RELAPSING POLYCHONDRITIS WITH AIRWAY INVOLVEMENT: A CLINICAL ANALYSIS OF 5 CASES WITH LITERATURE REVIEW

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Abstract

PURPOSE: Relapsing polychondritis (RP) is a unique, rare respiratory disorder, we did this job to enhance the investigation of Relapsing polychondritis (RP) with the involvement of the respiratory tract.

METHODS: The clinical features, auxiliary tests and prognosis of RP with airway involvement in 5 patients admitted in our hospital from 2003 were analyzed retrospectively.

RESULTS: The symptoms relapsing polychondritis (RP) in respiratory disorder included chest tight, dyspnea, breathless, cough, hoarseness. Thoracic CT scan with three dimensional reconstruction of the airways demonstrated a diffusely thickened tracheobronchial wall with tracheobronchial stenosis in earlier period of the disease and showed severe narrowing or calcification of both trachea and main bronchi in later period. Spirometry showed severe obstructive ventilatory disorder. Bronchoscopy revealed dynamic collapse of the proximal airways and dissolving of cartilagerings. In our 5 patients, after initially receiving the medical drugs such as corticosteroids and immunosuppressive drugs, symptoms were improved significantly in 2 patients, mild improved in 1 and progressed in 2 patients, in the progression cases, 1 case relieved after receiving the self- expanding metallic stent replacement in the airways(trachea and main bronchi). The following-up period was 2 to 29 month, 3 patients were still alive so far, 2 of them were died in the 1, 5 months after the diagnosis respectively.

CONCLUSION: RP with respiratory tract involvement is a rare respiratory disorder; we should diagnosis this disease with its typical clinical features, abnormal thoracic CT and bronchoscopy manifestation.

CLINICAL IMPLICATIONS: Generally, respiratory tract involvement of RP had a poor prognosis, however, early stage diagnosis and early therapy with prednisone, immunosuppressive agents could have a relative better prognosis, in the later stage, invasive methods such as tracheotomy, tracheal stent and mechanical ventilation could soothe dyspnea.
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