ORIGINAL RESEARCH PAPER

Pulmonary Medicine

RELAPSING POLYCHONDRITIS CAUSING AIRWAY OBSTRUCTION: A RARE ENTITY

KEY WORDS: polychondritis; bronchoscopy; PET; steroid.

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ABSTRACT

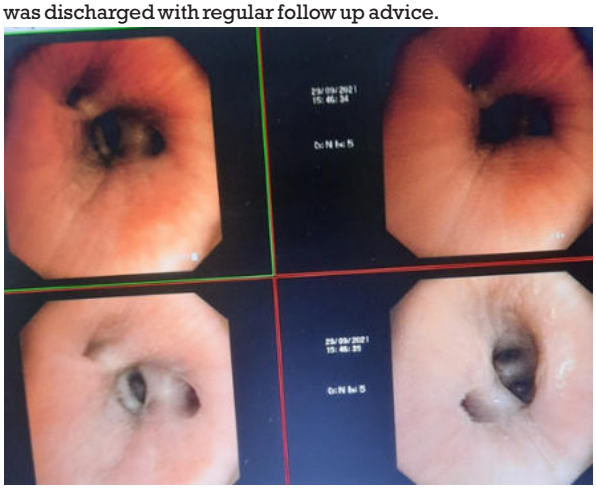
Relapsing polychondritis is one of the uncommon autoimmune chronic multisystem disorders characterized by recurrent episodes of cartilaginous tissue inflammation. We hereby present a case of 60 years old lady presented with fever, respiratory symptoms and multiple joints pain since 3 months. She had saddle nose, hoarseness of voice, rhochi and crepitation in chest. All routine investigations were done. Connective tissue panel markers were negative. Bronchoscopy showed airway edema and lavage microbiology was sterile. In whole body PET scan, FDG avid edema, subtle soft tissue thickening in cartilaginous portion of nasal septum, laryngeal cartilages, tracheobronchial tree, bilateral costochondritis and peripheral arthritis in bilateral upper limbs were found. Diagnosis of relapsing polychondritis was made based on Clinico-radiological finding and improvement with steroid and immuno suppressents (Modified Adam's criteria). She was managed with intravenous pulse steroid, immunosuppresent, antibiotics and other supportive cares. Patient improved clinically and was discharged with regular follow up advice. Diagnosis of relapsing polychondritis is challenging in reality and cause significant morbidities during diagnosis. It may be considered in differential diagnosis of airway obstruction as a rare entity.

INTRODUCTION

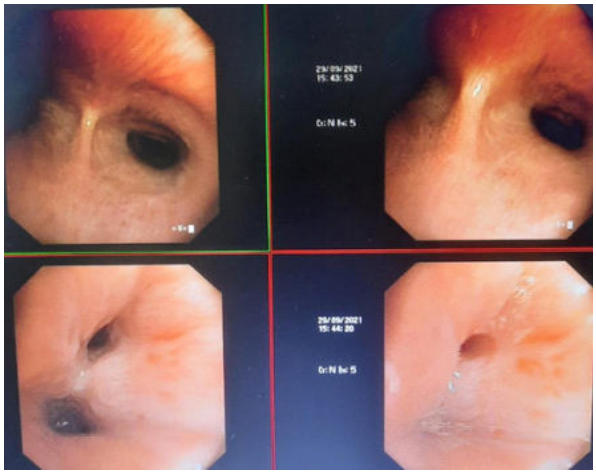
Relapsing polychondritis (RP) is a very rare autoimmune disorder. It is characterized by acute inflammation of cartilages that may recover spontaneously, but relapses frequently and attacks on cartilaginous tissue. Exact etiology is not known. IgG type antibodies against type 2 collagen are found to be the culprit in nearly half of all affected Patients[1]. Initially this disorder was known as "polychondropathy" by Jacksh-Wartenhorst in 1923. 'Chondromalacia or chronic atrophic polychondritis' was renamed it later. In 1960, Pearson et al. described a series of 12 cases presented with characteristics clinical features and used the term "Relapsing polychondritis"[2]. Estimated incidence is 3.5 cases in every 1 million population[3]. The prevalence is equal in men and women. It has been described in all races, but is more frequently diagnosed among Caucasians[4]. Age predominance is seen in 4th and 5th decades of life. Relapsing polychondritis is associated with recurrent inflammatory insults to cartilaginous and proteoglycan-rich structures. Most often, ear inflammation (involving the auricle with sparing the lobe), nasal chondritis, and arthralgia are manifested. One-third of patients are coexisting with autoimmune disease. The degree of systemic involvement is variable from nothing to notable, affecting the cardiovascular and respiratory systems and leading to life-threatening complications[5]. We hereby reported a rare case of relapsing polychondritis to alert the practicing physicians to keep it as differential diagnosis of airway obstruction.

CASE REPORT

60 years old lady presented to us with complaint of low grade, intermittent fever since 3 months. She also had cough with occasional sputum production, breathlessness on exertion and pain in the multiple joints (both hands and feet) on and off for same duration. On examination, she was conscious, oriented, afebrile with stable vitals. SpO2 was 93% on room air. She had saddle nose and hoarseness of voice. On chest examination, bilateral crepitations and rhonchi were present. On routine investigations, complete blood count showed white blood cell count of 10,260/mm^3 (N-70%), CRP of 141mg/lt, ESR of 105mm/hr, Procalcitonin of 0.1ng/ml, negative viral markers, normal RA factor, negative HLAB27, negative Anti-CCP, negative connective tissue panel markers, normal TSH, HbA1c of 8.2%, raised blood calcium with normal PTH, normal IgE and ACE level. Blood and urine culture were sterile. Mantoux test was negative. ECG had sinus rhythm. 2D echocardiography report showed LVEF of 60%, no valvular disease. X-ray pelvis was normal. ABPA panel was negative. Bronchoscopy was done and found airway edema (Figure: 1&2). Broncho-alveolar lavage stains and culture report were inconclusive. Whole body PET scan showed FDG avid edema, subtle soft tissue thickening in cartilaginous portion of nasal septum, laryngeal cartilages, tracheobronchial tree, bilateral costochondritis and peripheral arthritis in bilateral upper limbs. Diagnosis of relapsing polychondritis was made based on Clinico-radiological finding and improvement with steroid and immunosuppressents (Modified Adam's criteria). She was managed with intravenous pulse steroid, immuno suppresent, antibiotics, bronchodilator, oral hypoglycemic therapy and other supportive cares. Following treatment, patient improved clinically and CRP, ESR value reduced and was discharged with regular follow up advice.



[Figure: 1 showing airway edema on bronchoscopy]



[Figure: 2 showing airway edema on bronchoscopy]

DISCUSSION

Relapsing polychondritis is classically an autoimmune disease with a variable presentation, making a diagnostic challenge. One-third patients have coexisting autoimmune disease. Marcela Ferrada[6] et al. classified this disease into 3 subgroups. Type 1 group had ear chondritis (100%), tracheomalacia (100%), saddle-nose deformity (90%), and subglottic stenosis (80%) and had the shortest time to diagnosis (medium duration of 1 year), highest disease activity, and greater number of admission to the intensive care unit and tracheostomy. Type 2 group had tracheomalacia (100%) and bronchomalacia (52%), but no saddle-nose deformity or subglottic stenosis and needed the longest median time to diagnosis (10 years) and had highest percentage of work disability. Type 3 group was characterized by tenosynovitis/synovitis (60%) and ear chondritis (55%). In our study, we found mainly nasal and airway involvement without involving external ear. Histological diagnosis is not so easy. Because cartilage material is difficult to be obtained for analysis, or if obtained, findings are not so characteristic, principally in the advanced stages of disease when there is destruction of the structures and replacement by fibrous tissue. When there is no possibility of a histological diagnosis, diagnosis must be made clinically. Generally, the diagnostic criteria described by McAdam[7] et al. are:

1- Relapsing chondritis in both auricle;
2- Non-erosive inflammatory arthritis;
3- Chondritis in nasal cartilage;
4- Eye inflammation including conjunctivitis, keratitis, scleritis, episcleritis and/or uveitis;
5- Chondritis in the upper respiratory tract involving laryngeal and/or tracheal cartilage;
6- Cochlear or vestibular dysfunction.

The presence of three or more of the above mentioned conditions is considered as diagnostic for relapsing polychondritis. However, Damiani and Levine[8], and Michet[9] et al. made a modification of the McAdam criteria in 1979 and 1986 respectively. As per these modified criteria, diagnosis is considered when there are:

1- Presence of at least 3 or more McAdam criteria;
2- Presence of at least one or more McAdam criteria along with positive histopathological findings;
3- Chondritis of minimum 2 different locations with positive response to steroid or dapsone treatment.

Our patient is easily fit with the modified Adam's criteria number 3 for the essential diagnosis. Echocardiography or cardiac MRI is recommended for the demonstration of cardiac valvulopathy or aortic aneurism. In our patient, the valvupathy was ruled out. The most common computed tomography finding is thickening of the airway walls and fixed obstruction similar to our case. Moreover, subglottic stenosis and narrowing of tracheobronchial luminal, thickening of tracheal cartilage accompanied by dense calcification, peripheral bronchial narrowing, and bronchiectasis may also be the other findings. PET CT is useful for demonstrating multi-system cartilage anomalies along with the diagnosis of relapsing polychondritis[10]. Bronchoscopy visually observes the edema, thickening of bronchial wall, and disappeared cartilage ring. However, tracheobronchial lumina narrowing may also be found in infection, amyloidosis, tuberculosis etc. Tc-99m methylene diphosphonate (MDP) bone scintigraphy has also been used to assess in some case reports. In relapsing polychondritis, there is associated with a 30% mortality rate and prognosis of patient is variable. The most common cause of death is due to pneumonia secondary to airway anomalies and steroid use. Next common cause is airway collapse and cardiovascular complications. Early detection of respiratory system involvement considerably reduces mortality. There is no standard guideline of treatment. But disease management is involved with the therapeutic intervention by immunosuppressants like corticosteroids and dapsone. Additional medications used along with steroids include antirheumatic drugs like azathioprine, anakinra, and abatacept. Other drugs include cyclophosphamide, rituximab, tocilizumab, TNF-□ inhibitors and colchicine. In our case, combination of steroid and cyclophosphamide treatment worked quite well. Further research to evaluate the prognosis, mechanisms, and treatment may be necessary in the future.

CONCLUSION

Early diagnosis of relapsing polychondritis and proper treatment are essential to prevent disease related complications. FDG avid PET scan has a growing role in diagnosis and follow-up of this disease. Missed diagnosis may lead to disease progression and decrease survival.

REFERENCES

1. Edrees A. Relapsing polychondritis: a description of a case and review article. Rheumatol Int. 2011;31:707-13.
2. Pearson CM, Kline HM, Newcomer VD. Relapsing polychondritis. N Engl J Med. 1960;263:51-8.
3. Kent PD, Michet CJ Jr, Luthra HS. Relapsing polychondritis. Curr Opin Rheumatol. 2004;16:56-61.
4. Michet CJ. Relapsing Polychondritis of Koopman WJ. Arthritis and Allied Conditions. A Textbook of Rheumatology, 13th ed. 1997;82:1595-1603.
5. Vitale A, Sota J, Rigante D, et al. Relapsing polychondritis: an update on pathogenesis, clinical features, diagnostic tools, and therapeutic perspectives. Curr Rheumatol Rep. 2016;18:3.
6. Marcela Ferrada , Casey A Rimland , Kaitlin Quinn et al. Defining Clinical Subgroups in Relapsing Polychondritis: A Prospective Observational Cohort Study. Arthritis Rheumatol. 2020;72:1396–402.
7. McAdam L.P., A O'Hanlan M., Bluestone R., Pearson C.M. Relapsing polychondritis: Prospective study of 23 patients and a review of the literature. Medicine. 1976;55:193–215.
8. Damiani JM, Levine HL. Relapsing polychondritis-report of ten cases. Laryngoscope 1979;89:929-46.
9. Michet CJ Jr, McKenna CH, Luthra HS, O'Fallon WM. Relapsing polychondritis. Survival and predictive role of early disease manifestations. Ann Intern Med 1986;104:74-8.
10. Deng H, Chen P, Wang L, Li X, Yi J. Relapsing polychondritis on PET/CT. Clin Nucl Med 2012;37:712-5.