Chronological Assessment of Airway Lesions in Relapsing Polychondritis by Positron Emission Tomography

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Abstract

A 40-year old woman presented with pyrexia, productive cough, and bilateral precordial pain. Positron emission tomography (PET)-computed tomography (CT) showed high, diffuse F-18 deoxyglucose accumulation in the tracheal, peribronchial, and bilateral costal cartilage. We diagnosed her with relapsing polychondritis (RP) based on McAdam’s criteria. Airway lesions are a major prognostic indicator of RP, and so chronological assessment and control is essential. In this patient, PET-CT accurately reflected both the location and severity of the inflammation and helped to guide treatment decision-making and facilitated early detection of recurrence. However, its high cost is prohibitive to frequent use, making it necessary to comprehensively evaluate serum C-reactive protein levels, bronchoscopy, spirometry, and 3D-CT.

Key words: relapsing polychondritis, positron emission tomography, airway lesion

(Intern Med 54: 1099-1102, 2015)
(DOI: 10.2169/internalmedicine.54.3775)

Introduction

Relapsing polychondritis (RP) is a rare, chronic inflammatory disease involving recurrent inflammation and the destruction of cartilage, particularly of the external ears, nose, and respiratory tract. The cumulative incidence of airway lesions associated with RP is approximately 50-70% (1-3). Airway lesions are a major prognostic indicator of RP; therefore, chronological assessment and control is essential.

Case Report

In early April 2013, a woman in her forties presented with pyrexia, productive cough, and bilateral precordial pain that had gradually worsened over the preceding month. She reported having developed bilateral mixed hearing loss in 2008 and bilateral auricular pain in March 2012. On examination, the patient had an audible wheeze and a saddle nose was noted. Laboratory data revealed high serum C-reactive protein (CRP) levels but negative antinuclear antibody and anti-neutrophil cytoplasmic antibody levels. 3D-CT revealed luminal stenosis in the trachea and bilateral bronchial regions (Fig. 1a, b). Positron Emission Tomography (PET)-CT, which was performed to detect the distribution of the lesions and to determine the biopsy site, showed high and diffuse F-18 deoxyglucose (FDG) accumulation in the tracheal, peribronchial, and bilateral costal cartilages (Fig. 1c, d). Bronchoscopy revealed diffuse mucosal swelling, stenosis, and an indistinct outline of the bronchial rings in the trachea and bilateral bronchial regions. The trachea collapsed during expiration. A biopsy of the auricular and costal cartilage revealed damage to the cartilage tissue associated with inflammatory cell infiltration and a decrease in the basophilic matrix. She was also positive for anti-Type II collagen antibodies. RP was subsequently diagnosed according to McAdam’s criteria (1), namely bilateral auricular chondritis, nasal chondritis, respiratory tract chondritis, and cochlear and/or vestibular dysfunction.

On the 9th day after her first visit, we initiated prednisolone (50 mg/day) and noted a rapid improvement in both the patient’s subjective symptoms and CRP levels. 3D-CT performed 7 days later showed evidence of amelioration of tracheal stenosis and depicted the outline of peripheral bronchi that had not been detected prior to therapy. PET-CT confirmed the disappearance of the FDG accumulation one
When the dose of prednisolone was tapered to 25 mg/day in early July 2013, we noted the recurrence of both the wheezing and pain in her auricular and costal cartilage, and her CRP levels gradually elevated. Although 3D-CT did not show the progression of tracheal stenosis, PET-CT confirmed FDG accumulation in the trachea and costal cartilage (Fig. 2c). Following an increase in her prednisolone dose and the addition of cyclosporine (100 mg), the patient’s subjective symptoms improved and her CRP levels decreased. However, when we attempted to taper the dose of prednisolone, her symptoms and CRP levels worsened. Therefore, we exchanged cyclosporine for methotrexate and increased the dose to 20 mg. Subsequently, the patient’s symptoms and CRP levels improved, and a repeat PET-CT scan confirmed the disappearance of FDG accumulation (Fig. 2f). Thereafter, prednisolone was successfully tapered without recurrence.

**Discussion**

RP generally progresses slowly, with a 74% five-year survival rate and a 55% ten-year survival rate (2). Airway lesions are a major cause of morbidity and mortality. The most common cause of death is respiratory infection, followed by tracheal stenosis and airway collapse with respiratory failure (3). Although there is no specific test for RP, an assessment method is needed that can evaluate active laryngotracheal inflammation to guide treatment decision-making and facilitate early detection of recurrence.

Among laboratory data, only the serum CRP levels appear to correlate with symptoms such as costal and auricular cartilage pain; however, CRP is a nonspecific examination. Although anti-type II collagen antibodies reflect the disease state (4), the time needed to obtain test results is lengthy, making it difficult to use the results to guide treatment. Other laboratory data are generally considered to be unhelpful.

FDG PET-CT scanning showed FDG accumulation in cartilaginous structures, reflecting the infiltration of lymphocytes with a variable proportion of polymorphonuclear cells, monocytes, macrophages, and plasma cells. PET-CT is a potentially attractive option for following active airway inflammation and has been reported to be useful in small case series (5-7), but the value of this in routine clinical practice is not established. In this case, the extent of FDG accumulation in the lesion correlated with the patient’s subjective symptoms and CRP elevations (Fig. 2). PET-CT appeared to
reflect both the location and severity of inflammation and helped to guide treatment decision-making and facilitated early detection of recurrence. However, the main barrier for routine use of PET-CT is the high cost of the procedure. Another problem is radiation exposure; PET-CT exposes the patient to approximately 25 mGy for one test versus 10-20 mGy for a typical CT scan.

Spirometry and 3D-CT were useful when assessing the severity of luminal stenosis of the bronchi, but did not reflect the presence of active inflammation. We presumed that our patient had progressive tracheal stenosis but could not distinguish active disease from irreversible damage due to fibrous scarring or cartilage degradation. Bronchoscopy was also useful for assessing the severity of luminal stenosis and collapse during respiration, but it was highly invasive and thus may exacerbate mucosal swelling or cartilage inflammation via mechanical stimulation.

In conclusion, PET-CT was considered to be useful for following active airway inflammation associated with RP and it was found to be useful in the treatment decision-making process and facilitated the early detection of recurrence. However, its high cost is prohibitive and prevents its frequent use, thus making it necessary to comprehensively evaluate the serum CRP levels, while also performing bronchoscopy, spirometry, and 3D-CT.

The authors state that they have no Conflict of Interest (COI).

References

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