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References

Development and Co-existence of Sarcoïdosis With Lymphoproliferative Processes

Evolución y coexistencia de sarcoïdosis con procesos linfoproliferativos

To the Editor,

Sarcoïdosis is a multisystemic granulomatous disease during the course of which lymphoproliferative diseases, primarily Hodgkin lymphoma (HL), may be triggered.1,2 In these cases, differential diagnosis between both diseases may be problematic.

We report the clinical case of 26-year-old woman, resident medical officer, with no significant history or toxic habits. She consulted in June 2013 with a clinical picture consistent with respiratory infection. Radiograph revealed right lower lobe pneumonia and she was treated with amoxicillin–clavulanic acid for 10 days. The follow-up radiograph showed regression of the pneumonia and increased bilateral hilar structures suggestive of lymphadenopathies, confirmed on computed tomography (CT), located in the mediastinum and bilateral hilar, the largest being a conglomerate in the thymus. The patient was interviewed again, and reported a 2-year history of asthenia with no other symptoms. On examination, her breathing was normal with basal SaO2 99%, normal cardiopulmonary auscultation, no edemas or other findings. Lung function tests were normal: FVC 92% (3.320 ml), FEV1 100% (3.150 ml), FVC/FEV1 95%, DLCO 112%, TLC 96%, 6MWT 617 m (83% predicted), with no saturation or tachycardia. Clinical laboratory tests showed only mildly elevated angiotensin-converting enzyme 68 IU/L. Endobronchial ultrasound was performed and regions 7 and 10R were aspirated. The microbiological study was negative. Bronchoalveolar lavage immunophenotyping showed no changes. The pathology study reported sarcoïd-like non-necrotizing granulomatous structures with no signs of malignancy. Sarcoïdosis stage I was diagnosed, with no other systemic involvement, and no indication for pharmacological treatment.3

Six months later, the patient reported clinical worsening, but no change was observed with respect to earlier clinical and functional findings. CT revealed a reduction in size of the mediastinal lymphadenopathies, and an anterior mediastinal mass, which corresponded to the lymph node conglomerate located in the thymus, with uptake of 11.5 SUVmax on PET/CT. Left anterior mediastinotomy was performed, which was diagnosed as nodular sclerosis HL.

The patient received 4 cycles of doxorubicin, bleomycin, vinblastine and dacarbazine (ABVD) over 5 months. The post-chemotherapy follow-up CT showed a 65% reduction in the size of the mass, and radiation therapy of the affected field began for 1 month. The patient improved clinically, and the follow-up PET/CT at 3 months revealed a reduction in size and metabolic activity of the mass (4.5 SUVmax), with pathologically increased metabolic activity in the mediastinal (5 SUVmax) and bilateral hilar (7 SUVmax) lymphadenopathies, in the hepatic hilum (3.3 SUVmax) and the spiciform parenchyma, and no other evidence of malignant disease. The case was discussed in the multidisciplinary session, and with the patient’s consent we decided to adopt a wait-and-see attitude and to repeat the PET/CT after 3 months. This procedure revealed disappearance of the mediastinal lymphadenopathies and absence of pathological metabolic activity and malignant disease. Since then the patient has remained asymptomatic, and no changes have been observed in the follow-up CTs, with both diseases being in complete remission. The patient may have had both diseases all along, and the anterior mediastinal image initially thought to be a sarcoïd conglomerate in a lymph gland may actually have been HL.

Sarcoïdosis is a systemic granulomatous disease of unknown etiology, unspecific clinical signs and symptoms, and variable radiological pattern and progress. Diagnosis is obtained from the visualization of non-necrotizing granulomas on histology.

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Hodgkin disease is derived from an alteration in the maturation and activation of B cells in the lymph nodes. It is characterized by the presence of lymphadenopathies. Approximately 25% of patients have general symptoms consisting of the so-called B symptoms: fever, night sweats, and weight loss. Diagnosis is obtained by biopsy, showing characteristic Reed–Sternberg cells on cytology. The nodular sclerosis variant is the most common and has the best prognosis. Treatment is based on chemotherapy and radiation therapy, depending on staging.

The combination of sarcoidosis and lymphoma is unusual, and may be derived from a disordered immune system. Lymphoma–sarcoidosis syndrome was described by Brincker in 1986 after conducting 2 studies, the first in the Danish Clinical Epidemiology Institute, and another subsequent study. Brincker observed that the frequency of lymphoma in patients with pulmonary sarcoidosis was significantly higher than in the general population, the most common being Hodgkin disease, followed by non-Hodgkin lymphoma and other hematological cancers. He also concluded that sarcoidosis normally precedes the lymphoproliferative process by a short period of around 24 months; the inverse order is rarely seen.

The development of sarcoidosis after receiving treatment for a lymphoproliferative process may be due to a hyperresponse of the immune system against the tumor cells. Patients with sarcoidosis may also experience an exacerbation after receiving anticancer treatment. Sarcoid reactions have been reported that are histologically identical to sarcoidosis, and these might be a marker of antitumor response mediated by macrophages activated by T cells.

In conclusion, sarcoidosis and lymphoproliferative processes are diseases in which differential diagnosis can be complicated, but it is important to remember that both diseases may be found in the same patient, either consecutively or simultaneously.

References

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Atypical Chlamydia psittaci Pneumonia. Four Related Cases

Neumonia atípica por Chlamydia psittaci. Cuatro casos relacionados

To the Editor,

Psittacosis is a globally distributed zoonotic disease caused by Chlamydia psittaci (C. psittaci), an intracellular bacteria.1 Birds constitute its main reservoir and the mechanism of transmission is direct contact or inhalation of respiratory secretions or dry feces of infected birds.2 Individuals with occupational or recreational exposure to various birds have a greater risk of infection.3 Psittacosis generally occurs sporadically, but outbreaks have been reported.2 Presentation varies from subclinical infection to severe sepsis and multisystemic involvement.2,4 We describe an outbreak of 4 cases of C. psittaci pneumonia, the focus of infection being a store selling birds (Table 1).

Case 1

A 47-year-old man was admitted with a 5-day history of dyspnea and fever. On admission, he presented tachypnea and crackles in the right lung base, respiratory failure (RF) (PaO₂ 55 mmHg), neutrophilia (91%), elevated C-reactive protein (CRP) (49 mg/dl), procalcitonin (PCT) 1 mg/ml, and alveolar infiltrate in the right lower lobe. Antibiotic treatment with ceftriaxone, levofloxacin and doxcycline was administered for 2 weeks, followed by oseltamivir for 5 days. After 24 h he developed respiratory failure and progressive radiological infiltration, renal failure and shock, so was admitted to the intensive care unit (ICU). Invasive mechanical ventilation was initiated, with noradrenaline 0.4 mcg/kg/min and extrarenal depuration. In view of the lack of response, venovenous extracorporeal membrane oxygenation support (ECMO) was required. Progress was slow and weaning prolonged, and the patient was discharged from the ICU after 60 days, and from the hospital 81 days after admission. C. psittaci DNA was detected in respiratory samples (nasopharyngeal exudate and tracheal aspirate) by polymerase chain reaction (PCR). No other microorganisms were found, including influenza virus. Immunofluorescence testing for C. psittaci serology was positive (IgG seroconversion 1:256 after 14 days from the first sample).

Case 2

A 22-year-old man was admitted with a 1-week history of fever, dry cough, and general malaise. Physical examination showed fever, arterial hypertension, tachycardia, and crackles in the entire right hemithorax. The patient presented respiratory failure (PaO₂ 53 mmHg), neutrophilia (87%), elevated CRP 58 mg/dl, clotting disorders with prothrombin activity 55%, and multilobar infiltrate in right lung. He received high-flow oxygen therapy in the ICU. Ceftriaxone, levofloxacin and doxycline were administered for 2 weeks. C. psittaci serology was positive, with IgG seroconversion (1:256) in convalescent serum. All other microbiological results