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(CASE REPORT)

Atypical sarcoidosis: About a case

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Abstract

Chronic lung consolidation is often associated with various respiratory pathologies, ranging from chronic infections to interstitial diseases such as sarcoidosis, which is a benign systemic granulomatosis of unknown etiology. Interstitial parenchymal involvement is usual. The pseudo-alveolar form is atypical, often presenting acutely, difficult to diagnose, but tends to evolve rapidly favorably under corticosteroid therapy.

We report here a case of pseudo-alveolar sarcoidosis in a 58-year-old patient whose radiological and clinical presentation is unique and perplexing. In this study, we emphasize the rarity of this pseudo-alveolar sarcoidosis.

Keywords: Systemic granulomatosis; Corticosteroids; Chronic consolidations; Non-necrotizing granuloma; Pulmonary sarcoidosis

1. Introduction

Mediastinal-pulmonary sarcoidosis is the most common form of sarcoidosis, which is a systemic granulomatous disease of uncertain cause. Involvement of the hilar and mediastinal lymph nodes is one of the most frequently observed manifestations of the disease [1]. The pseudo-alveolar form is uncommon, often acute, and can be isolated or associated with mediastinal involvement. This atypical form is characterized by granulomatous infiltrates in lung structures resembling alveoli, but without the classic histopathological features of the alveolar granulomas typical of sarcoidosis. Recognizing this variant is crucial for accurate diagnosis and for differentiating pseudo-alveolar sarcoidosis from other interstitial lung pathologies [2]. Although spontaneous regression is possible, the disease can be chronic and can be effectively treated with oral corticosteroids.

2. Case presentation

Mrs. E.F, a 58-year-old woman with no professional or environmental exposure, was treated for pleuropulmonary tuberculosis 40 years ago, which was declared cured. She has a history of thyroidectomy two years ago and is on levothyroxine, a rheumatic fever without carditis treated with extencillin for 30 years (stopped three months ago), and hypertension treated with Tanzaar for one month. Four months prior to her admission, she presented with stage II dyspnea according to Sadoul, associated with a productive cough producing whitish sputum, without hemoptysis or chest pain, and without other thoracic or extra-thoracic signs. All of this occurred in the context of afebrility and preserved general condition. The pleuropulmonary examination revealed bilateral velcro crackles

The ophthalmological examination was normal except for a positive Schirmer test (Right eye 4mm; Left eye 5mm).

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The chest X-ray revealed an alveolo-interstitial syndrome with heterogeneous para-hilar bilateral opacities associated with diffuse bilateral micronodules ***(Figure 1).***

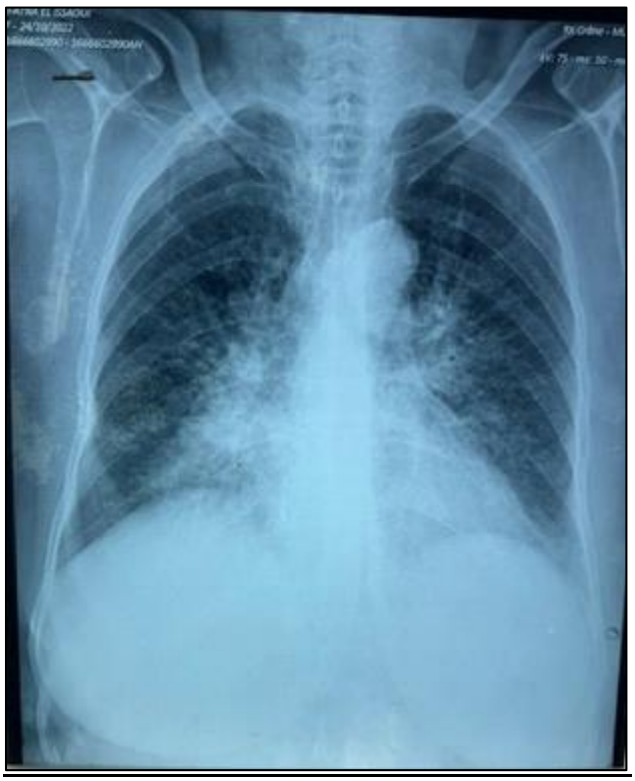

Figure 1 Bilateral alveolo-interstitial syndrome on chest X-ray

The search for mycobacterium tuberculosis in the sputum and the Xpert gene test in the sputum were both negative.

High-resolution chest computed tomography (CT) showed a focal area of pulmonary consolidation with a systematic air bronchogram in the middle lobe, associated with interstitial involvement featuring perilymphatic micronodules, peribronchovascular thickening, and calcified mediastinal lymphadenopathy ***(Figure 2).***

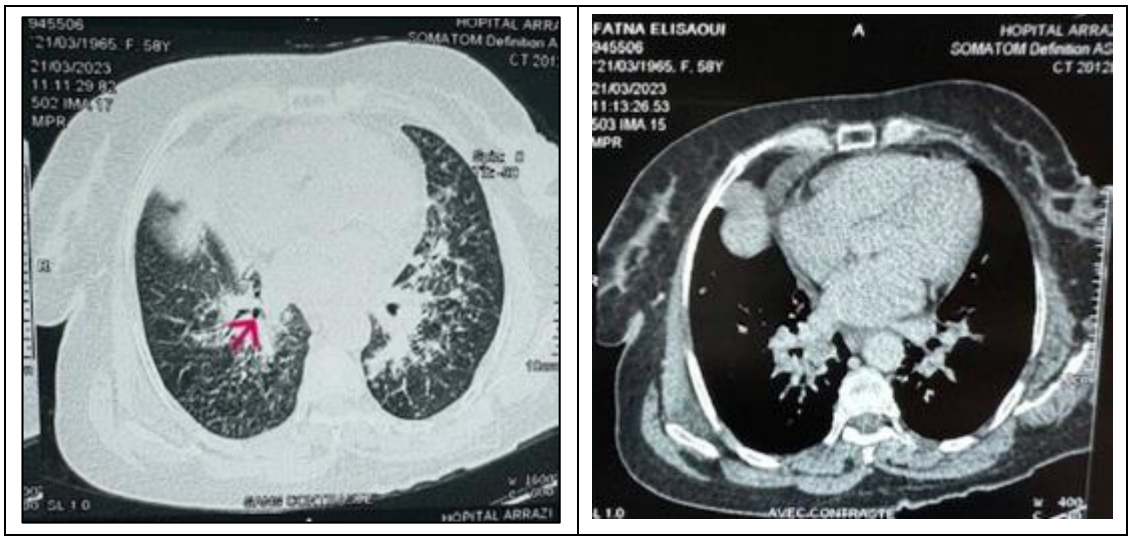

Figure 2 HR chest CT showing an area of alveolar consolidation associated with interstitial involvement

Bronchoscopy revealed an endoscopic appearance without particularities in the right and left bronchi; the orifices were clear, and the spurs were thin. No visible buds or granulomas were observed. The cytology was dystrophic and slightly inflammatory, with no suspicious cells. The search for acid-fast bacilli in the bronchial aspirates was negative, and the bronchoalveolar lavage (BAL) was inflammatory, predominantly lymphocytic, with a CD4/CD8 ratio estimated at 2. The staged bronchial biopsies showed epithelioid and giant cell granulomatous lesions, non-necrotizing of the bronchial mucosa, with no specific characteristics and no cytological or histological signs of malignancy ***(Figure 3).***

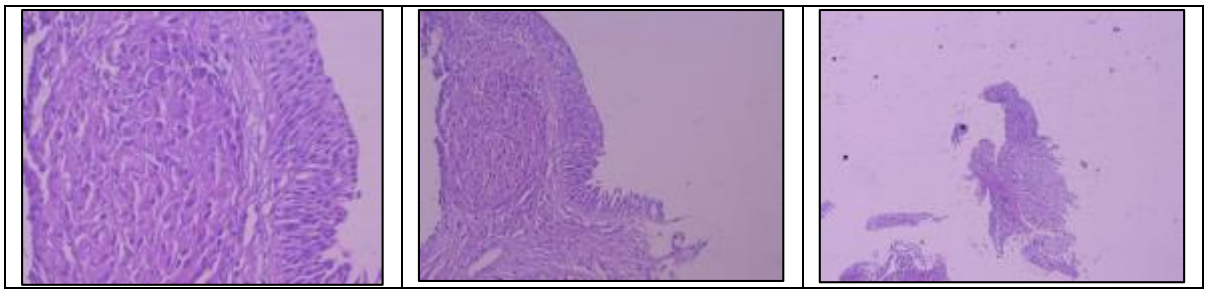

Figure 3 Histological appearance at high, medium, and low magnification of a granulomatous inflammatory lesion with epithelioid and giant cells, without caseous necrosis, in the staged bronchial biopsy

The measurement of angiotensin-converting enzyme revealed a serum activity of 68 UI/L. The calcium-phosphate assessment showed normal blood and urinary calcium and phosphate levels. Serum protein electrophoresis was negative. The complete immunological evaluation showed no abnormalities, and the tests for avian and farmer's lung precipitins were negative. The salivary gland biopsy showed subacute and chronic nonspecific sialadenitis of grade 2 according to Chisholm and Mason. A plethysmography was performed as part of the impact assessment, but the patient was non-cooperative. Cardiac evaluation was normal. Treatment with oral corticosteroids, specifically prednisone at 40 mg/day, was initiated, resulting in a regression of dyspnea.

3. Discussion

Sarcoidosis is an inflammatory, multisystem disease of unknown cause, with highly varied clinical manifestations. Although this disease can affect virtually any organ in the body, it primarily targets the lungs, lymphatic system, skin, eyes, or a combination of these sites, and is characterized by the formation of non-caseating granulomas [3].

Atypical manifestations, such as the pseudo-alveolar form, can indicate a range of differential diagnoses, which delays diagnosis.

In our study, we report the pseudo-alveolar form of sarcoidosis in a 58-year-old woman. This is consistent with the literature, which indicates a female predominance with a peak incidence between 45 and 60 years, corresponding to the perimenopausal peak [4].

Classic radiological manifestations include mediastinal lymphadenopathy and interstitial infiltrates [5]. However, in our study, the CT scan revealed parenchymal consolidation associated with interstitial involvement featuring perilymphatic micronodules, peribronchovascular thickening, and calcified mediastinal lymphadenopathy, thus suggesting a pseudo-alveolar form of sarcoidosis. This phenomenon can be attributed to the formation of sarcoid granulomas within the lung structures, leading to alteration of the surrounding tissues and prolonged infiltration [6]. The underlying mechanisms of this atypical presentation remain to be clarified. Previous studies have suggested that sarcoidosis can lead to changes in alveolar structures, often observed as consolidation or nodules [7, 8].

The identification of this pseudo-alveolar presentation is crucial for differential diagnosis. Clinicians must be attentive to these atypical manifestations to avoid misdiagnoses, such as pulmonary infections or neoplasms, which may present with similar radiological signs [9, 10]. Indeed, a rigorous diagnostic approach, including all biological, histological, bronchial biopsy, and radiological results, while ruling out other causes of granulomatous etiologies, allows for the establishment of a diagnosis of sarcoidosis [11]. The treatment of pseudo-alveolar sarcoidosis follows the general guidelines for sarcoidosis, with corticosteroids as the first-line treatment [12]. However, the response to treatment may vary depending on the clinical presentation and disease progression. Patients with atypical forms may require closer monitoring to adjust treatment based on clinical response and radiological evolution [13,14].

4. Conclusion

Pulmonary sarcoidosis in its pseudo-alveolar form is a rare and complex presentation that often poses a diagnostic challenge in respiratory pathology. It is crucial to recognize the rarity of this form and the associated diagnostic challenges, as well as to maintain early suspicion, especially when there is a discrepancy between the extent of alveolar abnormalities observed in imaging and the severity of clinical symptoms. A thorough understanding of these atypical forms of sarcoidosis can help improve diagnostic and therapeutic strategies, thereby offering a better quality of life to patients affected by this complex disease.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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