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CASE REPORT

Spontaneous increase and decrease of thoracic lymphadenopathy and SACE in sarcoidosis

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Spontaneous increase and decrease of thoracic lymphadenopathy and SACE in sarcoidosis. F. Lhote, D. Valeyre, Ch. Clerici, J. Amouroux, J.P. Battesti.
ABSTRACT: A case of sarcoidosis with iterative spontaneous increase and decrease of thoracic lymphadenopathy is described. No condition such as infectious disease, particular environmental exposure or medication could explain this unusual course of thoracic sarcoidosis.
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In sarcoidosis, the recurrence of lymphadenopathy is very rare after spontaneous and complete resolution, whereas it is often observed in patients treated with corticosteroids. We observed a case in which the course of sarcoid intrathoracic lymphadenopathies was characterized by a frank iterative spontaneous increase and decrease of their size in the absence of any obvious concurrent pathologic condition.

Observation

A 52-year-old caucasian house painter was admitted to the hospital for the first time in June 1977, with a three-week history of productive cough, asthenia and a recent weight loss of 7 kg. He was a smoker (20 cigarettes a day, 35 pack yrs). Clinical examination was normal and a chest X-ray revealed bilateral hilar lymphadenopathy. The Mantoux test (10 U) was negative as was cultural identification of *Mycobacterium tuberculosis*. The serum angiotensin converting enzyme activity (SACE) was 92 $U \cdot ml^{-1}$ (Normal: 22±8.4 $U \cdot ml^{-1}$), the calciuria 0.11 $mM \cdot kg^{-1}$ per day and the serum gammaglobulins 1.7 $g \cdot dl^{-1}$. Tracheobronchial fibroscopy and biopsies of bronchial mucosa were normal. Three months later chest radiography showed an increase of the mediastinal lymphadenopathies.

An open lung biopsy was performed and revealed noncaseating granulomas in the lung parenchyma and in mediastinal lymph node biopsy specimens. No foreign bodies, including birefringent particles, were found. The patient did not receive corticosteroids and was seen every six months. Spontaneous and incomplete roentgenographic resolution of the bilateral hilar lymphadenopathy was noted in May 1978 without development of pulmonary infiltration. In December 1979, a new enlargement of bilateral hilar lymphadenopathy and the appearance of a right para-tracheal lymphadenopathy occurred without clinical manifestation (fig. 1). The SACE was 107 $U \cdot ml^{-1}$. A new spontaneous incomplete roentgenographic resolution was noticed in May 1981 (fig. 2) and the SACE had decreased to 54 $U \cdot ml^{-1}$. In December 1981 the patient complained of asthenia and weight loss of 3 kg and a chest X-ray showed major enlargement of bilateral hilar and para-tracheal lymph glands without evidence of pulmonary infiltrates (fig. 3). The SACE was 106 $U \cdot ml^{-1}$. There were 285,000 cells per ml, with 37% lymphocytes in the fluid recovered by bronchoalveolar lavage. A mediastinoscopy was performed and lymph node biopsy specimens showed confluent noncaseating granulomas. The patient did not receive corticosteroids and in January 1987 was still presenting voluminous and isolated mediastinal lymph nodes.

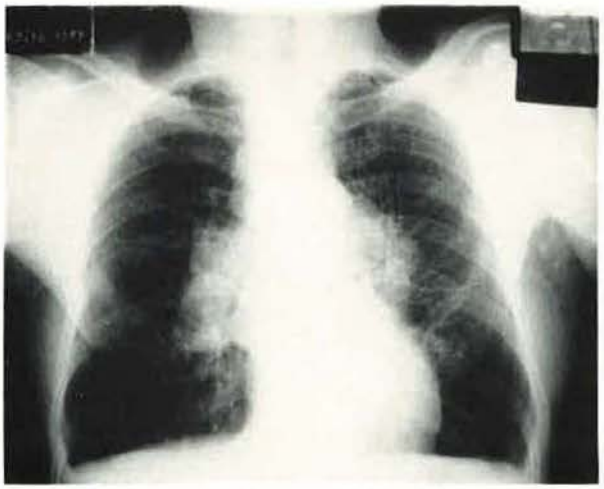

Fig. 1. – Enlargement of right paratracheal and right and left hilar lymph nodes.

Comments

The case reported has been followed for ten years and was characterized by two episodes of incomplete recovery of mediastinal and hilar sarcoid lymphadenopathy. Each episode was followed by an increase of their volume on chest X-ray which justified further

histopathological investigations (open lung biopsy and mediastinoscopy, respectively) to rule out the possibility of an additional disease, particularly lymphoma which occurs with an abnormally high frequency in sarcoid patients [1]. The successive increases and decreases in the size of the mediastinal and hilar lymphadenopathies were always observed in the same place whilst radiological involvement of lung parenchyma was never identified despite repeated chest X-rays.

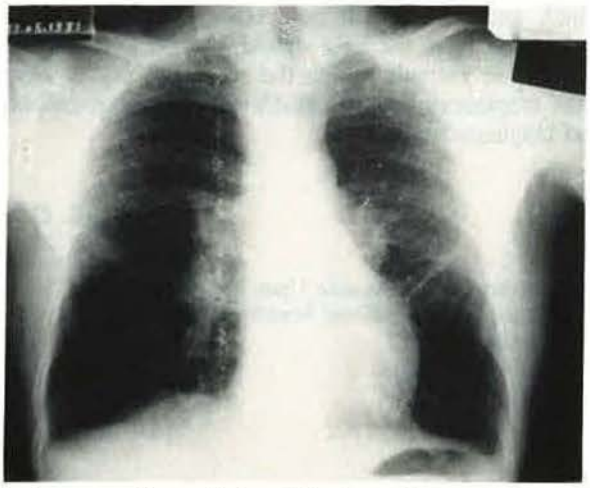

Fig. 2. – Incomplete resolution of hilar lymphadenopathy. Disappearance of right paratracheal lymph nodes.

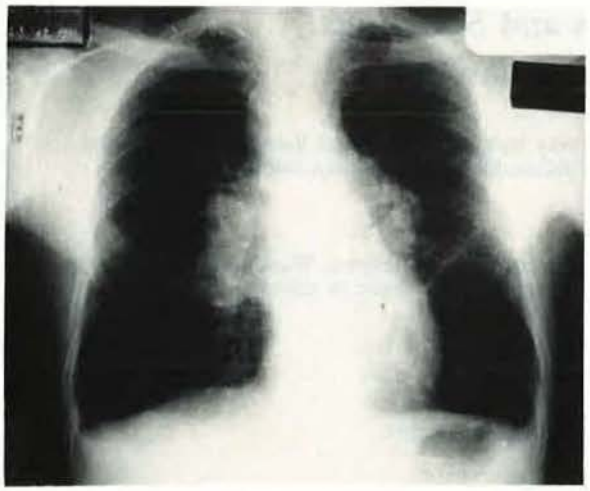

Fig. 3. – New enlargement of hilar and right paratracheal lymph nodes.

The variations of serum angiotensin converting enzyme activity (SACE) noticed in this sarcoid patient, who was free from any clinically overt extrathoracic sarcoid localization, were correlated with the course of thoracic lymphadenopathy. The lack of normalization of SACE level, when the volume of thoracic lymphadenopathy was minimal, indicated the persistence of active sarcoid lesions.

No condition was found to explain such an unusual course of thoracic sarcoidosis, *e.g.* no infectious disease or particular environmental exposure, no medication even given topically. Moreover, biopsy specimens did not reveal any foreign bodies including birefringent particles.

Isolated thoracic lymphadenopathies of sarcoid origin recover spontaneously in 65–80% of cases in less than 2 years [2–3]. In 8–10% of cases they can persist unchanged for many years [4] and are characteristic of stage 1 of chronic evolution [5]. Iterative relapses of the disease following complete recovery are not unusual after cessation of corticosteroid therapy or after delivery [5]. Very rarely, the relapse of sarcoid thoracic lymphadenopathy has been identified several years after an apparent spontaneous complete recovery [6–11].

To our knowledge, a spontaneous recurrence of successive increase and partial decrease of thoracic lymphadenopathy has not previously been recorded and raises questions about the mechanism of such an unusual aperiodic evolution in the absence of any recognized concurrent factor, infectious or environmental. We have also observed (unpublished data) a case of sarcoidosis, followed for four years, in which an identical evolution was noticed in peripheral lymphadenopathy with a parallel evolution of SACE level, however the enlarged thoracic lymphadenopathy in this case remained unchanged during the follow-up.

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RÉSUMÉ: Un cas de sarcoïdose ganglionnaire médiastinal caractérisé par des accroissements et des régressions spontanées du volume des adénopathies médiastinales est rapporté. Des modifications parallèles des taux sériques de l'enzyme de conversion de l'angiotensine ont été notées. Aucune condition: maladie infectieuse, exposition environnementale particulière, prise de médicaments, n'explique cette évolution inhabituelle de la sarcoïdose thoracique.