

CASE REPORT

Relapsing Polychondritis Possibly Caused by Chronic Infection with *Borrelia burgdorferi* – Case Report

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Introduction: Relapsing polychondritis has been described by Rudolph von Jaksch Warthenhorst in 1923 under the name of polichondropathy. Although the etiology is unknown, an autoimmune mechanism is likely due to formation of autoantibodies to type II collagen in the extracellular matrix of the cartilage with its consequent destruction.

Case report: We present a case with relapsing polychondritis according to McAdam and Damiani criteria and with positive Western blot reaction for Ac IgG anti *Borrelia burgdorferi*. Serological reaction became negative and the patient presented favorable evolution at 6 months follow-up after corticosteroid therapy and antibiotics.

Conclusion: Chronic infection with *Borrelia burgdorferi* may play a role in relapsing polychondritis determinism

Keywords: relapsing polychondritis, *Borrelia burgdorferi*

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Introduction

Relapsing polychondritis (RP) has been described by Rudolph von Jaksch Warthenhorst in 1923 under the name of polichondropathy. Relapsing term was introduced in 1960 by Pearson, Kline and Newcomer which highlights developments in inflammatory relapses followed by destruction of cartilage, which is replaced by fibrous tissue [1]. Clinical signs affects organs rich in cartilage with consequent damage such as ears, nose, trachea, bronchi, heart valves, joints, larynx, skin [2, 3]. Rarely kidneys, eyes, blood vessels and the heart [4, 5, 6].

Clinical manifestation in organs involved may be preceded by recurrent fever without an obvious cause [7]. Although the etiology is unknown, an autoimmune mechanism is due to formation of autoantibodies to type II collagen in the extracellular matrix of the cartilage with its consequent destruction [8].

Case presentation

We present a male patient, Caucasian, 50 years, which came at the Dermatology Clinic with swelling and erythema of the upper two thirds of the ears bilaterally symmetrical, spontaneous sensitive and at palpation (fig 1).

Symptoms associated with erythema of sclera (fig. 2). Both ocular lesions and those of the ears started simultaneously about three weeks before, being more pronounced

on the left side. Skin lesions were preceded by joint disruption in the form of erythema, edema, stiffness and pain in joints interphalangeal and metacarpal phalangeal in left hand. From previous medical history we noted a tick bite in several years ago, when the patient presented centrifugal erythema with spontaneously remission without determining anti-borrelia antibodies and no specific treatment.

Western blot test for *Borrelia* was positive for IgG and negative for IgM. Based on the McAdam and Damiani criteria we considered relapsing polychondritis.

After anti-infective therapy with doxycycline 200 mg/day for 21 days associated with systemic corticosteroids (prednisone 25 mg/day) and local (mometasone furoate 1 application/day) the lesions were remitted (fig. 3) and serology become negative at 6 months after treatment.

Discussions

Isolated reports, clinical features and treatment of this disease were reported between 1923 and 1975 in first systematic review on a sample of 159 patients performed in 1976 by McAdam et al. They found that disease is affecting both sexes equally, in the fourth decade is the highest incidence, approximately 30% of patients have an autoimmune or rheumatologic disease which precedes or coexist with relapsing polychondritis [2]. Affection of auricular cartilage was found in 88.5% of patients, with audiometric in 80% of those affected by RP [9, 10]. Death occurs in 30% of cases, the underlying cause being respiratory failure by affecting airway cartilage [2].

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Fig. 1. Bilateral chondritis



Fig. 2. Erythema of sclera



Fig. 3. Remission after treatment

Diagnosis is set on the basis of the clinical picture in most cases. Radiographs and joint histopathology help support the diagnosis. In atypical forms with primary respiratory impairment, it appeals to PET-CT, followed by cartilage biopsy of involved organ [11]. Histopathology shows degeneration and an inflammatory infiltrate composed of lymphocytes, plasma cells and neutrophils. The first diagnostic criteria was established by McAdam and imply the presence of at least three of the following clinical

manifestations: bilateral auricular chondritis, nasal chondritis, audio-vestibular affection, sero-negative arthritis, ocular inflammation and airway chondritis [2]. In 1979 Damiani sets new diagnostic criteria, namely the presence of one of the events described by McAdam with relevant histopathology or two clinical symptoms with favorable response to corticosteroid therapy [12]. In our case the diagnosis was supported by this bilateral ear chondritis with sclera inflammation, joint damage and favorable response to prednisone.

There have been cases cited of RP association with other autoimmune or inflammatory disease such as ulcerative colitis, ankylosing spondylitis, Behcet's disease, myelodysplastic syndromes [13, 14, 15, 16]. There are reported cases of central nervous system affection translated by encephalitis, meningitis, meningoencephalitis [17, 18]. Have been described so far three cases of RP with symptoms of Parkinson's disease, but the association was demonstrated by Atsushi and his team in 2014 by highlighting the Lewy bodies in a patient who present clinical resemble of Parkinson's disease and RP [19].

We introduced prednisone in dose of 25 mg/day, followed by gradual decrease when therapeutic response. This represents the treatment of choice, 75% of patients requiring a mean dose of 25 mg/day for the relief of symptoms [2]. It generally calls to prednisone 1 mg/kg/day with gradual decrease after getting control of clinical manifestations. Colchicine may also be used, cyclosporin A or methotrexate [20]. There are reports of biologic therapy benefits for etanercept or tocilizumab [21, 22]. We associated with doxycycline 2x100 mg /day for 21 days due to the presence of IgG anti *Borrelia burgdorferi*.

Infection with *Borrelia burgdorferi* is cited in literature coexisting with other autoimmune diseases such as morphea plaque and other localized scleroderma or dermatomyositis [23, 24]. It was reported the association between dermatomyositis and RP due to *Borrelia burgdorferi* infection [25]. In order to make a direct causal link between infection with *Borrelia burgdorferi* and some organic disease it is need to be showed the presence of spirochetes in affected tissue by molecular methods such as PCR or FFM [26]. In the case of our patient we could not perform molecular biology, but the history of the tick bite, a chronic migrant erythema, no antibiotic therapy at that time, with positive serology converting in negative subsequent to favorable evolution after antibiotic therapy lead us to claim that chronic infection with *Borrelia burgdorferi* may play a role in the development of relapsing polychondritis.

Conclusion

Relapsing polychondritis has an autoimmune etiology with multiple organs affected and the main target is collagen type II. Diagnosis can be sustain clinically in typical forms but etiology of disease it is still far from being fully clarified. Chronic infection with *Borrelia burgdorferi* may play some role in the determinism of the disease.

Although *Borrelia* IgG test can be false positive in autoimmune diseases, the presence of tick bite and chronic migratory erythema associated with remission symptoms, negative serology at 6 months after therapy, allows us to formulate the hypothesis that chronic infection with *Borrelia burgdorferi* could be involved in the etiopathogenesis of relapsing polychondritis.

References

1. Rhys Davies H, Kelsall AR. Atrophic Polychondritis: With the Report of a Case. *Ann Rheum Dis.* 1961;20: 189-193.
2. McAdam LP, O'Hanlan MA, Bluestone R, et al. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. *Medicine (Baltimore).* 1976;55:193-215.
3. Cantarini L, Vitale A, Brizi MG, et al. Diagnosis and classification of relapsing polychondritis. *J Autoimmun.* 2014;48-49:53-59.
4. Chang-Miller A, Okamura M, Torres VE, et al. Renal involvement in relapsing polychondritis. *Medicine (Baltimore).* 1987;66(3):202-217.
5. Ben Salah R, Frikha F, Hentati Y et al: Recurrent anterior uveitis with hypopyon revealing relapsing polychondritis. *Intern Med.* 2012;51(22):3173-3176.
6. Gautier M, Boutemy J, Ivascau C, et al. Cardiac involvement in relapsing polychondritis. *Eur Heart J.* 2014 Sep 1;35(33):2241.
7. Avila JN, Carvalho SB, Tavares G et al. Fever of unknown origin in a patient with red ears: relapsing polychondritis. *BMJ Case Rep.* 2014 Feb 7;2014. pii: bcr2013202670.
8. Puéchal X, Terrier B, Mouthon L, et al. Relapsing polychondritis. *Joint Bone Spine.* 2014;81(2):118-124.
9. Yi H, Dong M. Clinical analysis of 26 patients with relapsing polychondritis. *Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.* 2014;28(7):446-449.
10. Gusmão RJ, Fernandes FL, Guimarães AC, et al. Otorhinolaryngological findings in a group of patients with rheumatic diseases. *Rev Bras Reumatol.* 2014;54(3):172-178.
11. Wei, L, Da-Xiong Z, Tao C, et al. FDG PET-CT combined with TBNA for the diagnosis of atypical relapsing polychondritis: report of 2 cases and a literature review. *J Thorac Dis.* 2014; 6(9):1285-1292.
12. Damiani JM, Levine HL. Relapsing polychondritis. Report of ten cases. *Laryngoscope.* 1979;89(6):929-946.
13. Paras, K, Ranjan P, Pragya S, et al. The red hearing: swollen ear in a patient with ulcerative colitis. *J Community Hosp Intern Med Perspect.* 2014; 4(3): 10.3402/jchimp.v4.24659.
14. Azevedo VF, Galli NB, Kleinfelder AD, et al. Relapsing polychondritis in a patient with ankylosing spondylitis using etanercept. *Case Rep Rheumatol.* 2014;2014:353782.
15. Nascimento AC, Gasparido DB, Cortez TM, et al: Syndrome in question. MAGIC syndrome. *An Bras Dermatol.* 2014;89(1):177-179.
16. Braun T, Fenaux P. Myelodysplastic Syndromes (MDS) and autoimmune disorders (AD): cause or consequence?. *Best Pract Res Clin Haematol.* 2013;26(4):327-336.
17. Yan M, Cooper W, Harper C, et al: Dementia in a patient with non-paraneoplastic limbic encephalitis associated with relapsing polychondritis. *Pathology.* 2006;38(6):596-599.
18. Wang ZJ, Pu CQ, Wang ZJ, et al: Meningoencephalitis or meningitis in relapsing polychondritis: four case reports and a literature review. *J Clin Neurosci.* 2011;18(12):1608-1615.
19. Atsushi N, Yoko O, Takayuki K, et al. Perivasculitic Panencephalitis with Relapsing Polychondritis: An Autopsy Case Report and Review of Previous Cases. *Internal Medicine.* 2014;53 (11):1191-1195.
20. Burns T, Breathnach S, Cox N, et al. *Rook's Textbook of Dermatology*, 8th Edition, Wiley-Blackwell Publishing House, 2010, vol. 4, Ch. 45, pg. 2159.
21. Loricera J, Blanco R, Castañeda S, et al. Tocilizumab in refractory aortitis: study on 16 patients and literature review. *Clin Exp Rheumatol.* 2014 May-Jun;32(3 Suppl 82):S79-89.
22. Morariu SH, Badea M, Capreanu P, et al. Plaque-type morphea secondary to *Borrelia burgdorferi* infection. *DermatoVenerol.* 2013; 58:183-190.
23. Fraser DD, Kong LI, Miller FW, et al. Molecular detection of persistent *Borrelia burgdorferi* in a man with dermatomyositis. *Clin Exp Rheumatol.* 1992;10(4):387-390.
24. Arniaud D, Mattei JP, Pham T, et al. Coexistent dermatomyositis, relapsing polychondritis, and positive Lyme serology. A case-report. *Rev Rhum Engl Ed.* 1997;64(10):589-590.
25. Eisendle K, Baltaci M, Kutzner H. Detection of spirochaetal microorganisms by focus floating microscopy in patients from Central Europe. *Histopathology.* 2008; 52: 877-884.