 Bronchiectasis in Diffuse Panbronchiolitis: High Resolution CT Assessment

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Purpose: To evaluate the characteristics of the bronchiectasis in diffuse panbronchiolitis using HRCT.

Materials and Methods: We retrospectively studied 12 HRCT scans and two bronchography of 12 patients with diffuse panbronchiolitis (DPB). According to Akira et al., DPB was classified into four types: small nodules around the end of bronchovascular branchings (CT type I), small nodules in the centrilobular area connected with small branching linear opacities (CT type II), nodules accompanied by ring-shaped or small ductal opacities connected to proximal bronchovascular bundles (CT type III), large cystic opacities accompanied by dilated proximal bronchi (CT type IV). We compared the type and the extent of bronchiectasis, CT types of DPB, and pulmonary function test.

Results: Bronchiectasis was defined in 12 cases with the tubular type predominantly involving small and medium-sized bronchi. These bronchiectasis involved the proximal bronchi of the centrilobular lesions of DPB. Among eight cases of advanced DPB (CT type III & IV) which extended to both upper lobes, seven showed tubular bronchiectasis at the same area. Cystic bronchiectasis was shown in eight cases predominantly involving right middle lobe (n=7). There was no linear correlation between the values of pulmonary function test and CT types of DPB.

Conclusion: Characteristic feature of the bronchiectasis in DPB is the tubular ectasis predominantly involving the small and medium-sized bronchi. DPB with associated tubular bronchiectasis can involve whole lung field in advanced cases. HRCT is useful not only to depict the findings of DPB but also to demonstrate the extent of lesion.

Index Words: Bronchiectasis Bronchiolitis Lung, CT Computed tomography (CT), high-resolution

Diffuse panbronchiolitis (DPB) is a chronic inflammatory disease of the respiratory bronchioles of unknown etiology characterized clinically by chronic cough, expectoration, and dyspnea; physiologically by chronic airflow limitation; and histologically by typical bronchiolar lesions (1). Chest radiograph and high resolution CT have shown specific findings of the bronchiolar lesions (2, 3).

However, recent studies have shown that DPB causes inflammation and dilatation of not only the bronchioles but also the bronchi (4-7). Although there is no doubt that the major pathologic findings are present in and around the bronchiolar walls, it is not known whether the respiratory bronchioles are primarily or secondarily affected (8). Furthermore, the relationship of DPB to bronchiectasis, if there is any, remains to be elucidated (7).

We began this study to find the characteristics of
the bronchiectasis in DPB and to determine if there is any suggestion about the nature of the airway disease findings of HRCT.

Table 1. Summary of 12 Patients with Diffuse Panbronchiolitis

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/Sex</th>
<th>PFT, ABGA</th>
<th>CT Extent</th>
<th>Bronchiectasis</th>
<th>Size of Bronchi</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.57/F</td>
<td>39%</td>
<td>3/2</td>
<td>U*</td>
<td>RML</td>
<td>La*</td>
<td>17 years*</td>
</tr>
<tr>
<td>2.33/M</td>
<td>27%</td>
<td>4/3</td>
<td>U++ M++ L++</td>
<td>Whole*</td>
<td>RML</td>
<td>Bronchography</td>
</tr>
<tr>
<td>3.21/M</td>
<td>58%</td>
<td>2/1</td>
<td>U - M+ L++</td>
<td>Ls</td>
<td>La -</td>
<td>Bronchography</td>
</tr>
<tr>
<td>4.48/F</td>
<td>38%</td>
<td>4/4</td>
<td>U+ M+ L++</td>
<td>Whole</td>
<td>RML</td>
<td>Pseudomonas</td>
</tr>
<tr>
<td>5.24/F</td>
<td>61%</td>
<td>3/2</td>
<td>U+ M+ L+</td>
<td>Whole</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>6.42/M</td>
<td>58%</td>
<td>2/3</td>
<td>U - M+ L+++</td>
<td>RML</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>7.32/F</td>
<td>55%</td>
<td>3/2</td>
<td>U+ M+ L+++</td>
<td>Whole</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>8.26/M</td>
<td>48%</td>
<td>2/3</td>
<td>U - M - L+++</td>
<td>RLL</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>9.20/M</td>
<td>25%</td>
<td>4/4</td>
<td>U++ M+++ L+++</td>
<td>Whole</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>10.20/F</td>
<td>52%</td>
<td>2/3</td>
<td>U - M+ L++</td>
<td>LLL</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>11.51/F</td>
<td>42%</td>
<td>4/4</td>
<td>U+ M+ L+++</td>
<td>Whole</td>
<td>Tubular</td>
<td>La -</td>
</tr>
<tr>
<td>12.26/F</td>
<td>66%</td>
<td>3/4</td>
<td>U+ M+ L+++</td>
<td>Whole</td>
<td>Tubular</td>
<td>La -</td>
</tr>
</tbody>
</table>

Abbreviations
* U, M, L: Upper, Middle, Lower lung field
* La, Me, Sm: Large, Medium, Small-sized bronchi * Ls: Lingular segment BLL: both lower lobes * Whole: Whole lobes * Years: Duration of symptoms such as productive coughing and exertional dyspnea * NA: Not Applicable due to severe dyspnea
* Rt > Lt, Lt > Rt: Right or Left predominant involvement of lung * ++, +++, +++++: mild, moderate, severe involvement
* CT type I: small nodules around the end of bronchovascular branchings
CT type II: small nodules in the centrilobular area connected with small branching linear opacities
CT type III: nodules accompanied by ring-shaped or small ductal opacities connected to proximal bronchovascular bundles
CT type IV: large cystic opacities accompanied by dilated proximal bronchi
* CT type 3/2: Predominant CT type/Subsidiary CT type
**MATERIALS and METHODS**

The study group consisted of 12 patients (five men and seven women), who ranged in age from 20 to 57 years (mean 33 years). The diagnosis was based on the clinical, functional, and radiologic criteria of Homma et al. (1) and HRCT (Table 1). All cases had chronic pansinusitis. Pseudomonas aeruginosa was detected continuously in the sputum of one case. Histologic proof was obtained by means of open lung biopsy in one case. The CT scans were obtained on a Somatom Plus SI (Siemens) and GE 9800 (General Electric) with 1.0 to 1.5 mm collimation at 1.5 - 2.0 cm interval from the apex to the base of the lung during breath holding after full inspiration. A high spatial frequency algorithm was used for all patients. The high-resolution images were displayed at window levels appropriate for pulmonary parenchyma (1000 ~ 1500/ -700 ~ -600). In two cases, we reviewed the bronchography films taken one year before the diagnosis of DPB. Two diagnostic radiologists were participated in reading of the HRCT films. The CT findings of DPB were graded with CT types reported by Akira et al. (2). Several CT types seen concurrently in each case were categorized using slash (Predominant CT type/Subsidiary CT type, e.g. CT type III/II). The extent of the involvement of DPB was divided to upper, middle and lower third lung field. We observed the laterality of the lesions. Bronchiectasis and the thickening of the bronchial wall were assessed according to the size: large (main stem, lobar, and segmental bronchi), medium, and small (within the range of the diameter of accompanying pulmonary artery from 1 to 2 mm) bronchi by our arbitrary classification. The extent (involved lobes or segments), the type (cystic, varicose and tubular), and the laterality of involvement of bronchiectasis were also assessed. We correlated the findings of DPB and associated bronchiectasis with pulmonary function test and CT types of DPB. Wilcoxon rank sum test and Mantel-Haenszel chi-square test, and Spearman correlation coefficient were used in analysis of data.

**RESULTS**

Of the 12 cases with DPB, four cases had CT type II (CT type II/I in one, II/III in three), four had CT type III (CT type III/II in three, III/IV in one), and four had CT type IV (CT type IV/III in one, IV/IV in three). None was in the CT type I. The extent of DPB was predominant in lower lung zone, but there were upper lung zone extension in eight cases all of which had advanced disease (CT type III and IV). Bilateral symmetrical involvement was found in 10 cases (Fig. 1, 2), and there were two cases of unilateral predominant involvement (Fig. 3).

Bronchiectasis was defined in 12 cases with the tubular type predominantly involving small and medium-sized bronchi. These bronchiectasis involved the proximal bronchi of the centrilobular lesions of DPB. In two cases of unilateral predominant involvement of DPB, ipsilateral involvement of tubular bronchiectasis and thickening of bronchial wall were found (one in the left lower lobe, the other in the right lower lobe). Among the eight cases (CT type III and IV) involving both upper lung fields with DPB, seven cases showed tubular bronchiectasis. Tubular bronchiectasis was not found in the lobar or segmental area which was not involved with bronchiolar lesion.

In addition, there were cystic bronchiectasis in eight cases predominantly involving right middle lobe (RML) (n=7) (Fig. 2). Other sites of cystic bronchiectasis were left lower lobe (LLL) in one case and right upper lobe (RUL) anterior segment in another (combined RML lesion). The cases with cystic bronchiectasis had CT type II in two, CT type III in three, and CT type IV in three cases. Large bronchi were involved with tubular bronchiectasis only in one case, but all cases (n=12) showed thickening of the bronchial wall.

In two cases, previous bronchography showed mild tubular bronchiectasis at the medium-sized bronchi, broncholitis, stenosis of long segment of small airways, tapered obstruction of the small and medium-sized bronchi, and absence of the acinar filling in the involved area. (Fig. 2, 4)

There was no linear correlation between CT types and FEV1% (p = -0.526, p = 0.09), % FVC (p = -0.477, p = 0.13), and FEV1/FVC (p = -0.481, p = 0.13) though there was a tendency that the values of pulmonary function test decreased as CT type increased. With higher CT types, DPB and associated bronchiectasis involved the upper lung field (P = 0.01).

**DISCUSSION**

Clinical DPB based on the diagnostic criteria accord-
ing to Homma et al. (1) can include the case with not only pathologic DPB but also unclassified bronchiolitis and bronchiolectasis(7). But several recent studies on DPB showed that typical HRCT findings are enough to diagnose DPB when correlated with clinical feature(2, 3). Open lung biopsy is not necessary for the diagnosis of DPB when typical HRCT features, clinical findings and pulmonary function test(1) results exist.

The HRCT findings of DPB include centrilobularly distributed, small rounded areas of attenuation; branched linear areas of attenuation, contiguous with the small rounded areas; dilated airways with thick walls, also common outside secondary pulmonary lobules; and decreased lung attenuation in peripheral areas due to air trapping caused by the bronchiolar obstruction(8). Bronchial wall thickening was thought to be due to inflammation of bronchial wall or retained secretion(8). Figure 3 shows one case of crescentic wall thickening on dependent portion which can be due to retained secretion.

Cystic bronchiectasis was found in 8 cases which af-

![Fig. 2. Case 2. a. Cystic bronchiectasis in RML. There is also dilatation of medium-sized bronchi with thickened wall (arrows). b. Bronchography one year before. There are tubular bronchiectasis in right lower lobe and abrupt narrowing of small airway suggesting long segment stenosis (arrow). c. Left lower lobe shows poor filling of acini inspite of adjacent normal acinar filling.](image)

![Fig. 3. Case 10, Predominant left lung involvement of DPB with ipsilateral mild tubular bronchiectasis in medium-sized airways with thickening of bronchial wall. The thickened portion of bronchi probably suggests the retained secretion because the thickening is only seen in the dependent portion (arrow). Centrilobular lesions are seen in peripheral lung field of RML and LLL.](image)

![Fig. 4. Case 3. Bronchography shows tubular branching linear shadows about 5 mm apart from pleural surface compatible with bronchiolectasis.](image)
fect ed predominantly the right middle lobe known to be the vulnerable site due to its anatomical orientation. Considering the specific features of DPB which is bilateral, symmetrical and diffuse, we consider that cystic bronchiectasis in DPB is more likely to be due to recurrent secondary infection. We propose that the presence of cystic bronchiectasis should not influence the diagnosis of DPB in the reading of the HRCT films.

Tubular bronchiectasis involving the small and medium-sized bronchi is considered as the characteristic feature of the bronchiectasis in DPB. With the view point of the bronchiectasis, DPB can be considered as a cause of the diffuse bronchiectasis (such as cystic fibrosis and immotile cilia syndrome etc.), although the relationship of DPB and bronchiectasis was not elucidated. In our cases, there was no early DPB(CT type I) which had only centrilobular small nodules without bronchiolectasis or bronchiectasis. However, it is suggested that the bronchiectasis would eventually develop in vicinity of the bronchiolar lesions and progress proximally if the patient is neglected as shown in our cases.

It is possible but not proved that the dilatation of the proximal bronchi is secondary due to bronchiolar obstruction in DPB as proposed by Akira et al. We have shown two cases of DPB examined with bronchography which was done 1 year before under the suspicion of bronchiectasis. These cases show somewhat unusual bronchographic features for usual bronchiectasis such as irregular stenosis and dilatation of the small airways, areas of the bronchiolectasis, and difficulty of acinar filling at the involved area despite forceful inspiration. These findings were reported previously from Japan but considered as nonspecific for DPB(10, 11). With long term follow-up of the bronchographies in DPB, they found that the bronchiectasis in medium-sized bronchi is the findings of advanced cases in DPB(10) which could have been the theoretical basis insisting that the proximal bronchiectasis is secondary to bronchiolar obstruction. But two factors are required for the occurrence of the bronchiectasis: bronchiolar wall damage and a dilating force(12). It is not clear whether the proximal bronchiolar or bronchial dilatation can be caused by just physiologic dilating force produced by the distal bronchiolar narrowing or obstruction.(8)

In our study, there was no clear correlation between the CT types and the values of pulmonary function test, though there was a tendency that the values of pulmonary function test decreased with higher CT type. This is in contradiction to the report by akira et al(2) that there was a good correlation between the CT types and values of pulmonary function test. The CT types of Akira et al(2) seem to be too simplified to be in accordance with the clinical severity of DPB as there were several CT types in most patients in our study. Probably not only the CT types but also the extent of involvement of DPB should be considered in the correlation with pulmonary function test. We suggest that the CT types should be cautiously applied in clinical practice, and larger number of subject should be studied to determine clinical usefulness of the application of the CT type.

In conclusion, the characteristic bronchiectasis in DPB occurs specifically in the small and medium-sized bronchi, mainly with the tubular type. At first, the disease begins in basal lung fields. These bronchiectasis and corresponding bronchiolitis can involve whole lung field as the disease process advances. HRCT is useful to depict the centrilobular lesions and associated proximal bronchial lesions in DPB, and also useful to demonstrate the extent of DPB. To clarify the relation between the centrilobular lesions and dilated bronchi in DPB, prospective long-term studies and studies for the etiology are needed.

**REFERENCES**

미만성 세기관지염에서의 기관지확장증: 고해상 CT 소견

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목 적: 미만성 세기관지염(Diffuse panbronchiolitis, DPB)에서 잘 동반되는 기관지확장증의 특징을 알고자 하였다.

대상 및 방법: DPB로 진단된 환자 12명의 고해상 전산화 단층촬영(HRCT) 12예와 기관지조영술 2예를 후향적으로 분석하여 기관지확장증의 형태와 분포양상을 DPB의 진행정도와 폐기능검사치(FEV1, FVC)와 비교하였다. DPB의 진행정도는 Akira 등의 분류에 따라 HRCT상 type I은 기관지혈관분지부의 끝에 소결절이 있는 것, type II는 중심소엽성 소결절이 소분지음영과 연결된 것, type III는 결절이 근위부 기관지혈관대에 연결된 환형 혹은 관상형 음영과 동반될 때, type IV는 큰 난형음영이 확장된 근위부기관지와 동반될 때로 분류하였다.

결 과: 12예 모두에서 범발성의 소형 혹은 중형 기관지로 침범하는 관상형 기관지확장증이 보였다. 이들은 모두 세기관지 병소의 근위부 기관지에서 나타났고 세기관지병소가 침범한 엽과 소엽부위에서 보였다. 병변이 진행된 8예(C type III and IV) 중 7예에서는 양측상엽에서도 같은 특징의 기관지 확장증이 보였다. 8예에서는 난형 기관지확장증이 보였으며 이는 우중엽에 7예로 많았다. CT type이 진행정도에 따라 폐기능검사치가 감소하는 경향은 보였으나 유의한 상관관계는 보이지 않았다.

결 론: DPB의 특징적인 기관지 확장증은 주로 중상형 기관지의 관상형 기관지확장이다. 이 질환은 세기관지 뿐만아니라 근위부 중상형 기관지의 염증과 확장을 동반하여 폐기저부에서 병소가 시작하지만 치료없이 진행하면 전 폐와 동반된다. HRCT는 이 질환의 진단 및 진행정도 뿐만아니라 치료방향을 아는데 유용하다.