Atypical Peritracheobronchial Vasculitis and an Effective Treatment

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We report an interesting case of vasculitis in which the inflammatory lesion was limited to the peritracheobronchus. This case showed positive antineutrophil cytoplasmic antibodies, diffuse peritracheobronchial swelling, and vasculitis in its histology. Steroid therapy was effective for both roentgenological and serological findings. Although the biopsy specimen shows only inflammation and does not satisfy the WHO criteria of Wegener's granulomatosis (WG), a possible diagnosis of WG should not be disregarded.

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Key words: tracheal swelling, vasculitis, steroid therapy, c-ANCA

Introduction

We report an atypical peritracheobronchial vasculitis obtained by open chest biopsy. Stein et al reported diffuse tracheal stenosis of Wegener's granulomatosis (WG), and discussed the difficult differential diagnosis such as sarcoidosis, amyloidosis and relapsing polychondritis (1). The present case does not satisfy the WHO criteria for the localized form of WG (2). Several reports discuss difficult and delayed diagnosis of WG (3, 4). In one case, the final diagnosis was made at the time of autopsy, thus it is critical not to overlook a clinical diagnosis of WG if the biopsy specimen shows only inflammation (3). In the present case, therefore, considering WG, a close follow-up is necessary. It will also be important to observe tracheal swelling. CT scan was very useful in the detection of the main lesions where steroid therapy was very effective.

Case Report

A 56-year-old male was admitted with fever (38°C), tenderness of the parasternal area, and weight loss. There were no abnormal findings in nasopharyngeal lesion, palpable masses, hepatosplenomegaly, or lymph node enlargement. The ESR was 131 mm/h, CRP 10.4 mg/dl, and no hematuria. Antineutrophil cytoplasmic antibodies (cANCA) were positive ($\times 20$) (normal, $< \times 10$). A chest X-ray showed diffuse tracheal thickening but no abnormal shadow in both lung fields. A chest CT also showed diffuse tracheobronchial thickening (Fig. 1A). Bronchoscopic findings showed slight redness and narrowing

of the trachea. Histological examination of a transbronchial biopsy of the carina showed only a slight submucosal edema. As these findings were not sufficient to explain the diffuse tracheal thickening, we performed an open chest biopsy of the peritrachea for histological diagnosis. Peritrachea was smooth and edematous. Histological findings of peritrachea showed vasculitis with plasma cells, neutrophils, macrophages and lymphocytes without showing necrosis (Fig. 2).

As shown in Fig. 3, two days after steroid therapy with prednisolone (60 mg/day), his complaints of high fever and tenderness of the parasternal area were diminished. ESR and CRP were also improved within 10 days. Furthermore, the thickness of the tracheobronchus was decreased on chest X-ray and CT scan at one month after the therapy (Fig. 1B, Fig. 3).

Discussion

In the present patient, an atypical vasculitis of inflammatory lesion was found only in the peritracheobronchus. The endobronchial lesion showed marked improvement with prednisolone therapy. As shown in Fig. 3, the patient's complaints of high fever and tenderness of the parasternal area were diminished only two days after prednisolone therapy. The reason for this rapid improvement might be explained by the fact that the main inflammatory lesion was only around the small vascular and the small artery was intact (Fig. 2). So far as we know, this is the first case report of a limited vasculitis with diffuse peritracheobronchial inflammation.

One of the interesting findings of the present case was the

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Fig. 1. Chest CT on admission (A), after prednisolone therapy (B).

Fig. 2. Biopsy specimen of peritrachea (HE stain, $\times 100$).

positive cANCA test, although the titer was low but significant. This test has been recently recognized as a marker for WG (5) and microscopic polyarteritis vasculitis (6). As the cANCA test turned to negative after prednisolone therapy, cANCA probably participated in the pathogenesis of this disease. Furthermore, this is one of the useful tests for markers of vasculitis activity and treatment effectiveness.

An important diagnostic point of the present case was diffuse tracheal stenosis due to the swelling, which was discussed by Stein et al (1). It is necessary to differentiate from WG. Other granulomatous diseases (e.g. sarcoidosis), amyloidosis, or relapsing polychondritis were thought to be unlikely, since the patient had no other symptoms and signs including histopathological findings.

Typical bronchoscopic findings of WG are redness, swelling, bleeding, engorgement of small vessels, irregularity, ulceration, thickening, and elevated lesion (7). In contrast,

Fig. 3. Clinical course.

transbronchial biopsies of atypical WG cases have often revealed only nonspecific inflammatory tissue. In all of these cases, nasal or pulmonary biopsy enabled histological diagnosis (8). In the present case, the inflammatory lesion was only in the swelling of the tracheobronchus. Bronchoscopic findings showed slight redness and slight narrowing of trachea. Histological findings of a transbronchial biopsy of the carina showed only submucosal edema.

Although the present case differs from the WHO criteria of WG regarding biopsy findings, Scully et al showed that only irregular focal deposition of fibrin in the lamina propria with intact epithelium and without evidence of vasculitis or granuloma is compatible with and characteristic of early WG (9). Additionally, Shah et al reported an autopsy case of classical vasculitis and granuloma only in the liver and spleen (3). They stressed that, unable to make a final diagnosis until autopsy, it is critical not to negate a possibility of the clinical diagnosis of WG if the biopsy specimen of a patient with the clinical impression of WG shows only inflammation. In the present case, therefore, considering WG, a close follow-up is necessary with particular consideration to the appearance of additional symptoms or signs such as nasopharyngeal or renal lesion. Observation of tracheal swelling on CT scan was very useful to detect the main lesions, where steroid therapy was very effective.

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