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Relapsing Polychondritis, an Unusual Cause of Respiratory Failure: A Senegalese Case Report

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Abstract

***Background:** Relapsing polychondritis is a rare systemic disease which is characterised by recurrent inflammation of cartilaginous structures. The involvment of laryngo-trachio-bronchial pathway is the most serious condition of this disease. We report the first complete observation of RP in our area, revealed by chondritis of the respiratory tree with respiratory failure in a Senegalese patient.*

***Case Presentation:** A 39-year-old female Senegalese black patient was admitted in our Institution for a diagnostic challenge and the management of respiratory failure. Clinical examination also showed a saddle nose deformity and a right chronic chondritis of the pinna of the ear. Laboratory investigations revealed a biological inflammatory syndrome and antineutrophil cytoplasmic autoantibodies were negative. Cervical computed tomography showed a complete obstruction of the tracheobronchial pathway. The immediate change was marked by respiratory arrest, requiring its transfer in reanimation. The patient was stabilised by mechanical ventilation and administration of methylprednisolone.*

***Conclusions:** In our observation, the RP has been revealed by chondritis of the respiratory tree. This condition may challenge the vital prognosis. Its evolution can be favourable, butit requires early diagnosis and "aggressive" management.*

Keywords: *Relapsing polychondritis; Emergencies; Africa south of the Sahara; Case report*

Introduction

Relapsing polychondritis (RP) is a rare multisystem disease that was first described in 1923 by Rudolf Jaksch von Wartenhorst, and which is characterised by recurrent inflammation of cartilaginous structures [1, 2, 3]. Among its many clinical manifestations, the involvement of the laryngo-tracheo-bronchial pathway is not uncommon and may challengethe vital prognosis [2]. We report an observation of RP revealed by chondritis of the respiratory tree with respiratory failure in a Senegalese patient.

Case Presentation

A 39-year-old female Senegalese black patient was referred to our institution for a diagnostic evaluation.

In her clinical history, she had been followed for one year for unclassified seronegative arthritis. She presented with a 2-day history of dysphonia, a dry cough and acute dyspnoea with stridor. The haemodynamic constants were as follows: blood pressure: 110/80 mmHg, heart rate: 102 beats/minute, and temperature: 36.5°C. There was acute respiratory failure with polypnoea at 38 breaths/minute and signs of writhing requiring oxygenation. The remainder of the examination revealed a saddle nose deformity (fig.1) and a right chronic chondritis of the pinna of the ear with a soft aspect sparing the ear lobe. In this stage, RP and granulomatosis with

polyangitis were the two main diagnoses considered.

The immediate change was marked by respiratory arrest, requiring its transfer in reanimation. Intubation was difficult to achieve with a paediatric catheter. Mechanical ventilation stabilised the symptoms.

Laboratory investigations revealed a biological inflammatory syndrome. The blood count showed white blood cells at 11.8 Giga/l, haemoglobin at 12.2 g/dl, and platelets at 313 Giga/l. The rate of erythrocyte sedimentation was 100 mm in the first hour and C-reactive protein was 103 mg/l. Immunologically, rheumatoid factors, antinuclear antibodies, anti-CCP and ANCA were negative.

The remainder of the biological assessment (serum creatinine, blood urea, ASAT, ALAT, blood glucose, serum uric acid, and 24-hour proteinuria) was normal, with viral serologies (HBsAg, anti-HCV, and retroviral serology) that were negative.

Cervical computed tomography showed the almost complete obstruction of the tracheobronchial pathway (Fig.2).

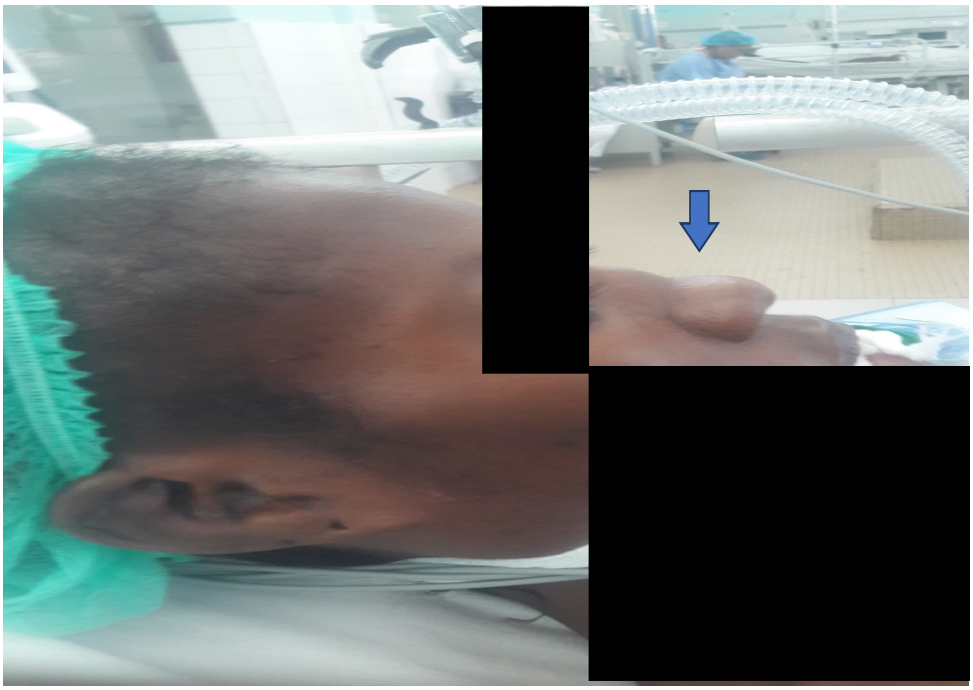

Figure 1. *Saddle nose deformity in our patient (Blue arrow)*

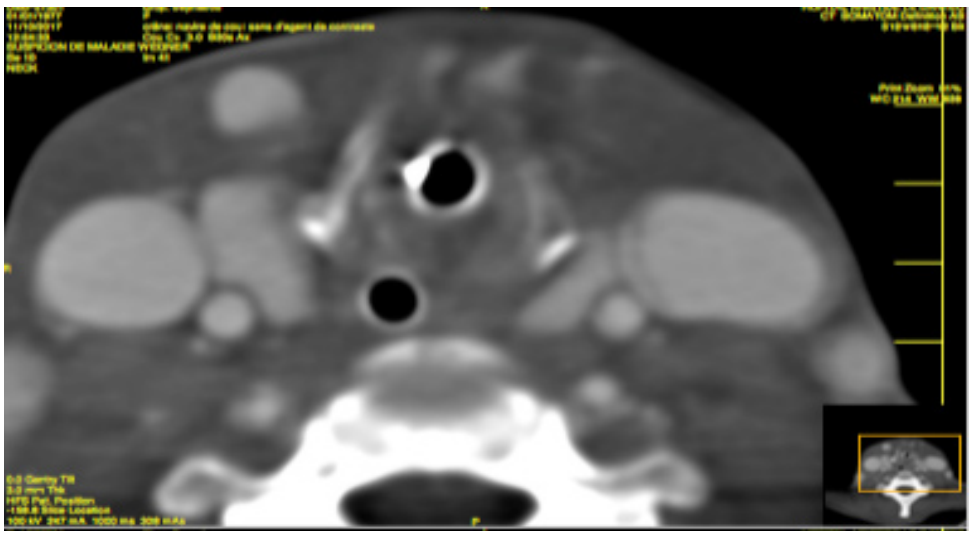

Figure 2. *Cervical computed tomography in transverse view (with intubation catheters). Complete obstruction of the tracheobronchial pathway.*

The diagnosis of RP with laryngo-tracheo-bronchial involvement was retained.

The bolus administration of methylprednisolone (10 mg/kg/day for 3 days) relayed by prednisone at 1 mg/kg/day enabled the patient to be weaned from mechanical ventilation.

The short-term evolution was marked by the less severe recurrence of symptomatology when the glucocorticoid was tapered(below 20 mg of prednisone), justifying the initiation of methotrexate at 15 mg/week.

Evolution is stable to this day.

Discussion

RP is a rare disease; its incidence has been estimated at 3.5 new cases/million inhabitants per year [2, 4, 5]. Since its first description in 1923, about 600 cases have been reported worldwide [1, 2]. In the African literature, to the best of our knowledge, it has been the subject of only three observations from the Maghreb and a scientific communication by a Senegalese group[6, 7, 8, 9]. We report, therefore, the first complete description of this disease in sub-Saharan Africa, revealed by a chondritis of the respiratory tree.

The patient's pathogenesis remains poorly known. This results from an autoimmune disorder,mainly directed against types II, IX and XI collagen and other cartilaginous tissue proteins such as cartilage matrix protein (or matrillin 1) and cartilage oligomeric matrix proteins (COMP) [2, 4, 10]. RP has been associated with HLA DR4 alleles[l3, 4].

RP is a systemic disease with an involvement that may be of interest to ENT specialists, and those of the respiratory, musculoskeletal, cardiovascular, ophthalmological, dermatological, renal and neurological systems[11].

This diagnosis is a real challenge, with a diagnostic delay ranging from 2 to 9 years [12].

Upper airway involvement or laryngotracheal bronchial chondritis has been reported in 20–25% of cases in the early phase of RP and in 50% of cases during its evolution [2, 13, 14, 15]. In our observation, RP and granulomatosis with polyangitiswere the two main diagnoses considered. The key to the diagnosis was the demonstration of extra-respiratory signs characteristic of RP[14, 16]. Indeed, the coexistence of sequelae of auricular and nasal chondrites strongly suggested the diagnosis [10, 14]. Our patient met the criteria proposed by McAdam *et al*., and the criteria of Michet*et al.*[1, 5]. The diagnosis is essentially clinical; a biopsy is not required [11].

In addition, immunologically, antineutrophil cytoplasmic autoantibodies (ANCA) were negative. However, these autoantibodies may be encountered in a true RP, and RP-GPA overlaps have also been reported [3, 4]. Other differential diagnoses to be considered for laryngo-tracheal involvement were sarcoidosis, amyloidosis, rhinoscleroma, respiratory papillomatosis and chondro-osteopathic tracheobrochopathy[11, 14].

The tomodensitometric aspects of airway chondritis are of the type of wall thickening, calcification and narrowing of the airway lumen [2, 3, 5, 17]. In our patient, the imaging was consistent with the severity of the clinical presentation. We noted an almost complete occlusion of the tracheobronchial lumen.

Chondritis of the airways is the most severe disease and the leading cause of death [2, 3, 4, 11]. The treatment of RP is empirical[1, 2, 4]. The severe clinical presentation in our patient had justified the initiation of corticosteroid therapy, methotrexate and the use of an invasive mechanical ventilation procedure [10, 15]. In the absence of early and adapted management, the evolution of this form leads to death in 10–59% of cases or tracheobronchomalacia[2, 5, 11, 13].

Conclusion

RP is an autoimmune inflammatory disease that can affect multiple organs and systems. Laryngo-tracheobronchial chondritis is the most serious condition of the disease. It's an unusual cause of respiratory failure. The key to the diagnosis was the demonstration of extra-respiratory signs characteristic of RP. Its evolution can be favourable, butit requires early diagnosis and "aggressive" management.

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