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Intermittent fever and cough in a 56-year-old patient: Relapsing polychondritis and extranodal NK/T-cell lymphoma

Qin Huang, Danyu Cui, Jianhui Chen, Hao Ren*, Min Yang

Department of Rheumatology and Immunology, Nanfang Hospital, Southern Medical University, Guangzhou 510515, Guangdong Province, China

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Abstract

Relapsing polychondritis (RP) is a rare autoimmune disease in which recurrent and progressive chondritis occurs throughout the body. We report a case of a 56-year-old female subject presented as intermittent fever and cough, who was found obvious luminal stenosis and intense 18F-fluorodeoxyglucose (FDG) uptake in her larynx and trachea *via* bronchoscopy and FDG positron emission tomography/computed tomography (PET/CT). The auricular cartilage biopsy demonstrated chondritis. At first she was diagnosed as RP and treated by glucocorticoid and methotrexate, leading to completely response. Fever and cough recurred after 18 months, and FDG PET/CT were performed again and targeted a newfound nasopharyngeal lesion, where the biopsy proved to be an extranodal natural killer (NK)/T-cell lymphoma, nasal type.

Keywords

relapsing polychondritis • extranodal natural killer/T-cell lymphoma • nasal type • 18F-fluorodeoxyglucose positron emission tomography/computed tomography

Case Presentation

A 56-year-old female patient was admitted with the complaint of intermittent fever (about 38.5 ℃), dry cough for 3 months. She denied chills, notable sweats, sore throat, stridor, dyspnea, or weight loss. She received empiric antibiotics for suspected infections with no improvement. Tests showed increased C-reactive protein (CRP), and no positive results of infection, autoantibodies or tumor biomarkers. Her bronchoscopy examination (Figure 1A-1D) revealed obvious luminal stenosis of glottis and upper segment of trachea, with the loss of trachea cricoid cartilage, and she had no abnormalities of the ears, nose, eyes, or joints. Then the FDG PET/CT was performed and showed intense FDG uptake in her larynx and tracheobronchial tree, which led to the consideration of relapsing polychondritis (RP) (Figure 1E). The diagnosis of RP was confirmed by an auricular cartilage biopsy demonstrating inflammatory cell infiltration in cartilaginous tissue (Figure 1F). Prednisone 50 mg daily and methotrexate 10 mg weekly were prescribed, and her body temperature returned to normal and the dry cough gradually resolved. CRP began to subside and the patient was discharged.

However, while on a maintenance dose of 10 mg prednisone daily and methotrexate 10 mg weekly, her fever and cough recurred after 18 months, leading to readmission. She was suspected of active RP and received Methylprednisolone 40 mg daily, but the fever and cough could be relieved shortly and recurred. We performed FDG PET/CT to probe the cause of intermittent fever and cough, which showed FDG accumulation in the top of nasopharynx while the previous intense FDG uptake in larynx and tracheobronchial tree had gone back to normal (Figure 2A). A biopsy of the nasopharynx was performed and upon examination proved to be an extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKTCL-NT) (Figure 2B-2E). The patient was transferred to the hematology department for further treatment.

Discussion

RP is a rare autoimmune disease in which recurrent and intense chondritis occurs throughout the body, particularly in the ears, nose, respiratory tract, joints, and cardiovascular system. Patients with RP often present with other autoimmune

Address for correspondence:

*Hao Ren, Department of Rheumatology and Immunology, Nanfang Hospital, Southern Medical University, 1838# North Guangzhou Avenue, Guangzhou 510515, Guangdong Province, China. E-mail: renhao67@aliyun.com

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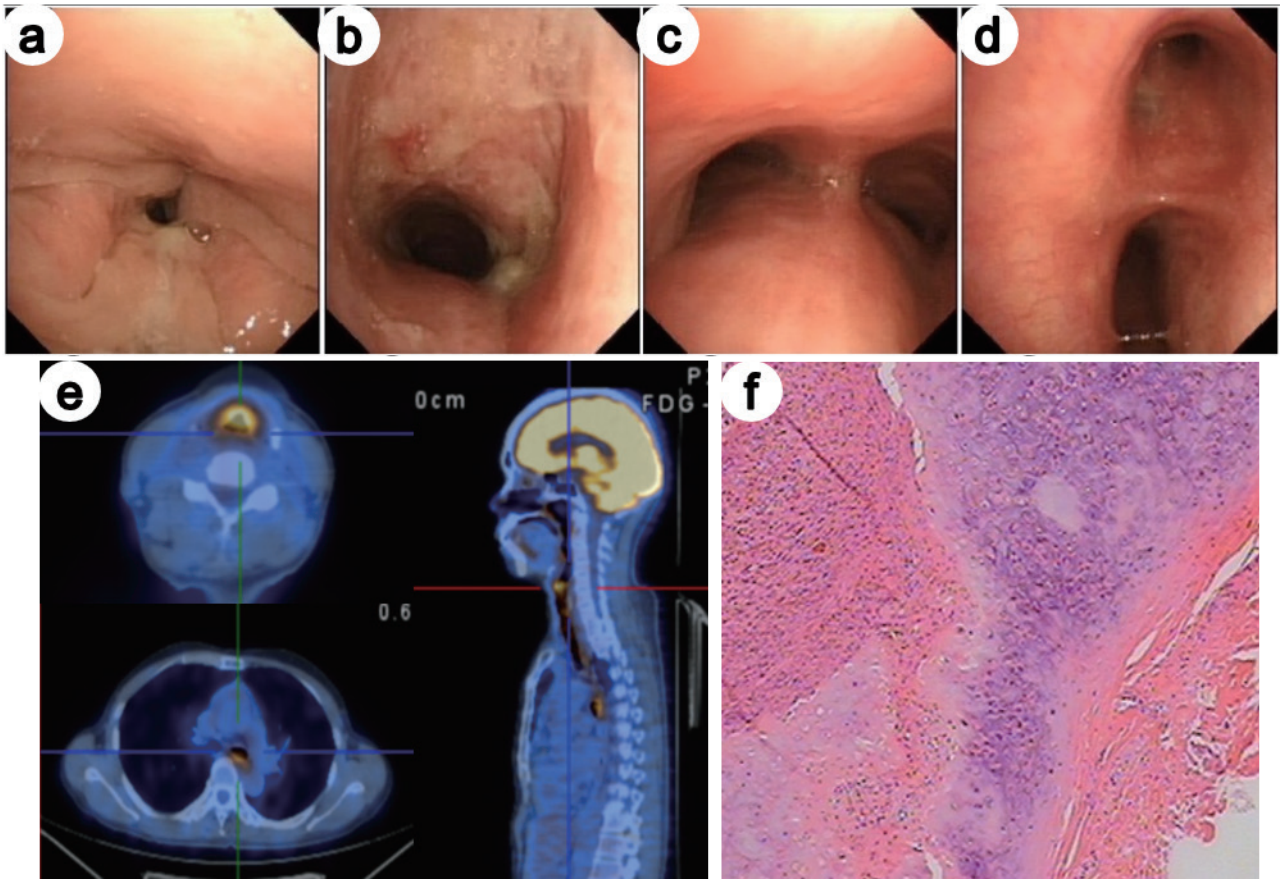

Figure 1: (a-d) The bronchoscopy images of glottis, upper segment of trachea, left principal bronchus, right principal bronchus, with luminal stenosis and loss of trachea cricoid cartilage before start of treatment. (e) FDG PET/CT findings of the larynx and tracheobronchial tree before the start of treatment. (f) Pathological finding for the auricular cartilage showed cartilaginous infiltration of inflammatory cells. H&E staining. Original magnification: X 100. FDG PET/CT, 18F-fluorodeoxyglucose positron emission tomography/computed tomography; H&E, hematoxylin and eosin.

diseases but association with malignancy is rare except with myelodysplastic syndrome.[1] ENKTCL-NT is a rare subtype of non-Hodgkin's lymphoma (NHL) characterized by an aggressive clinical manifestation involving the nose and upper aerodigestive tract (80%).[2] In this case, the patient was initially diagnosed with RP according to Damiani and Levine criterion and had good response to glucocorticoid treatment.[3] However, the fever and cough recurred after 18 months of initial treatment. FDG PET/CT were performed again and targeted the newfound nasopharyngeal lesion, and ENKTCL-NT was finally diagnosed.

We reported an infrequent case of RP associated with ENKTCL-NT, illustrating a rare combination of RP and lymphoma. Only 5 case reports suggesting an association between RP and lymphoma have been reported to date,[4-8] including: Hodgkin lymphoma, orbital mucosa-associated lymphoid tissue type lymphoma, nodal NHL and splenic NHL. It has been a controversial issue what was the real association of RP and lymphoma? Do they overlap or mimic each other? In these reported cases, the time interval between diagnosis of RP and the specific lymphoma differed. There were 2 cases of RP preceding lymphoma, and the time interval from RP to lymphoma were 6 months and 12 years respectively.[5,8] RP occurred after lymphoma with the interval of 3 years and 21 years in 2 cases.[4,7] RP and lymphoma occurred simultaneously in 1 case.[6] In our case study, the patient developed ENKTCL-NT about 18 months after the initial diagnosis of RP. Other studies have reported 2 cases of lymphoma mimicking RP, considering that some RP cases may occur as a paraneoplastic condition of the hematological malignancies.[9-10] In our case, we believe that ENKTCL-NT was overlapped with RP, rather than mimicking RP, because there was little FDG uptake in her nasopharynx at the time of diagnosing RP.

Therefore, physicians treating RP must be vigilant in searching for underlying malignancies, particularly with unexplained fever during the follow-up. Whether it is mimicking or combination, lymphoma should be considered an important differential diagnosis for patients with RP.

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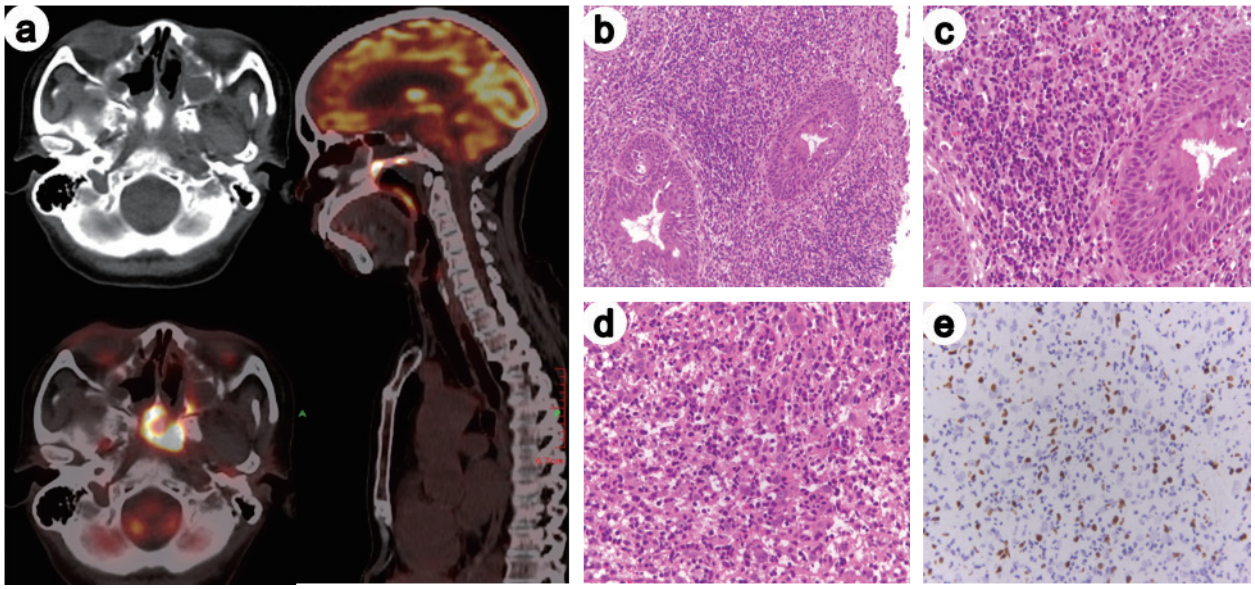

Figure 2: (a) FDG PET/CT findings showed that FDG accumulation in the top of nasopharynx and few FDG uptake in larynx and tracheobronchial tree after 18 months of initial treatment. (b-e) Pathological findings of the nasopharynx showed atypical lymphocytes varied in size, with CD2 (+), CD3 (+), CD7 (+), CD56 (+), TiA-1 (+), GZB (+), Ki-67 (+, 60%) in immunohistochemistry. EBER was positive in situ hybridization. H&E staining and situ hybridization. Original magnification: X 40, X 100, X 400, X 400 respectively. FDG PET/CT, 18F-fluorodeoxyglucose positron emission tomography/computed tomography; EBER, Epstein-Barr virus-encoded small RNA; H&E, hematoxylin and eosin.

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Author contributions

Writing- original draft preparation, Huang Q and Cui D. Writing-review and editing, Chen J and Ren H. Supervision, Yang M. All authors have read and agreed to the published version of the manuscript.

Informed Consent

The authors certify that they have obtained all appropriate patient consent documents. In the documents, the patient has given her consent for her images and other clinical information to be published in the journal.

Ethical Statement

Not applicable.

Conflict of Interest

The authors declare no conflict of interest.

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