy with nitrofurantoin.7

Due to this variable clinical presentation the existence of desquamative interstitial pneumonia as a separate entity has been questioned by Scadding and Hinson,8 who believe that desquamative interstitial pneumonia is just a phase in the development of diffuse interstitial fibrosis of the lungs. The purpose of this report is to call attention to still another variant of this condition, consisting of a localized form of desquamative interstitial pneumonia found at the periphery of specific pulmonary lesions. To our knowledge, this is the first description of this reaction, which bears obvious implications for the interpretation of small biopsies of the lung.

CASE REPORTS

Table 1 summarizes pertinent clinical and pathologic data in the six cases in which desquamative interstitial pneumonia-like areas accompanied other pulmonary lesions. Thoracotomy in all six cases was performed because of an abnormal chest

<table>
<thead>
<tr>
<th>Case, Age (yr), Sex, Race, Location of Process, Displaying</th>
<th>Radiographic Procedure</th>
<th>Surgical Procedure</th>
<th>Accompanying Lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, 38, W, M, Right lower lobe, of lung, wedge resection</td>
<td>Wedge resection</td>
<td>Rheumatoid nodule</td>
<td></td>
</tr>
<tr>
<td>2, 45, W, M, Bilateral bases of lungs, open lung biopsy</td>
<td>Open lung biopsy</td>
<td>Eosinophilic granuloma</td>
<td></td>
</tr>
<tr>
<td>3, 55, W, M, Right lower lobe, open lung biopsy</td>
<td>Open lung biopsy</td>
<td>Intrapulmonary lymph node</td>
<td></td>
</tr>
<tr>
<td>4, 55, W, M, Right upper lobe, lobectomy</td>
<td>Lobectomy</td>
<td>Chondromatous hamartoma</td>
<td></td>
</tr>
<tr>
<td>5, 49, W, F, Bilateral bases of lungs, open lung biopsy</td>
<td>Open lung biopsy</td>
<td>Eosinophilic granuloma</td>
<td></td>
</tr>
<tr>
<td>6, 43, W, M, Bilateral bases of lungs, open lung biopsy</td>
<td>Open lung biopsy</td>
<td>Eosinophilic granuloma</td>
<td></td>
</tr>
</tbody>
</table>

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x-ray film and the need for a histopathologic diagnosis (Fig. 1). In all cases, either open lung biopsies or lobectomies were performed, yielding pulmonary tissue with consolidated parenchyma at gross inspection. In addition, cases 1, 3, and 4 revealed the characteristic gross appearance of the circumscribed lesions listed in Table 1.

Routine sections stained with hematoxylin-eosin were prepared from paraffin-embedded blocks of pulmonary tissue fixed with formaldehyde solution (formalin), and reactions with PAS and Prussian blue were also obtained. Tissue from three patients (cases 1, 5, and 6) was subjected to ultrastructural study following fixation with osmium tetroxide of blocks fixed with formaldehyde solution, embedding in epoxy resin, sectioning with an ultramicrotome, and staining with uranyl acetate and lead citrate.

All cases revealed well-recognized histopathologic features of the accompanying lesions listed in Table 1, having in common a solid, space-occupying diffuse or nodular con-

FIGURE 1. Chest x-ray film. Note well-circumscribed lesion in right lower lobe surrounded by focal consolidation of parenchyma (case 1).

FIGURE 2. Pulmonary tissue adjacent to lesion seen in Figure 1. Notice rheumatoid nodule surrounded by area indistinguishable from desquamative interstitial pneumonia (hematoxylin-eosin, original magnification × 80).

FIGURE 3. Tissue in right half of Figure 2. Alveoli filled with macrophages and hyperplastic alveolar lining cells are characteristic of desquamative interstitial pneumonia (hematoxylin-eosin, original magnification × 150).

FIGURE 4. Desquamative interstitial pneumonia-like area (case 1). Epithelial cells lining alveolar walls are type 2 (granular pneumocytes). Cells in lumen of air space are free alveolar macrophages with numerous lysosomes (uranyl acetate and lead citrate, original magnification × 5,000).

TABLE 1. Lesions Listed in Figures 1-4

<table>
<thead>
<tr>
<th>Case</th>
<th>Lesion Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Well-circumscribed lesion</td>
</tr>
<tr>
<td>2</td>
<td>Diffuse consolidation</td>
</tr>
<tr>
<td>3</td>
<td>Focal consolidation</td>
</tr>
<tr>
<td>4</td>
<td>Well-circumscribed lesion</td>
</tr>
</tbody>
</table>

Notice rheumatoid nodule surrounded by area indistinguishable from desquamative interstitial pneumonia.

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cytoplasmic lysosomal granules of ordinary free alveolar macrophages and lacked structures of the Langerhans' type. In all cases a diagnosis of desquamative interstitial pneumonia was eschewed because of the recognition of other accompanying pulmonary lesions.

**DISCUSSION**

This report emphasizes the danger of attempting histopathologic diagnoses in limited samples of tissue. It was evident in all six cases that small biopsies from the periphery of the localized lesions would have been interpreted as characteristic desquamative interstitial pneumonia. This pitfall is more likely with the widespread use of transbronchial biopsies, which, indeed, have been used to make the diagnosis of desquamative interstitial pneumonia on at least one occasion.

When originally described, desquamative interstitial pneumonia referred to a diffuse process affecting large portions of the pulmonary parenchyma.1 Scadding and Hinson2 were the first to call attention to the fact that a nonuniform histologic picture, including desquamative interstitial pneumonia-like and usual interstitial pneumonia-like areas, may occur within the same lung. Our current observations indicate that other underlying lesions have not been necessarily ruled out when the pattern of desquamative interstitial pneumonia is noted in small biopsies of the lung.

Currently, a diagnosis of desquamative interstitial pneumonia carries with it a good prognosis and an indication for conservative medical treatment.1 In three of our six cases, the therapy of choice is surgical removal. Administration of corticosteroids, the therapy of choice for desquamative interstitial pneumonia, is clearly without justification in all of the six current cases. This form of therapy could even be harmful if the underlying lesion were infectious in nature. Although not an etiologic agent in any of our cases, tuberculosis has been reported to elicit a perifocal desquamative reaction, histologically indistinguishable from desquamative interstitial pneumonia.10 The potentially disastrous consequences of treating such a case with corticosteroids are obvious.11

The pathogenesis of desquamative interstitial pneumonia-like reactions in the vicinity of pulmonary lesions is not clear, but the cells accumulated in the air spaces are free alveolar macrophages, as previously described by electron-microscopic studies and currently confirmed by our own ultrastructural findings.12 Of the six cases the three localized nodular lesions are obviously not histogenetically related to each other or to the cells accumulated in the alveoli. The three remaining cases of eosinophilic granuloma are of histiocytic origin, but electron-microscopic studies left no doubt as to their derivation from interstitial histiocytes of the Langerhans' type, characteristic of the differentiated histiocytosis.13

Hyperplasia of type 2 cells is now recognized as a nonspecific reaction commonly accompanying damage to type 1 cells. The presence of hyperplasia of type 2 cells, therefore, suggests some epithelial injury. Increased numbers of macrophages in the air spaces can come about by diminished removal via the airways, recruitment from the circulation, or local proliferation. It has been suggested that ventilatory movements may be involved in the clearance of macrophages and other materials from alveoli.14 If this is the case, it would not be surprising that focal lesions producing low compliance in adjacent air spaces would result in accumulation of macrophages. On the other hand, Liebow et al1 clearly demonstrated mitotic activity in the mononuclear cells in the air spaces in diffuse desquamative interstitial pneumonia, suggesting that at least in that condition, proliferation of macrophages in the air spaces is also a factor.

Regardless of its pathogenesis, the occurrence of large areas fulfilling the histologic criteria for desquamative interstitial pneumonia, in association with other pulmonary lesions, adds to the burden of accepting desquamative interstitial pneumonia as a specific diagnosis. These findings, along with the various associated features previously described, indicate that desquamative interstitial pneumonia should probably be regarded as a pattern of pulmonary reaction, rather than a well-defined disease entity.15,16 Until more experience is gained, the best approach is perhaps the cautious posture of making the diagnosis of desquamative interstitial pneumonia only when other possible underlying lesions have carefully been ruled out. This can be accomplished by close scrutiny of the clinical data, as well as by a thorough examination of the pathologic specimen.

**REFERENCES**

5. Corrin B, Price AB: Electron microscopic studies in des-
Certifying Examination in Pulmonary Disease

The American Board of Internal Medicine will administer the certifying examination in the subspecialty of Pulmonary Disease on June 27, 1978. Application forms may be requested beginning August 1 and must be returned by November 1, 1977. Information and application forms are available from: Registration Department, ABIM, 3930 Chestnut Street, Philadelphia 19104.