Endoluminal Stenosis of Proximal Bronchi in Sarcoidosis*

Aim: Endoluminal stenosis of proximal bronchi (ESPB) is a potentially severe manifestation of sarcoidosis. Unusual clinical presentation and variable response to medical treatment require specific attention to diagnosis and follow-up.

Design: Of 2,500 patients with sarcoidosis seen at our institution, we retrospectively identified 18 patients with stage 1–3 sarcoidosis and ESPB. Clinical manifestations, endoscopic findings, pulmonary function tests, follow-up, and therapeutic response were assessed.

Results: Respiratory symptoms were present in 17 patients (94%): cough and dyspnea (89% each), wheezing (83%), and hemoptysis (11%). Generalized symptoms (67%) and extrapulmonary manifestations (72%) of sarcoidosis were frequent. Three bronchoscopic patterns were observed: single stenosis (n = 3), multiple stenoses (n = 12), or diffuse narrowing of the bronchial tree (n = 3). The two former groups accounted for 45 ESPBs located in the left upper lobe (44.5%), the right upper and middle lobes (15.5% each), and the left lower lobe (11%). ESPBs were due to mural thickening of bronchi (n = 16) or associated with extrinsic compression by lymphadenopathy (n = 2). Endobronchial biopsies uniformly confirmed the presence of granulomas. FEV1/FVC ratio was < 70% in 12 patients (66.7%), with a correlation between the decrease of FEV1/FVC ratio and the number of ESPBs (R² = 0.31; p = 0.02). Patients treated with oral corticosteroids (n = 12) or methotrexate (n = 1) within the first 3 months had a good prognosis, whereas patients in whom treatment was delayed by > 3 months (n = 4) or who did not receive any systemic treatment (n = 1) acquired fixed ESPB and persistent ventilatory defects.

Conclusions: ESPB is a rare and serious complication of sarcoidosis. Its clinical hallmarks include multiple respiratory symptoms, multiorgan involvement, and generalized symptoms. Treatment has to be started early to avoid the development of fixed stenotic lesions and irreversible pulmonary function impairment.

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Key words: airway obstruction; bronchi; bronchoscopy; glucocorticoids; prognosis; respiratory function tests; sarcoidosis

Abbreviations: ESPB = endoluminal stenosis of proximal bronchi; PFT = pulmonary function test; T1 = time from the onset of the bronchial symptoms (cough or dyspnea, wheezing, ronchi) to the diagnosis of endoluminal stenosis of proximal bronchi; T2 = time from the diagnosis of endoluminal stenosis of proximal bronchi to the beginning of treatment; TLC = total lung capacity

Sarcoidosis is a systemic granulomatous disease of unknown origin that commonly affects the bronchial tree.1,2 Granulomatous lesions usually occur in the bronchial submucosa, facilitating bronchoscopic diagnosis by either endobronchial and/or transbronchial lung biopsy.3 The bronchial mucosa in sarcoidosis often appears inflamed with small or large nodules containing noncaseating granulomas.4,5 Granulomatous lesions more frequently involve the distal bronchial tree.6 In stage 4 sarcoidosis, bronchial involvement often leads to bronchial narrowing by intricate mechanisms such as endobronchial and/or peribronchial fibrotic lesions, or is rarely associated to an extrinsic compression by fibrotic and calcified lymph nodes.7,8

In stage 1–3 sarcoidosis, the pathophysiologic mechanisms leading to bronchial narrowing are different and consist of various degrees: (1) inflamma-

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*From Services de Pneumologie (Drs. Turbie, Nunes, Battesti, and Valeyre) et Radiologie (Dr. Brauner), Avicenne University Hospital, Bobigny; and Laboratoire des Explorations Fonctionnelles (Dr. Chambellan), Nantes University Hospital, Nantes, France.

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Correspondence to: Dominique Valeyre, MD, Service de Pneumologie, Centre Hospitalo-Universitaire Avicenne, 125 route de Stalingrad, 93009 Bobigny Cedex, France; e-mail: dominique.valeyre@acp.ap-hop-paris.fr
tory, edematous lesions and/or granulomatous infiltration located in the bronchial mucosa, submucosa, and the peribronchial tissue; extrinsic compression by enlarged lymph nodes; or (3) an endobronchial mass lesion. These uncommon manifestations are often associated with airway-related symptoms, obstructive ventilatory defects, and a typical bronchoscopic presentation. The rationale of this study is to retrospectively analyze patients with stage 1–3 sarcoidosis and endoluminal stenosis of proximal bronchi (ESPB). Because of heterogeneous responses to treatment previously reported in the literature, we would like to characterize their clinical presentation, bronchoscopic findings, pulmonary function tests (PFTs), and response to therapy to bring out predicting factors of outcome.

**Materials and Methods**

**Patients**

Of a cohort of 2,500 patients with sarcoidosis evaluated at our institution between 1980 and 2000, we retrospectively identified 18 patients with ESPB. The mean age was 38.8 ± 10.9 years (±SD) [6 men and 12 women]. Eight patients (44.4%) were black. All patients fulfilled the following criteria: (1) sarcoidosis was diagnosed according to the American Thoracic Society/European Respiratory Society criteria (ie, clinicoradiologic findings supported by histologic evidence of nonnecrotizing epithelioid cell granulomas, with exclusion of other known causes of granulomatous lesions), (2) ESPBs were defined as a bronchoscopic narrowing of at least 50% of the bronchial lumen from the main bronchus up to the segmental bronchus, and (3) other mechanisms of airway obstruction were excluded (ie, diffuse interstitial pulmonary fibrosis or predominately extrinsic compression as the mechanism of the ESPB). Two time periods were defined: the time from the onset of the bronchial symptoms (cough or dyspnea, wheezing, ronchi) to the diagnosis of ESPB (T1), and time from the diagnosis of ESPB to the beginning of treatment (T2). Patients were classified in two groups according to time T2. Group A was defined as patients treated within 3 months of the diagnosis of ESPB (T2 < 3 months), and group B was defined as patients in whom treatment was delayed by > 3 months (T2 > 3 months).

**Radiologic Features**

Chest radiographic staging was performed according to the Siltzbach classification (Table 1). Thirteen of 18 patients underwent a high-resolution CT scan at the time of diagnosis. The analysis was focused on the bronchial tree as well as the lung parenchyma and the mediastinum.

**Bronchoscopic Features**

ESPB was diagnosed bronchoscopically. The examination was carried out by one of the authors (P.T.) experienced with tracheobronchial findings in sarcoidosis who was not in charge of the patients. Bronchial narrowing of at least 50% of the lumen was defined as stenosis. According to the number and extent of the stenoses, patients were classified into three groups: single (one ESPB), multiple (at least two ESPBs but not diffuse), and diffuse (the whole bronchial tree was involved). Characteristics such as the macroscopic description of the mucosa (ie, inflammation, infiltration, mass, or scarred lesions), the localization of the stenosis and possible extrinsic compression of the bronchi

**Table 1—Clinical and Radiologic Findings of the 18 Patients at Baseline**

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex/Age, yr</th>
<th>Geographic Origin</th>
<th>Smoking History, pack-yr</th>
<th>Respiratory Symptoms*</th>
<th>Radiographic Stage†</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Group A</strong></td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Female/35</td>
<td>African</td>
<td></td>
<td>C, D(1), W</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>Male/54</td>
<td>West Indian</td>
<td></td>
<td>C, D(3), W</td>
<td>2</td>
</tr>
<tr>
<td>6</td>
<td>Female/39</td>
<td>White</td>
<td>5</td>
<td>C, D(2)</td>
<td>2</td>
</tr>
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<td>7</td>
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<td>West Indian</td>
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<td>D(1), W</td>
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<tr>
<td>9</td>
<td>Female/36</td>
<td>White</td>
<td></td>
<td>C, D(1), W</td>
<td>1</td>
</tr>
<tr>
<td>11</td>
<td>Female/37</td>
<td>West Indian</td>
<td>5</td>
<td>C, D(2), H, W</td>
<td>1</td>
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<td>North African</td>
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<td>C, D(2), W</td>
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<td>5</td>
<td>C, D(1), W</td>
<td>2</td>
</tr>
<tr>
<td>14</td>
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<td>West Indian</td>
<td></td>
<td>C, D(1)</td>
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<tr>
<td>15</td>
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<td>North African</td>
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<td>C, D(1)</td>
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<tr>
<td>17</td>
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<td>C, D(1), W</td>
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<tr>
<td>18</td>
<td>Female/37</td>
<td>West Indian</td>
<td>5</td>
<td>C, D(3), W</td>
<td>2</td>
</tr>
<tr>
<td><strong>Group B</strong></td>
<td></td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>1</td>
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<td>5</td>
<td>C, D(1), W</td>
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</tr>
<tr>
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<td>Female/32</td>
<td>White</td>
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<td>C, D(4), W</td>
<td>2</td>
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<tr>
<td>4</td>
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<td>C, D(5), H, W</td>
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<tr>
<td>8</td>
<td>Female/35</td>
<td>White</td>
<td></td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>Female/67</td>
<td>White</td>
<td></td>
<td>C, D(1), W</td>
<td>3</td>
</tr>
</tbody>
</table>

*C = cough; D(x) = dyspnea (Sadoul classification); H = hemoptysis; W = wheezing.

†Siltzbach classification. Stage 0 = normal chest radiograph; stage 1 = bilateral hilar lymphadenopathy; stage 2 = stage 1 plus pulmonary infiltration; stage 3 = pulmonary infiltrations; stage 4 = pulmonary fibrosis.
were reported. Bronchial biopsies were performed at various sites including stenoses, carina, and other lesions of the mucosa when present.

**PFTs**

Parameters of the flow-volume curve were measured with a spirometer (Gould Instrument Systems; Valley View, OH). Lung volumes were measured with a bodyplethysmograph (Jaeger MasterScreen Body; Jaeger, GmbH; Wurzburg, Germany). All values were expressed at body temperature, barometric pressure, and saturated with water vapor, and as percentage of predicted normal values determined by the European Respiratory Society according to sex, age, and height. An obstructive ventilatory defect was defined as a FEV1/FVC ratio < 70%. The severity of the airway obstruction was based on the FEV1 value: mild, FEV1 ≥ 70% of predicted value; moderate, FEV1 < 70% and ≥ 50% of predicted value; and severe, FEV1 < 50% of predicted value. A restrictive ventilatory defect was defined as total lung capacity (TLC) < 80% of the predicted value.

**Treatment**

Sixteen patients received corticosteroids, 1 patient received methotrexate (patient 5), and 1 patient received inhaled corticosteroids (patient 16). Group A included 13 patients: 12 patients who received corticosteroids at a daily dose of 0.5 to 0.75 mg/kg, and 1 patient who received low-dose methotrexate (10 mg/ wk). Group B included five patients: four of these patients were treated with corticosteroids a second time, after 3 months of antituberculous treatment, and the fifth patient received inhaled corticosteroids only.

**Follow-up Data**

Clinicoradiologic features, endoscopic data, and PFTs were first evaluated 3 to 6 months after the initiation of treatment and at the end of the treatment. Four items were defined: (1) complete resolution (recovery of clinical and radiographic signs, of stenosis at bronchoscopy, and of ventilatory defects), (2) partial resolution (clinicoradiologic and/or bronchoscopic improvement with an improved of the ventilatory defect, ie, FEV1 and/or FEV1/FVC ratio of at least 10% compared to the previous test), (3) stabilization (previous items remain unchanged), and (4) aggravation (persistence or worsening of the clinicoradiologic signs, bronchoscopic data, and worsening of the ventilatory defect of at least 10% compared to the previous test).

**Statistical Analysis**

Comparison of elapsed T1 and T2 between groups A and B was performed using the Mann-Whitney rank-sum test. FEV1 and FEV1/FVC ratio were compared between baseline, at 3 to 6 months, and at the end follow-up using a paired t test in each group and a one-way analysis of variance between group A and group B. Positive biopsy results at baseline were analyzed using a χ² test, and were compared between groups A and B at the end of the follow-up using a Fisher exact test. Statistical significance was assumed to be present at p ≤ 0.05. The correlation between the number of ESPBs per patient and the FEV1/FVC ratio was established by a linear regression analysis fit with the R² correlation coefficient using analysis of variance.

**Results**

**Clinical and Radiologic Features at Diagnosis of ESPB**

The diagnosis of sarcoidosis with ESPB was established in 14 patients (78%) during their evaluation for respiratory symptoms. In four patients (22%), ESPB occurred a second time. In this latter group, respiratory symptoms triggered an evaluation of ESPB at 2, 6, 12, and 14 years after the diagnosis of sarcoidosis. Airway-related symptoms were present in 17 patients (94%) and consisted of cough, dyspnea (n = 16 each; 89%), and wheezing (n = 15; 83%), as shown in Table 1.

Generalized symptoms were present in 12 patients (67%). Symptoms related to an extrapulmonary sarcoidosis were present in 13 patients (72%). Ocular manifestations consisted of xerophthalmia (n = 4; 22%) and conjunctival follicles (n = 3; 17%). Symp-
Symptoms related to the involvement of the upper respiratory tract were present in five patients, including nasal obstruction (n = 4; 22%) and chronic purulent rhinorrhea and/or epistaxis (n = 3). One patient displayed Lofgren syndrome (Table 2, Fig 1). Serum angiotensin-converting enzyme was increased in 10 patients (55%). The radiographic features at presentation are outlined in Table 3.

Flexible Bronchoscopy

Three main patterns were observed at baseline: a single stenosis (n = 3; 16.5%), multiple stenoses (n = 12; 67%), or a diffuse narrowing of the bronchial tree (n = 3; 16.5%). A single stenosis was observed, at the subdivisions of the middle lobe bronchus in two patients as a mass lesion, and at the right upper subdivision in one patient. ESPB described as diffuse was due to the intense inflammation at the lobar and segmental level.

The 12 patients in the multiple stenoses subgroup had a total of 42 ESPBs (3.5 ± 1.6 per patient; mean ± SD). The reduction of the bronchial lumen was complete (pinpoint opening) in 11 ESPBs (26%). It involved the main bronchus in 2 ESPBs...
The bronchial mucosa appeared thickened edematous and inflamed at the site of the stenosis in all cases (Fig 3). The mucosa outside the stenosis was inflamed in all patients, with fine granulations and nodules in 50% of patients. Enlargement of the carina due to extrinsic compression lymphadenopathy was reported in two patients (11%).

In all cases, at least one bronchial biopsy showed granulomas. Of the 62 bronchial biopsies performed at baseline, 48 biopsies (77.4%) showed noncaseating granulomas. Macroscopic nodules always contained granulomatous lesions (n = 5).

**PFTs**

PFTs at baseline showed an obstructive ventilatory defect without any reversibility in 12 patients (66.7%) [Table 4]. It was moderate in nine patients and severe in three patients. A restrictive ventilatory defect was associated in three patients (mixed ventilatory defect, 16.7%) and was pure in five patients (28%). PFT results were normal in one patient. The decrease of the FEV1/FVC ratio correlated with the number of ESPBs observed (Fig 4).

**Follow-up**

The mean follow-up of the patients was 56.5 ± 25 months, with a duration of corticosteroid treatment of 37.4 ± 19 months. No statistical differences were observed regarding follow-up and duration of therapy between groups A and B. Five patients are presently undergoing treatment.

Our findings suggest that T1 and T2 are important. For group A, T1 was 0 ± 2.5 months (median ± SD), whereas it was 8 ± 8 months in group B (p = 0.004). T2 was 1.0 ± 0.8 months in

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### Table 4—PFT Results at Baseline*

<table>
<thead>
<tr>
<th>Group/ Patient No.</th>
<th>FEV1, % Predicted</th>
<th>FEV1/FVC, % Predicted</th>
<th>TLC TCO % Predicted</th>
</tr>
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<tbody>
<tr>
<td>A</td>
<td></td>
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<tr>
<td>3</td>
<td>47</td>
<td>50 (58)</td>
<td>82.2 44</td>
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<tr>
<td>5</td>
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<td>6</td>
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<td>83 (95.7)</td>
<td>63 68</td>
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<tr>
<td>7</td>
<td>60</td>
<td>60 (70.7)</td>
<td>74.5 66</td>
</tr>
<tr>
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<td>81</td>
<td>61 (70.6)</td>
<td>104 84</td>
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<tr>
<td>10</td>
<td>64</td>
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<tr>
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<td>76</td>
<td>72 (82.5)</td>
<td>81 70.1</td>
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<tr>
<td>12</td>
<td>66</td>
<td>64 (76.7)</td>
<td>80 73</td>
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<td>13</td>
<td>82</td>
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<td>68 51</td>
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<td>15</td>
<td>98</td>
<td>62 (74)</td>
<td>111 102</td>
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<td>66.0 68.9</td>
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<td>81.6 70</td>
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<td>16</td>
<td>55</td>
<td>69 (83)</td>
<td>78 64</td>
</tr>
</tbody>
</table>

*TCO = transfer coefficient of carbon monoxide.

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**Figure 4.** Relationship between FEV1/FVC and the number of ESPBs at baseline.
group A and 11.5 ± 4.1 months in group B (p = 0.004). In group B, the delay was equally distributed between T1 and T2. The differences in the clinical course, radiographic results, bronchoscopic results, and PFT follow-up are outlined in Figure 5. Clinical symptoms, including dyspnea on exercise and cough, persisted in three of the five patients of group B (60%), but only mild symptoms persisted in two patients in group A (15%).

During their follow-up, the patients underwent an average of 4.6 ± 2.6 bronchoscopic examinations (mean ± SD). When the treatment was initiated immediately following bronchoscopic diagnosis (group A), ESPB resolved completely (n = 7; 87%), except in one case probably related to the delay between onset of symptoms and diagnosis (8 months), and another patient who presented bronchial distortion (Table 5). Despite improvement of bronchoscopic lesions in this group, the bronchial mucosa often remained macroscopically inflamed (n = 5; 62%). As shown in Table 5, only patients in group B had persistent stenotic lesions. Interestingly, irrespective of bronchoscopic resolution of the stenosis, bronchial biopsies performed at the end of the follow-up period still revealed noncaseating granulomas in 11 of 20 biopsies (55%) in group A and in 4 of 9 biopsies (44%) in group B.

Significant improvement of PFTs as assessed by the FEV1 and the FEV1/FVC ratio only occurred in group A (Fig 6). The improvement occurred in the first 3 to 6 months after therapy had been started. Patients in group A also had significantly improved TLC, from 4.56 ± 0.25 to 4.98 ± 0.90 L (p = 0.005) after 3 to 6 months of treatment.

Seven patients (five in group A and two in group B) had a relapse of ESPB either during tapering (n = 4) or after discontinuation of corticosteroid therapy (n = 3); 3, 16, and 25 months after discontinuation of corticosteroids. All patients improved either spontaneously (n = 2) or after restarting of corticosteroids (n = 5). The patient treated with methotrexate also had improved clinical and radiologic symptoms during treatment.

Figure 5. Global evolution of groups A and B at the end of the follow-up period (18 patients).
In patients with sarcoidosis, EPBS can present clinically with a wide range of respiratory symptoms and is associated with multisystemic involvement. Airway-related symptoms, especially wheezing, are unusual in sarcoidosis. The clinical presentation is often misdiagnosed as asthma, COPD, or pulmonary tuberculosis. This often results in a significant delay of the diagnosis and therapy in these patients. Bronchoscopy is very important. At baseline, it not only allowed the diagnosis of ESPB, but also provided valuable information concerning the location, extent of involvement, and the mechanism of the stenosis, as well as the degree of the endoluminal narrowing.

One of the most important observations of our study is that successful response to treatment of ESPB requires prompt initiation of the treatment. Early treatment is crucial to obtain complete resolution, not only from a clinical and bronchoscopic perspective, but also in terms of pulmonary function. As a matter of fact, the resolution of obstructive ventilatory defects was closely related to the early onset of the treatment. The prognostic value of an early treatment might be explained by either the rapid development of fibrotic endobronchial lesions or the fact that untreated granulomas may become more resistant to corticosteroids. Moreover, the prolonged course of therapy needed to treat these lesions supports the hypothesis that ESPB is a severe and chronic complication of sarcoidosis.

Few studies have emphasized the clinical, functional, and bronchoscopic presentations of ESPB. They included small patient numbers, mostly representing fibrotic stages of sarcoidosis and extrinsic bronchial compression. This makes it difficult to draw definitive conclusions. The reported incidence ranges from 1.33 to 8.1 and even 25.7 per 100 patient-years. The incidence in our study was much lower (0.72 per 100 patient-years). Although none of these studies provided a complete evaluation at baseline and follow-up with treatment, they did provide a broad description of this rare complication, and pointed out some typical features of ESPB in sarcoidosis.

These patients shared a typical clinical presentation, consisting of dyspnea, cough, wheezing, and extrapulmonary manifestations or generalized symptoms. They often have a poor prognosis and acquire fibrotic EPBS. The presence of generalized symptoms (ie, fatigue, weight loss, and fever) frequently leads clinicians to consider other diseases, especially tuberculosis. This emphasizes the importance of bronchoscopy with endobronchial biopsies if patients present with bronchial and general symptoms.

As indicated by Sharma and Lewis and Horak, various mechanisms could lead to narrowing of the bronchial lumen. The common localization of the strictures at the origin of the lobar and segmental bronchi suggested the possibility of direct pressure or spread of the granulomatous process from enlarged lymph nodes to the bronchi, but the lymph node capsule acts as an effective barrier to such an extension. Although bronchial compression due to enlarged lymph nodes may account for occlusion of the middle lobe, disease of other lobes, especially the right upper lobe, most likely results from endobronchial occlusion caused by sarcoid granulomas.
mas spreading in the submucosa. This is often associated with a thickened and inflamed mucosa.\textsuperscript{27} These two mechanisms are not mutually exclusive and can occur simultaneously.\textsuperscript{4} A third mechanism reported is the development of an endobronchial mass lesion, which remains anecdotal.\textsuperscript{14}

Diagnosis of ESPB requires a bronchoscopy. Axial or helical CT scans are helpful to detect focal

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure6}
\caption{Top: FEV\textsubscript{1} follow-up values (18 patients). Bottom: FEV\textsubscript{1}/FVC follow-up values (18 patients). Patients from group A (●) were compared to patients from group B (○); values are shown as mean ± SEM. *p < 0.05; **p < 0.001.}
\end{figure}
bronchial lesions, but are inaccurate in the prediction of whether a given abnormality is endobronchial, submucosal, or extrinsic. Moreover, CT scans lead to false-positive results, incorrectly predicting the presence of focal bronchial abnormalities in 7.9%–18 and 14.3% of patients.26 An obstructive ventilatory defect is commonly seen when PFTs are performed in patients with sarcoidosis.15,16,20,28 Obstructive defects have been linked to certain ethnic groups20,31 and are associated with increased mortality.28

Corticosteroid therapy remains the most commonly used treatment with proven efficacy for these lesions.14,24,27,29 Ventilatory defects have been reported to improve with corticosteroid therapy.16,27,33,34 In other studies,21,22,35 the response of airway stenosis to treatment with proven efficacy for these lesions was uncertain. As supported by our data, this may be due to the development of early cicatricial stenosis and a decrease of efficiency of the corticosteroid on chronic ESPB. In our study, most patients in whom treatment was initiated within 3 months of diagnosis displayed a significant improvement.

We are aware that our results have significant limitations. This is a retrospective study including few patients with incomplete data collection. Our study design reflects the rarity of the disease. Future prospective studies would be extremely helpful to clarify the optimal diagnosis, treatment, and the follow-up of these patients.

In conclusion, sarcoidosis complicated by proximal endobronchial stenosis is an uncommon but severe disease manifestation. Bronchoscopic diagnosis is easy, and treatment should be initiated early. Prognosis is closely related to the rapid onset of treatment since fibrotic lesions develop rapidly and appear to be resistant to corticosteroids.

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