Laryngotracheal stenosis requiring emergency tracheostomy as the first manifestation of childhood-relapsing polychondritis

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RESUMO

A policondrite recidivante é uma doença rara na infância, de etiologia desconhecida, caracterizada por lesões inflamatórias, recorrentes e destrutivas de cartilagem. A condrite pode ser generalizada e envolve principalmente as cartilagens hialinas laríngea e auricular. Relatamos uma menina de 9 anos e 4 meses de idade, que apresentou laringotraqueítes agudas recorrentes e estenose laringotraqueal como primeira manifestação de policondrite recidivante, e foi submetida à traqueostomia de emergência. Apresentou também condrite auricular e artrite, sendo tratada com prednisona e metotrexato. Concluindo, relatamos um caso raro de policondrite recidivante que apresentou uma complicação laringo-traqueo-brônquica com risco de vida exigindo traqueostomia. Sugerimos que o diagnóstico de PR deva ser considerado em pacientes que apresentem laringotraqueítes agudas recorrentes associadas a outros tipos de condrites, assim como manifestações musculoesqueléticas.

Palavras-chave: Policondrite; Infância; Estenose laríngea; Traqueostomia.

ABSTRACT

Relapsing polychondritis is a rare childhood disorder of unknown etiology, characterized by inflammatory, recurrent and destructive cartilage lesions. The chondritis could be widespread and involves generally laryngeal and auricular hyaline cartilages. We described a 9 years and 4 months old girl, who presented recurrent acute laryngotracheitis and laryngotracheal stenosis, which were the first manifestations of relapsing polychondritis, and was submitted to emergency tracheostomy. She also had ear condritis and arthritis, being treated with prednisolone and methotrexate. In conclusion, we reported a rare case of relapsing polychondritis that presented a life-threatening laryngo-tracheo-bronchial disorder requiring tracheostomy. We suggest that the diagnosis of relapsing polychondritis should be considered for patients who present recurrent acute laryngotracheitis with other types of condritis, as well as musculoskeletal manifestations.

Keywords: Polychondritis; Childhood; Laryngeal stenois; Tracheostomy.

INTRODUCTION

Relapsing polychondritis (RP) is a rare childhood disorder of unknown etiology, characterized by inflammatory, recurrent and destructive cartilage lesions. The chondritis could be widespread and involves generally auricular, nasal and laryngeal cartilages[1-5].

Respiratory symptoms include coughing, inspiratory stridor, hoarseness, wheezing, aphonia and dyspnea[6]. Of note, recurrent acute laryngotracheitis leading to laryngotracheal stenosis was rarely described in pediatric population with RP as the first manifestation of the disease, mostly case reports or case series[6-10]. Moreover, to our knowledge the prevalence of this rare life-threatening laryngo-tracheo-bronchial disorder was not studied.

From January 1983 to May 2012, 5,682 patients were followed at our Pediatric Rheumatology Unit, Instituto da Criança da Faculdade de Medicina da Uni-

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versidade de São Paulo. Only one of them had RP (0,017%) that presented recurrent acute laryngotracheitis with subsequent laryngotracheal stenosis and tracheostomy, as the first manifestation. This RP patient was the only case followed by our service during 29 years and was reported herein.

CASE REPORT

A 9 years and 4 months old female patient had recurrent acute laryngotracheitis without any upper airway involvement that required glucocorticosteroids during episodes. At 10 years old, she had worsening cough, hoarseness, dyspnea, progressive respiratory stridor and respiratory insufficiency due to airway obstruction, and emergency tracheostomy was performed in other service. She had also painful edema with limitation on motion of knees and ankles joints, and methotrexate (0.5 mg/kg/week) was introduced. At 11 years and 3 months old, she was admitted to our Pediatric Rheumatology Unit due to auricular chondritis of the cartilage portion in the left ear without nasal involvement (Figure 1). At that moment, oral laryngo-tracheo-bronchoscopy revealed laryngeal edema with severe narrowing, decreased mobility of the vocal cords and vocal cord polyps, moderate tracheomalacia and severe bronchial thickening with bronchomalacia. The histology showed chronic inflammation of the perichondrium with infiltration of lymphocytes, monocytes and plasma cells, and laryngeal nodule without granulomatous inflammation. The laboratory findings showed hemoglobin 10.5 g/dL, white blood cell count (WBC) 10,300/mm³ (60% neutrophils, 34% lymphocytes, 4% monocytes, 1% eosinophils and 1% basophils) and platelets 670,000/mm³. C-reactive protein (CRP) was 56 mg/dL (normal <5.0), erythrocyte sedimentation rate (ESR) was 55 mm/1st hour, C3 was 149 mg/dL (range 79-152) and C4 was 26 mg/dL (range 16-38). Immunological tests showed antinuclear antibodies (ANA) 1:320 (fine speckled pattern), and negative anti-double-stranded DNA (anti-dsDNA), anti-Sm, anti-RNP, antineutrophil cytoplasmic autoantibodies (ANCA), IgG and IgM anticardiolipin antibodies. Ophthalmologic examination and thoracic X-ray were normal. Therefore, RP diagnosis was established according to diagnostic criteria of Michet et al[11]. Prednisolone (1.0 mg/kg/day) and methotrexate (0.7 mg/kg/week) were used, and glucocorticoid was soon tapered. At 12 years and 8 months old, she was on clinical remission with ESR 26 mm/1st hour, CRP of 8.7 mg/dL and receiving methotrexate (0.4 mg/kg/week). The tracheostomy was maintained due to extensive narrowing of the airways.

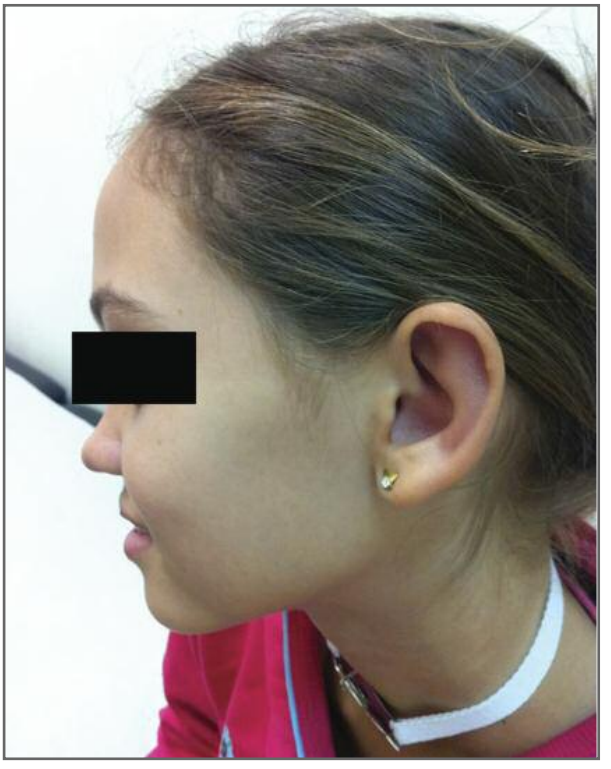

FIGURE 1. Auricular chondritis of the cartilage portion of the left ear without nasal involvement in a young girl with relapsing polychondritis.

DISCUSSION

We described herein a rare case of RP that presented recurrent acute laryngotracheitis with subsequent laryngotracheal stenosis, as the first manifestation of the disease.

Of note, RP is a rare immune mediated disease of unknown etiology[12] characterized by recurrent episodes of inflammation in cartilaginous tissue[6] throughout the body, particularly hyaline cartilage[12]. The incidence is approximately 3 per million population with onset around 40-60 years old[4] and the female-to-male ratio 3:16. Less than 5% of the RP cases occurred in pediatric population, especially in pre-adolescent period, as observed herein[10].

RP diagnosis is performed according to the Michet

et al criteria and requires inflammatory episodes involving at least 2 of 3 sites (auricular, nasal, or laryngotracheal cartilage) or any one of these sites associated with two other manifestations, including ocular inflammation (conjunctivitis, keratitis, episcleritis or uveitis), hearing loss, vestibular dysfunction and soronegative inflammatory arthritis[11], as evidenced in the present case. Additionally, there are no specific laboratory tests for RP diagnosis[13].

The most common cartilaginous tissues involvement in RP are inflammatory hyaline cartilage lesions of ears (85%) and nose (62%), whereas musculoskeletal manifestations (50-75%) usually affects parasternal joints (the sternoclavicular, costochondral, and manubriosternal articulations) and peripheral joints, typically with a non-erosive and asymmetric pattern[14,15]. Involvement of the respiratory tract may vary from asymptomatic to life-threatening complication. Usual symptoms include non-productive coughing, inspiratory stridor, wheezing, hoarseness, aphonia and dyspnea[15]. Laryngotracheobronchial tree chondritis seems to be more frequent in young patients[16] and more severe when compared to adults, which might suggest a greater need for tracheostomy[17]. This involvement generally is triggered by airways infections[6]. Nonetheless, airways obstruction requiring urgent tracheostomy was rarely reported in pediatric population with RP, as observed in our patient as initial manifestation[10,17]. Furthermore, the most important differential diagnosis of RP with laryngotracheobronchial involvement includes primary vasculitis, especially Wegener granulomatosis (recently renamed to granulomatosis with polyangiitis)[18], sarcoidosis, amyloidosis and tuberculosis[15], emphasizing the importance of biopsy performed in the present case. Interestingly, the new validated criteria for Wegener granulomatosis, including patients of our Pediatric Rheumatology Unit, observed that laryngotracheobronchial involvement had a sensitivity and specificity of 22% and 99.8%, respectively[18], however our patient did not fulfill this vasculitis criteria.

The evaluation of the anatomical respiratory involvement in RP is performed by conventional techniques, such as: chest radiography, computed tomography scan and magnetic resonance imaging. Beside, others imaging tests could be realized: bone scintigraphy or positron emission tomography. Bronchoscopy can be helpful in patients with significant affection of airways and regarding obstruction functional, pulmonary function testing could be done[12].

Of note, the empiric use of immunosuppressive agents at disease onset was introduced to our patient due to a possible undifferentiated connective tissue disease[12]. The most important immunosuppressive drugs used in active RP patients are corticosteroids and methotrexate, as used herein. In addition, death in RP patients occurs in up to 50% of the cases due to tracheobronchial obstruction[5,19].

In conclusion, life-threatening laryngotracheobronchial disorder demanding tracheostomy might be the first presentation of RP. Hence, RP diagnosis should not be overlooked in patients who present recurrent acute laryngotracheitis with other types of condritis, as well as musculoskeletal manifestations.

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REFERENCES

1. Letko E, Zafirakis P, Baltatzis S, Voudouri A, Livir-Rallatos C, Foster CS. Relapsing polychondritis: a clinical review. Semin Arthritis Rheum 2002;31:384-395.
2. Dolan DL, Lemmon GB Jr, Teitelbaum SL. Relapsing polychondritis. Analytical literature review and studies on pathogenesis. Am J Med 1966;41:285-299.
3. Foidart JM, Abe S, Martin GR, et al. Antibodies to type II collagen in relapsing polychondritis. N Engl J Med 1978;299: 1203-1207.
4. Kent PD, Michet CJ Jr, Luthra HS. Relapsing polychondritis. Curr Opin Rheumatol 2004;16:56-61.
5. McAdam LP, O`Hanlan MA, Bluestone R, Pearson CM. Relapsing polychondritis: prospective study of 23 patients and a review of the literature. Medicine (Baltimore) 1976;55:193-215.
6. Kim CM, Kim BS, Cho KJ, Hong SJ. Laryngotracheal involvement of relapsing polychondritis in a korean girl. Pediatric Pulmonology 2003;35:314-317.
7. Lipnick RN, Fink CW. Acute airway obstruction in relapsing polychondritis: treatment with pulse methylprednisolone. J Rheumatol 1999;18:98-99.
8. Clark LJ, Wakeel RA, Ormerod AD. Relapsing polychondritis – two cases with tracheal stenosis and inner ear involvement. J Laryngol Otol 1992;106:841-844.
9. Luckey P, Kemper J, Niehues T, Schroten H, Furst G. Diagnostic role of inspiration and expiration CT in a child with relapsing polychondritis. AJR 2001;176:61-62.
10. Belot A, Duquesne A, Job-Deslandre C, et al. Pediatric-onset relapsing polychondritis: case series and systematic review. J Pediatr 2010;156:484-489.
11. Michet CJ Jr., McKenna CH, Luthra HS, O`Fallon WM. Relapsing polychondritis. Survival and predictive role of early disease manifestations. Ann Intern Med 1986;104:74-78.
12. Lahmer T, Treiber M, von Wender A, et al. Relapsing polychondritis: An autoimmune disease with many faces. Autoim-

munity Reviews 2010;9:540-546.
13. Isaak BL, Liesegang TJ, Michet CJ Jr. Ocular and systemic findings in relapsing polychondritis. Ophthalmology 1986;93: 681-689.
14. Letko E, Zafirakis P, Baltatzis S, Voudouri A, Livir-Rallatos C, Foster CS. Relapsing polychondritis: a clinical review. Semin Arthritis Rheum 2002;31:384-395.
15. Edrees A. Relapsing polychondritis: a description of a case and review article. Rheumatol Int 2011;31:707-713.
16. Trepel RJ, Lipinick RN, D`Angelo L. Relapsing polychondritis in an adolescent. J Adolesc Health Care 1989;557-560.
17. Fonseca AR, de Oliveira SK, Rodrigues MC, Aymoré IL, Domingues RC, Sztajnbok FR. Relapsing polychondritis in childhood: three case reports, comparison with adulthood disease and literature review. Rheumatol Int. 2012 (in press)
18. Ozen S, Pistorio A, Iusan SM, et al. Paediatric Rheumatology International Trials Organisation (PRINTO).EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. Ann Rheum Dis 2010;69:798-806.
19. Pearson CM, Kline HM, Newcomer VD. Relapsing polychondritis. N Engl J Med 1960;263:51-58.

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