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Case Report

Carrington's Disease: A Case Report

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Abstract

Chronic Idiopathic Eosinophilic Chronic Pneumopathy (EICP) or Carrington's disease is a chronic inflammatory lung disease, rare, the diagnosis is made in front of the association of predominantly peripheral pulmonary opacities and circulating and/or alveolar eosinophilia, of chronic evolution, with no obvious cause. The clinical picture is progressive, disabling, consisting of dyspnea, cough, chest pain and altered general condition. The time between symptoms and diagnosis is on average more than 3 - 4 months. Corticosteroid therapy allows a rapid regression of symptoms and radiological images. New research may lead to a therapeutic alternative to corticosteroids in the future. We report here the case of a patient with ECDL. We will then present the diagnostic approach, as well as the treatment of this pathology.

Keywords: *Carrington's Disease; Circulating and/or Alveolar Eosinophilia; Corticosteroid Therapy*

Introduction

Chronic Idiopathic Eosinophilic Chronic Pneumopathy (EICP) or Carrington's disease is a chronic inflammatory lung disease, rare, the diagnosis is made in front of the association of predominantly peripheral pulmonary opacities and circulating and/or alveolar eosinophilia, of chronic evolution, with no obvious cause.

Clinical Cases

50-year-old patient, professional hairdresser, without toxic habits, never treated for tuberculosis and without tuberculosis contagion, without personal or family atopy or stay in a tropical area, who had for 7 months gradually developed a permanent dry cough without hemoptysis, associated with Sadoul Stage III assessed stress dyspnea and right-point thoracic pain in the right side accompanied by inflammatory polyarthralgia affecting the ankles and elbows, recurrent xerostomia and oral aphthosis with no other associated thoracic or extra-thoracic signs, all evolving in a context of febrile sensation and altered general condition (asthenia, anorexia and weight loss at 15 kg/7 months).

At admission, the clinical examination objectified a conscious patient, polypneic at 30cpm, sign of respiratory struggle: under costal draw, 84% ODS2 in ambient air, tachycardia at 145bpm, pulmonary auscultation objected to the presence of bilateral crackling moans, without signs of heart failure or thrombophlebitis with the presence of pruritic papules on the dorsal surface of the feet and hands and a pruritic brownish erythema opposite the first right metacarpus.

The chest radiograph showed a dense heterogeneous opacity coarsely rounded right apical seat of some clarity within it with a dense triangular heterogeneous hilo-axillary left retractable opacity and right pleural thickening and a polycyclic hilar right opacity in relation to adenopathy (Figure 1).

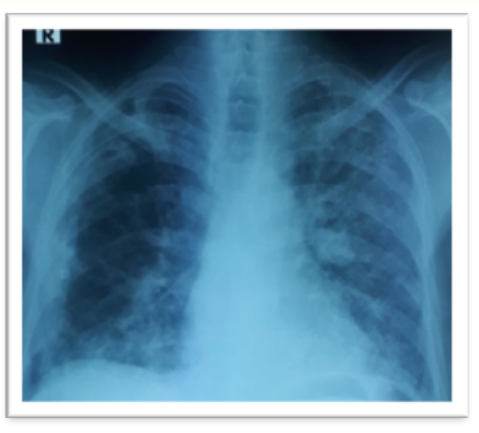

Figure 1: *Chest x-ray of the patient's face on admission.*

Thoracic CT objectified foci of bilateral parenchymal condensation with multiple nodules and micronodules continuing with a right pleural reaction and thickening, several hilar mediastino adenopathies especially of the Barety Lodge and a right pleural effusion of low abundance (Figure 2).

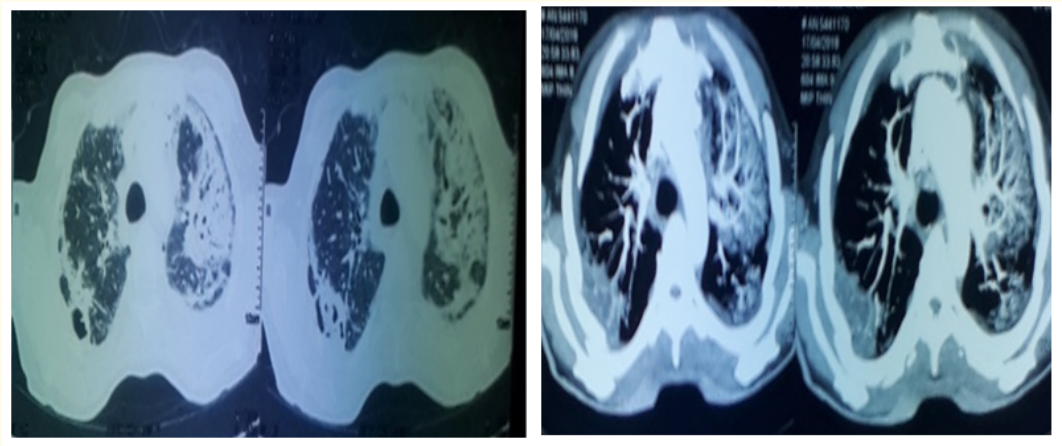

Figure 2: *Thoracic CT was objective of peripheral bilateral parenchymal condensation foci.*

The initial biological assessment had shown hypereosinophilia at 9400 elements/mm^3 with a CRP of 220 and a SV of 96, the immunological assessment was negative, parasitologies in the stool and aspergillus serology were negative, the cardiovascular assessment had not revealed any abnormalities, endoscopic exploration had shown a diffuse inflammatory state in the 2^{nd} degree in the bronchial tree, more marked on the left with an appearance of whitish granulations on the surface of the bronchial mucosa and thickened spurs, biopsy of the thickened spurs and granulations for anatomopathological study had revealed acute bronchitis lesions in acute thrust, rich in eosinophilic

polynuclear cells, bronchoalveolar lavage was rich in PNE, bronchial biopsy for BK culture and aspiration for BK research (ED+C) and expert gene were negative.

The diagnosis of chronic idiopathic to eosinophilic pneumonitis was selected. The patient was put on corticosteroid treatment with a remarkably favourable course: eupneic, ODS2 84% increase to 93% in ambient air, biological PNEs had decreased to 410 elements/mm^3 and radiological (Figure 3).

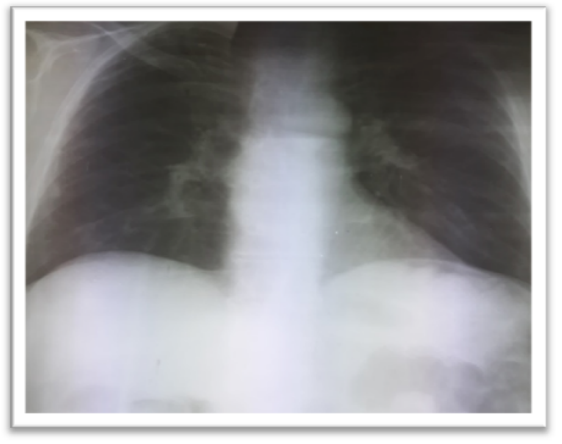

Figure 3: *Radiological cleaning after corticosteroid therapy.*

Discussion

Chronic idiopathic eosinophilic lung disease (CEFL), also known as Carrington's disease, named after the author who first described it in 1969, is part of eosinophilic lung disease without a specific cause [1]. It corresponds to predominantly peripheral pulmonary infiltrates, associated with circulating and/or alveolar eosinophilia [2]. It is a rare disease, the exact incidence and prevalence of which remain unknown [3]. It represents 0 to 2.5% of diffuse infiltrating pneumonitis [4]. There is a female predominance with a sex ratio of 2:1 on average [5,6]. In all published series, two-thirds of patients often have an atopic context with a history of asthma that can precede the disease by more than 20 years [5,6].

A positive diagnosis is made when a combination of arguments is used: the presence of respiratory symptoms and most often general symptoms of a subacute or chronic nature, the presence of blood and/or alveolar eosinophilia, the presence of opacities most often of a alveolar nature in thoracic imaging and the exclusion of any specific cause of parasitic eosinophilic lung disease, in particular helminthiasis or a drug etiology [4].

Indeed, clinical symptomatology is non-specific. It combines respiratory signs with coughing, dyspnea, chest pain and constantly, a change in general condition with fever and vesperal sweating, sometimes suggesting tuberculosis. Pulmonary auscultation finds crackling or sibilant in one third of patients. Extra respiratory manifestations are exceptional and should, a priori, cause the diagnosis to be rejected. Imaging is essential for diagnosis. Classically, it reveals alveolar opacities with poorly defined, unsystematized edges, located in peripheral pulmonary territories, mainly in the upper and middle regions. This rather evocative peripheral topography produces a

negative of the butterfly wing image of pulmonary edema (PAO), however, this typical aspect is present only in a minority of ECDLs. The upper and middle prevalence of lesions distinguishes Carrington's disease from obliterative bronchiolitis with organizational pneumonia (BOOP), whose alveolar opacities are often peripheral but rather localized in the lower lobes. Other unusual radiographic aspects have been reported: systematized alveolar opacities, micro or reticulonodular opacities or pleural effusions [7]. The main interest of thoracic CT is to better define the peripheral topography of the lesions when it is not evident on standard radiography, and the alveolar nature of the opacities ranging from frosted glass appearance to condensation with aerial bronchography. Bronchiectasis is typically absent [8,9]. It is not unusual to observe mediastinal adenopathies or minimal pleural effusions not visible on standard radiography, this was the case for our patient [10,11].

It is the leukocyte formula that most often leads to diagnosis, peripheral blood eosinophilia is constantly very marked. It is generally greater than 1000 elements/mm^3, a biological inflammatory syndrome is frequently reported. An increase in immunoglobulin E levels is present in half of all cases. Alveolar eosinophilia is particularly high, above 40% [2].

Respiratory functional explorations are important for monitoring the disease, especially in the context of frequent association with asthmatic disease. In the period acute, they often show a restrictive ventilatory disorder and a decrease in carbon monoxide transfer. The occurrence of an obstructive ventilatory disorder is common in the subsequent course of the disease even in the absence of a clinical or radiological relapse. The presence of a 'high level' of eosinophilic polymorphs in the BAL at the beginning of the disease is associated with a high risk of developing bronchial obstruction [12,13].

The treatment of ECDL is based on systemic corticosteroid therapy, which results in a dramatic improvement in clinical and radiological signs. General signs recede in a few hours, and radiological abnormalities in a few days. Blood eosinophilia disappears within 24 hours. The initial improvement in corticosteroids is most often total and dramatic. The long-term evolution of the poorly known ECDL is generally favourable [14]. A fibrotic course in the areas initially affected by radiological opacities is exceptional [1]. Significant alveolar hypereosinophilia at diagnosis appears to be associated with an increased risk of developing long-term bronchial obstruction [12].

Conclusion

Carrington's eosinophilic lung disease remains a diagnosis of elimination. The etiological diagnosis in the face of pulmonary eosinophilia must be certain so as not to ignore a particular drug or infectious cause, in particular parasitic, and to conduct treatment in a way that is appropriate for each entity. Finally, it is necessary to know how to identify an eosinophilic lung linked to angelitis and an eosinophilic syndrome because multivisceral involvement often requires appropriate and multidisciplinary management.

Bibliography

1. Carrington CB., *et al*. "Chronic eosinophilic pneumonia". *The New England Journal of Medicine* 280.15 (1969): 787-798.

2. Merchant E and Cordier JF. "Chronic idiopathic to eosinophilic pneumonitis". *La Revue des Maladies Respiratoires* 23.4 (2006): 13S99-1108.

3. Merchant E and Cordier JF. "Chronic idiopathic to eosinophilic pneumonitis". *La Revue des Maladies Respiratoires* 23 (2006): 13S99-113S.

4. Thomeer MJ., *et al*. "Comparison of registries of interstitial lung diseases in three European countries". *European Respiratory Journal* 18 (2001): 114s-118s.

5. Jederlinic PJ., *et al*. "Chronic eosinophilic pneumonia. A report of 19 cases and a review of the literature". *Medicine* 67 (1988): 154-162.

6. Naughton M., *et al*. "Chronic eosinophilic pneumonia. A long-term follow-up of 12 patients". *Chest* 103 (1993): 162-165.

7. Attic P. "Pneumopathies with idiopathic eosinophils". Chest imaging of the adult. Medicine and Science. Paris: Flammarion (1996): 543-544.

8. Mayo JR., *et al*. "Chronic eosinophilic pneumonia: CT findings in six cases". *The American Journal of Roentgenology* 153 (1989): 727-730.

9. Arakawa H., *et al*. "Bronchiolitis obliterans with organizing pneumonia versus chronic eosinophilic pneumonia: high-resolution CT findings in 81 patients". *The American Journal of Roentgenology* 176 (2001): 1053-1058.

10. Ebara H., *et al*. "Chronic eosinophilic pneumonia: evolution of chest radiograms and CT features". *Journal of Computer Assisted Tomography* 18 (1994): 737-744.

11. Marchand E., *et al*. "Idiopathic chronic eosinophilic pneumonia. A clinical and follow-up study of 62 cases". *Medicine* 77 (1998): 299-312.

12. Durieu J., *et al*. "Chronic eosinophilic pneumonia or Carrington's disease". *La Revue des Maladies Respiratoires* 10 (1993) 499-507.

13. Durieu J., *et al*. "Study Group in Interstitial Pathology of the Northern Chest Pathology Society. Long-term follow-up of pulmonary function in chronic eosinophilic pneumonia". *European Respiratory Journal* 10 (1997): 286-291.

14. Naughton M., *et al*. "Chronic eosinophilic pneumonia. A long-term follow-up of 12 patients". *Chest* 103 (1993): 162-165.

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