

eISSN: 2581-9615 CODEN (USA): WJARAI Cross Ref DOI: 10.30574/wjarr Journal homepage: https://wjarr.com/

WJARR	KISSN 2581-9615 CODEN (UBA): BUARAI
W	JARR
World Journal of	
Advanced	
Research and	
Reviews	
	World Journal Series INDIA
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Sarcoidosis revealed by long-term fever at the Vierzon Hospital: About a case and literature review

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World Journal of Advanced Research and Reviews, 2023, 18(02), 572-575

Publication history: Received on 02 April 2023; revised on 09 May 2023; accepted on 12 May 2023

Article DOI: https://doi.org/10.30574/wjarr.2023.18.2.0864

Abstract

Sarcoidosis is a systemic disease mainly affecting the mediastinum and the lungs. The other manifestations are much rarer and lead to diagnostic and therapeutic wanderings. The present study reports a case of lymph node sarcoidosis revealed by a long-lasting fever. The diagnosis is based on clinical and paraclinical arguments and the elimination of other causes of lymphadenopathy. The evolution is favorable after 6 months of corticosteroid therapy.

Keywords: Sarcoidosis; Long-Term Fever; Vierzon; France

1. Introduction

Sarcoidosis is a systemic disease responsible for high mortality compared to the general population [1]. Despite its description for the first time since 1978, the etiological factors still remain unclear [2]. Current data suggest the involvement of environmental factors associated with genetic ones that favor the formation of granuloma [3]. The latter are composed of lymphocytes, macrophages, fibroblasts, epitheloid cells and promote the destruction of the tissues within which they develop [4]. Sarcoidosis may remain asymptomatic for a long time; the manifestations most frequently encountered are often pulmonary and mediastinal, followed by ocular and cutaneous involvement [5]. General signs such as night sweats, asthenia, weight loss and long-term fever are rarely observed [4]. The present study reports a rare presentation of sarcoidosis. This is a young adult woman with long-term fever associated with peripheral lymphadenopathy without pulmonary or mediastinal involvement. After having eliminated the differential diagnoses, the evocative clinical and paraclinical context made it possible to retain sarcoidosis. The evolution was favorable under corticosteroid therapy. A literature review will be made on the manifestations and treatment of sarcoidosis.

2. Medical observation

This is the AF p restorer atient of black Caribbean origin, aged 35, by profession, with no particular history, who consults for fever evolving for 2 months having required several consultations without satisfaction. She did not report any other symptoms apart from a 3 kg weight loss. The examination noted a good general condition, the mucous membranes are normally colored, a fever at 38°2 C with the other vital constants normal. Physical examination showed bilateral cervical and axillary lymphadenopathy without any sign of inflammation. Somatic examination of the other devices was normal.

On the paraclinical level, the biology noted a slight anemia with hemoglobin level 10.3 g / dl, hyperleukocytosis at 12 G / l with neutrophil predominance (83%) without quantitative anomaly of the platelets, a normal sedimentation rate 21

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mm at the first hour and a C reactive protein at 6 mg/l. There was no hypercalcemia or hypercalciuria. Angiotensin 1 converting enzyme was not measured. The search for rheumatoid factors, anti-citrullinated peptide, anti-native DNA, anti-Sm antibodies was negative. The tuberculin skin test was negative. The chest X-ray showed no pleuropulmonary and mediastinal abnormalities. Abdominal ultrasound showed no lymphadenopathy or organ abnormality. Anatomopathological examination of a cervical lymph node had objectified the presence of gigantocellular and epitheloid granulomas without caseous necrosis. The mycobacteriological examination of the biopsy specimen had not noted the presence of pathogenic agents. Faced with the absence of exposure to silica and beryllium, drug intake, the clinical and paraclinical context, the diagnosis of lymph node sarcoidosis was retained.

She received corticosteroid therapy (prednisone 0.5 mg/kg/day) which resulted in apyrexia after 7 days and complete regression of lymphadenopathy after one month of treatment. Corticosteroid therapy was gradually reduced and then stopped after 6 months of treatment. Seen at the check-up after 1 month after stopping treatment, she showed no signs.

3. Discussion

The present study reports a case of sarcoidosis in a young adult revealed by a long-term fever in whom the examination objectified peripheral adenopathies. Sarcoidosis is a disease that affects all ages with a predominance of subjects whose age is between 15-45 years and female subjects [6]. The mediastinum-pulmonary manifestation is the most frequent and takes place in 4 stages during which lymphadenopathy is observed at the hilar and mediastinal level, then the lungs are affected and finally pulmonary fibrosis [1]. Extra-pulmonary manifestations are rarer and consist of cutaneous (sarcoids, lesions on scars, lupus pernio), ocular uveitis, renal or glandular manifestations [5, 7]. The same is true for long-term fever, asthenia or weight loss [8]. Long-term fever was the mode of revelation of sarcoidosis in the studies by Matsuda et al. [9], de Goncalves et al. [10]. Thomas et al. [11] and Kamath et al. [12]. In the study Wang et al. [13], longterm fever and weight loss were observed respectively in 22.5% and 16.25% of subjects followed for sarcoidosis. Haematological involvement, in particular thrombocytopenia, has been described by Pasli et al. [14]. In the present study only the red line is affected with a slight anemia at 10.3 g/dl. Sedimentation rate and C-reactive protein are mostly normal as reported in the present study. Löfgren's syndrome is an acute manifestation of sarcoidosis that combines fever, hilar and mediastinal adenopathy, polyarthritis and erythema nodosum with inflammatory biological signs [15]. Angiotensin-1 converting enzyme is elevated in only 50% of sarcoidosis cases [3, 13]. Hypercalcemia and hypercalciuria are not constant and may be responsible for renal lithiasis complications [16, 17]. Lymph node biopsy provides diagnostic guidance. However, sarcoidosis should be considered as a diagnosis of exclusion because several diseases or situations can mimic the same signs [6, 18]. In the present study, these different differential diagnoses were ruled out. This is the case of tuberculosis by the absence of pulmonary manifestations, tuberculous anergy, the absence of Koch's bacilli and caseous necrosis. The absence of rheumatoid factors, anti-citrullinated peptide, native anti-DNA and anti-Sm antibodies have ruled out auto-immune diseases such as rheumatoid arthritis and systemic lupus erythematosus. The absence of otorhinolaryngological, asthma, pulmonary, eosinophilia or renal impairment signs ruled out vasculitis such as granulomatosis with polyangiitis and eosinophilic granulomatosis with polyangiitis. The absence of exposure to silica and beryllium or drug intake made it possible not to retain the toxic or drug causes. In case of doubt between sarcoidosis and a malignant tumor, positron emission tomography is a useful examination but its routine use is not yet unanimous [19]. Therapeutically, abstention is recommended in cases of asymptomatic stage 1 or 2 lung involvement [3, 20]. In the case of Löfgren's syndrome, treatment is symptomatic, including non-steroidal anti-inflammatory drugs. Corticosteroids represent the first-line treatment that has proven itself in the presence of symptoms, organ failure and in chronic forms [20, 21]. In the event of contraindications or ineffectiveness or in order to reduce the adverse effects of corticosteroid therapy, anti-TNF alpha agents, in particular infliximab, constitute the second-line treatment [22, 23]. Other molecules such as methotrexate, azathioprine, mycophenolate mofetil, leflunomide can be used but with lower levels of scientific proof [3]. Other molecules such as antifibrotics are being tested in cases of pulmonary fibrosis [24]. The evolution of the sarcoid is marked by a spontaneous regression which can reach 50-90% in stages 1 and 2 of mediastinal-pulmonary involvement [23]. In the more advanced forms, this spontaneous regression is rare and is canceled in the event of pulmonary fibrosis. Mortality is also high in severe pulmonary forms and can reach 18% after 5 years of evolution [1, 3].

4. Conclusion

Sarcoidosis is a systemic disease with multiple manifestations. The presence of a long-term fever should lead to the search among many other causes for sarcoidosis. Its diagnosis is based on clinical and paraclinical arguments with elimination of other causes. The treatment of sarcoidosis includes therapeutic abstention in certain situations. Corticosteroids are the molecules of first intention in case of therapeutic need.

Compliance with ethical standards

Acknowledgments

All the staff of the Vierzon hospital in particular ESSAYAN Alain, AGBODJAN Jonas and MEURILLON Chrysoline.

Disclosure of conflict of interest

No conflict of interest.

Statement of ethical approval

Respect for patient confidentiality and fidelity to the information provided by the authors have been guaranteed.

Statement of informed consent

The patient's consent was obtained. Confidentiality of information has been respected

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