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Exuberant cystic destruction of lung parenchyma

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All authors were involved in the clinical care of the patient and contributed to and approved the final manuscript.

Conflicts of interest

The authors declare that they have no conflict of interest.

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*Written consent was obtained from the patient.

Introduction

Diffuse cystic lung diseases encompass a broad set of pathophysiologically heterogeneous processes characterized by the presence of multiple thin-walled parenchymal lucencies. Many diseases can present with a tomographic pattern including lymphangioliomyomatosis, pulmonary Langerhans cell histiocytosis, Birt–Hogg–Dubé syndrome, lymphocytic interstitial pneumonia, pneumocystis jiroveci pneumonia, as well as other pathologies[1]. We herein report atypical evolution of the results of the chest computed tomography (CT) of a young patient with hypersensitivity pneumonitis (HP) evolving from typical images of HP to that of an unusual extensive cystic destruction of the lung parenchyma.

Case presentation

A 22 year-old never-smoking male who kept a grey parrot (*Psittacus erithacus*) and a cockatiel (*Nymphicus hollandicus*) in his living room was diagnosed with bird fancier's lung (BFL) on the basis of dyspnea, a restrictive pattern on pulmonary function tests (PFTs) with a total lung capacity (TLC) of 66 % of predicted (*table 1*), typical diffuse centrilobular ground glass nodules on chest CT (*figure 1*), CD8-predominant lymphocytosis (85%) on bronchoalveolar lavage and a positive serum antibody testing for both birds. After well managed treatment with systemic corticosteroids at a conventional dose of 0.75 mg/kg/day for a month followed by gradual reduction over 4 months, the dyspnea disappeared and the chest CT improved considerably (*figure 2*). PFTs also improved up to a TLC of 81 % of predicted (*table 1*). He then resumed exposure to his birds and was lost to follow-up. Five years later he presented with slowly progressive dyspnea and hypoxemia. After thorough

history taking, clinical and blood examination, we concluded that he had no particular inhalational exposure except for avian antigens, no symptoms suggestive of anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitis or tuberous sclerosis, normal protein electrophoresis and alpha-1-antitrypsin levels and negative ANCA. Pulmonary function tests showed a “pseudo-normalization” of the initial restrictive spirometry pattern with a TLC of 7.86 L (104 % of predicted) with a low CO diffusion (51 % of predicted) (*table 1*). A chest CT showed almost exclusively diffuse cystic lung destruction (*figure 3*).

Discussion

BFL is a common form of HP. Patients with acute episodes may evolve to chronic HP but some patients with no identified acute episodes present with an insidious form of chronic HP.[2] Although it is widely unknown why some patients develop an acute form of the disease while others develop chronic HP, it is probable that the type of the antigen, some environmental, antigen-related and genetic factors like telomeropathies play a role in the evolution of the disease and its tomographic pattern. [2–5] The prognosis varies with age, the number of acute exacerbations, the duration of antigen exposure, smoking and other factors. [2,5] The most effective treatment consists of antigen avoidance. Corticosteroids are very effective in the acute context, but do not seem to alter long-term prognosis.[6] In our case report, there are three aspects that are interesting to note.

1- Tomographic presentation

Chest CT may be normal in 8–18% of patients with proven HP. Whenever abnormal, ground glass opacities, mosaic attenuation, gas trapping and poorly defined small centrilobular

nodules are the predominant HRCT findings in acute HP. In chronic HP, reticulations and images of fibrosis associated with the aforementioned signs are frequent.[5] Thin walled cysts and/or emphysematous changes have been reported in up to 20 % of patients with HP [5,7,8]. In a series of 86 patients with bird fancier's lung, areas of emphysema were seen in 7 (17%) of the patients among whom 5 were never-smokers.[10] Cysts are defined as round parenchymal lucency or area of low attenuation, usually thin walled and containing air.[9] They can sometimes be mistaken for emphysema whenever cyst walls are very thin. Cysts in HP are often few and associated with ground-glass opacities or other changes suggestive of HP, unlike in our patient. [10] The pathophysiology of cystic changes in HP is yet to be understood. This may be due to partial bronchiolar obstruction by the peribronchiolar lymphocytic infiltrate [7]. The images shown in this report are very illustrative of the rapid tomographic modification of the patterns of HP in a relatively short period of time. There is a marked difference between the typical early tomographic presentation of the disease, and the far less suggestive tomographic lesions seen in later stages of the disease, whether fibrotic or cystic as in our patient. This makes the diagnosis of HP more challenging when patients are seen at an advanced stage of the disease, in the absence of previous imaging.

2- Functional modification

Although lung function may be normal especially in patients with acute HP, the most frequent functional pattern in patients with chronic HP is restrictive with moderate to severe reduction of the carbon monoxide diffusing capacity. [2,6] An obstructive or mixed pattern is seen in 0.5–33% of the patients in large series of HP. [5,10,11] It has already been reported in the literature that patients with HP and emphysema demonstrate preservation of the

volume of the lungs relative to patients without emphysema. [12] Our patient's PFTs, framed at various moments of the evolution of the lung disease, are particularly interesting because they show that the development of cysts improves lung function test results. The DLCO was the only parameter that worsened in this patient. This pseudo-normalization of PFTs linked to the cystic evolution of the disease is probably a pitfall that clinicians should be aware of.

3- Management of the disease

Concerning the management of HP, compliance of patients to eviction is the key element in clinical improvement. Nonetheless, the emotional value of the antigen source or professional obligations can complicate eviction. In this case, although the risks of persistent exposure to avian antigens were clearly explained to our patient, he disregarded medical advice, kept his birds and was lost to follow-up. He only came back after major and irreversible worsening of his disease. Pulmonologists probably need to be more explicit and straightforward in explaining that the risk of progression is largely avoidable with appropriate antigen eviction. [4] The importance of follow-up should also be stressed in order to avoid end stage lesions in patients with a potentially reversible disease such as HP.

In conclusion, the presence of cysts in chronic HP has been reported in many case series. Nonetheless, this brief report is particularly didactic as it shows a spectrum of the functional and tomographic evolution in one patient as it goes from the typical acute restrictive form to the more unusual chronic cystic form in a particularly short period of time due to the persistence of exposure to antigenic aggression.

References

- [1] Francisco FAF, Souza AS, Zanetti G, Marchiori E. Multiple cystic lung disease. *Eur Respir Rev* 2015;24:552–64. <https://doi.org/10.1183/16000617.0046-2015>.
- [2] Selman M, Pardo A, King TE. Hypersensitivity Pneumonitis. *Am J Respir Crit Care Med* 2012;186:314–24. <https://doi.org/10.1164/rccm.201203-0513Cl>.
- [3] Bouros D, Tzouveleakis A. Telomeropathy in Chronic Hypersensitivity Pneumonitis. *Am J Respir Crit Care Med* 2019;200:1086–7. <https://doi.org/10.1164/rccm.201907-1483ED>.
- [4] Nogueira R, Melo N, Novais e Bastos H, Martins N, Delgado L, Morais A, et al. Hypersensitivity pneumonitis: Antigen diversity and disease implications. *Pulmonology* 2019;25:97–108. <https://doi.org/10.1016/j.pulmoe.2018.07.003>.
- [5] Quirce S, Vandenplas O, Campo P, Cruz MJ, Blay F de, Koschel D, et al. Occupational hypersensitivity pneumonitis: an EAACI position paper. *Allergy* 2016;71:765–79. <https://doi.org/10.1111/all.12866>.
- [6] Chan AL, Juarez MM, Leslie KO, Ismail HA, Albertson TE. Bird Fancier’s Lung: A State-of-the-Art Review. *Clin Rev Allergy Immunol* 2012;43:69–83. <https://doi.org/10.1007/s12016-011-8282-y>.
- [7] Remy-Jardin M, Remy J, Wallaert B, Müller NL. Subacute and chronic bird breeder hypersensitivity pneumonitis: sequential evaluation with CT and correlation with lung function tests and bronchoalveolar lavage. *Radiology* 1993;189:111–8. <https://doi.org/10.1148/radiology.189.1.8372179>.
- [8] Baqir M, White D, Ryu JH. Emphysematous changes in hypersensitivity pneumonitis: A retrospective analysis of 12 patients. *Respir Med Case Rep* 2018;24:25–9. <https://doi.org/10.1016/j.rmcr.2018.03.012>.
- [9] Hansell DM, Bankier AA, MacMahon H, McLoud TC, Müller NL, Remy J. Fleischner Society: glossary of terms for thoracic imaging. *Radiology* 2008;246:697–722. <https://doi.org/10.1148/radiol.2462070712>.
- [10] Morell F, Roger À, Reyes L, Cruz MJ, Murio C, Muñoz X. Bird Fancier’s Lung: A Series of 86 Patients. *Medicine* (Baltimore) 2008;87:110–130. <https://doi.org/10.1097/MD.0b013e31816d1dda>.
- [11] Erkinjuntti-Pekkanen R, Kokkarinen JI, Tukiainen HO, Pekkanen J, Husman K, Terho EO. Long-term outcome of pulmonary function in farmer’s lung: a 14 year follow-up with matched controls. *Eur Respir J* 1997;10:2046–50. <https://doi.org/10.1183/09031936.97.10092046>.
- [12] Jacob J, Odink A, Brun AL, Macaluso C, Lauretis A de, Kokosi M, et al. Functional associations of pleuroparenchymal fibroelastosis and emphysema with hypersensitivity pneumonitis. *Respir Med* 2018;138:95–101. <https://doi.org/10.1016/j.rmed.2018.03.031>.

Table 1: Evolution of pulmonary function tests during the follow-up of the patient.

	FEV1 (%)	FVC (%)	FEV1/FVC	TLC %	DLCO (%)
Baseline	62	58	0.86	66	67
After treatment	95	93	0.86	81	78
After loss to follow-up	108	115	0.75	104	51

Figures

Fig. 1. First chest computed tomography showing typical centrilobular nodules of ground glass density compatible with hypersensitivity pneumonitis

Fig. 2. chest computed tomography showing substantial improvement on corticosteroids. Centrilobular nodules of ground glass are no longer seen.

Fig. 3. Chest computed tomography 5 years later, after continuous exposure to bird antigens, showing extensive cystic destruction of the lungs.





