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Abstract

We present the case of a 39-year-old man with a severe glottis stenosis. The saddle nose, images of laryngotracheal stenosis, and the (FDG) positron-emission tomography/computed tomography (PET/CT) lead to a final diagnosis of relapsing polychondritis. In the patient a coexistent myelodysplastic syndrome was diagnosed. Moreover, the elevated total IgG4 exceeding 135 mL/dL requested additional immunochemistry for detection of IgG4 bearing plasma cells in the biopsies. The patient underwent an allogenic stem cell transplantation and died on the day 40 after the transplantation because of an acute steroid-resistent graft-vs-host.

Case report. A 39-year-old man with a history of asthma presented with inspiratory stridor, and parasternal and costochondral pain triggered by deep inspiration. Physical examination showed a saddle nose (Figure 1c), without inflammation or deformity of the auricle (Figure 1a). Laryngoscopy disclosed a significant swelling of the vocal cords with a circular subglottic stenosis and a severe laryngotracheal stenosis (LTS) (Figure 1b). A tracheostomy had to be performed (Figure 2a). ESR was 118 mm/h (n.v. ≤12), CRP 8.2 mg/dl (n.v. < 0.3), and hemoglobin 10.2 g/dl (n.v. 13–17). ANA, ANCA, anti-CCP and rheumatoid factor were negative. The serum concentration of IgG4 was 219 mg/dl (n.v. < 135). Serology for syphilis, HBV, HCV, EBV and CMV was negative. TBC test was negative. The CT of the thorax and bronchoscopy were normal.

The FDG-PET/CT (Figure 2) revealed an increased FDG uptake in the posterior aspect of the of the larynx, the rib cartilages and the costochondral joints. These findings supported the diagnosis of Relapsing Polychondritis (RP). There was no indication of underlying neoplasia or systemic vasculitis. Cardiac, renal and ocular involvement was ruled out. An associated Myelodysplastic Syndrome classifiable as refractory cytopenia with multi lineage dysplasia (RCMD) was diagnosed. Light microscopy of the biopsy from the vocal cords and trachea showed a stromal pleomorphic lymphocytic infiltrate; storiform fibrosis and obliterative phlebitis were absent. No more than 15–20/HPF IgG4 positive plasma cells were found.

The patient was treated with prednisolone 1 mg/kg and azathioprine 2 mg/kg with an initial prompt clinical improvement. However during follow-up glucorticoids needed to be soon taperend because of development of a aseptic necrosis of femoral head. Because of the worsening of the respiratory symptoms together with development of anemia requiring blood transfusion more than one time patient undervent an allogenic stem cell transplantation. Patient died on day 40 after transplantation after developing an acute steroid-resistent graft-vs-host.

Discussion. RP is a rare disease characterized by progressive episodic inflammation of the cartilaginous structures of the ear, nose, peripheral and axial joints, and tracheobronchial tract.[1] Eyes, heart, blood vessels and kidneys, may also be involved.

In absence of specific laboratory tests, the diagnosis of RP is made according to definite clinical criteria.[1,2]

Typically, auricular chondritis occurs in almost 90% of RP patients. However, as demonstrated by our patient, its absence does not rule out the diagnosis.

In this case the differential diagnosis should include granulomatosis with polyangitis (GPA, formerly defined as Wegener's Granulomatosis), sarcoidosis, rheumatoid arthritis, amyloidosis and TBC.

Involvement of the respiratory tract in RP is not uncommon. However, in patients with predominant laryngotracheobronchial involvement, the diagnosis can be easily overlooked, leading to a organ threatening situation. Furthermore, in hindsight it is reasonable to conclude that the initial symptoms reported by the patient led to the earlier misdiagnosis of RP as bronchial asthma.

ESR and CRP are normally elevated during disease flares. ANA, ANCA, RF, anti-CCP and complement are useful for the diagnosis of associated diseases (e.g. SLE) or to differentiate between diseases with similiar clinical features (e.g. GPA).

FDG-PET/CT is the preferred method to identify inflammation caused by RP, especially in the respiratory tract.[3] Approximately one third of patients with RP suffer from associated diseases such as myelodisplasia (for review [2,4]).

As recently reported, the destruction of cartilage observed in RP may be associated with infiltration of plasmacytes expressing IgG4.[5] Histopathological confirmation of storiform fibrosis and infiltration of > 30/HPF IgG4+ plasma cells is required. This case can be conidered not a definitive, but a possible IgG4-related disease.

Treatment of RP is empirical. Organ threatening situations, such as laryngotracheal tract involvement with LTS, usually requires high dose corticosteroids combined with steroid sparing agent. The role of biologic agents is difficult to assess.

This case shows that isolated involvement of the respiratory tract may lead to a delay in diagnosis. Early diagnosis and aggressive treatment are vital in case of organ threatment symptoms. Finally, RP may coexist with other diaseases.

Authors contributions

All the authors contributed equally to the manuscript.

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Figure 1. (**A**) The patient's tracheostomy. (**B**) Fiberoptic laryngoscopy procedure revealed a marked edema of the vocal cords (long arrows) and a circular subglottic stenosis grade III according to the Myer–Cotton classification score (short arrow). (**C**) The typical deformity known as saddle nose.

Figure 2. (**A** and **B**) A FDG-PET/CT showed markedly increased FDG uptake bilaterally in the posterior aspect of the of the larynx (white arrows), (**C**) costochondral joints (black arrows), and (**D**) rib cartilages (white arrows). The cardiac muscle's physiologic uptake of the tracer was filtered out in order to avoid its projection on costhocondral uptake of the tracer.