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Case Report

SINGLE NODULAR SARCOIDOSIS: Rare and Intriguing Presentation

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Abstract

A 50 years old Caucasian male driver, from Rio de Janeiro, Brazil, sought care in September 2019 for presenti an asymptomatic right lower lobe nodular radiological alteration. He reports having had sarcoidosis in 201 abandoning follow-up without treatment in 2013, but with regression of the condition. In order to rule out confirm the hypothesis of cancer, the patient underwent a series of routine tests ending with a thoracotomy th revealed a chronic granulomatous inflammatory process.

Key Words: single nodular sarcoidosis; lymph nodes; sarcoid necrotizing granulomatosis

Introduction

Sarcoidosis is a granulomatous disease characterized by the presence of granulomas without necrosis, however some degree of necrosis can be found [1]. Intra thoracic involvement (lymph nodes and lung) is predominant, with a pattern of pulmonary involvement preferentially diffuse and bilateral along the bronchovascular bundles. Some presentations considered atypical are well described in sarcoidosis. Among these, nodular sarcoidosis deserves some attention not only for its differential diagnosis sometimes difficult with neoplasms, but also with other diseases. Another important aspect is the existence of necrosis, which can affect up to 14% of cases in nodular sarcoidosis [2]. Other non-neoplastic diseases can also present in their course of evolution in a nodular form with a certain degree of necrosis such as tuberculosis, fungal diseases and Sarcoid Necrotizing Granulomatosis (SNG) to name a few.

Case Report

Patient EG, a 50 years old white male driver, living in São Gonçalo/RJ-Brazil, sought care in September 2019 for presenting asymptomatic radiological alterations (Figure 1A and 1D). He reports having had sarcoidosis in 2011 (Figure 1B), abandoning follow-up without treatment in 2013, but with regression of the condition (Figure 1C). He also has Systemic Arterial Hypertension, Asthma and report of kidney stones. He smoked 40 packs/year until 2004. Normal physical examination. Blood count, biochemistry and angiotensin-converting enzyme were normal. Fungal serologies (histoplasma, Paracoccidioidomycosis, cryptococcosis and aspergillus) were negative. Quantiferon-TB and PPD were negative. Fiberoptic bronchoscopy was normal. The bronchoalveolar lavage (BAL) study for tuberculosis (AFB research, GeneXpert and culture) fungi (direct and culture) and neoplastic cytology were negative. PET-CT and 68Ga DOTATOC PET-CT were negative. Histopathological study of material removed by thoracotomy showed a chronic granulomatous inflammatory process with participation of giant cells, epithelioid cells and necrosis. Presence of asteroid body in giant cell. Special stains were negative for AFB and fungi. No malignancy was observed in the examined sample (Figure 1E, 1F, 1G and 1H). Subsequently, all materials (BAL and biopsy fragment) sent for culture (*M. tuberculosis and fungi*) came with negative results, concluding the diagnosis of Nodular Sarcoidosis.1.

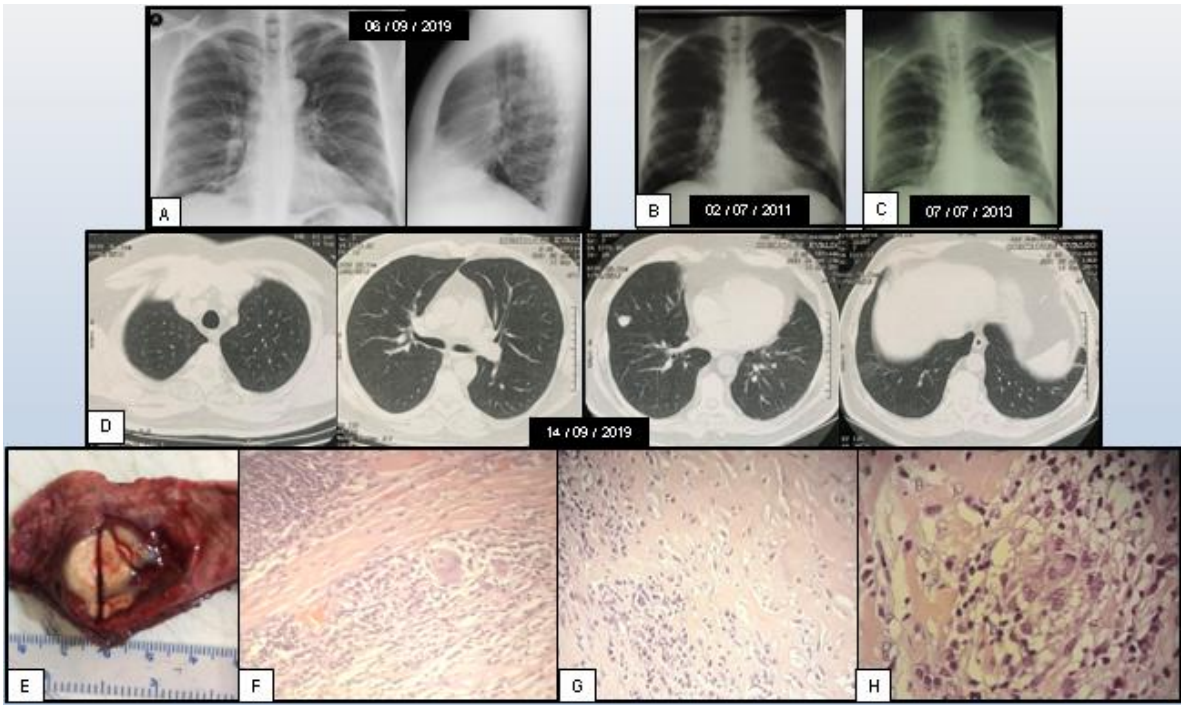

Figure 1: A) 2019 thorax X-ray showing a nodular lesion in the right lower lobe; **B)** 2011 thorax X-ray showing bilateral hilar lymphadenopathy; **C)** 2013 thorax X-ray showing bilateral hilar lymphadenopathy regression; **D)** 2019 Thorax High Resolution Computed Tomography study; **E)** Nodular fragment of lung tissue with a smooth white lesion; **F)** Encapsulated area of granulomatous inflammatory process with multinucleated giant cell presenting an asteroid boby; **G)** Fibrinoid necrosis; **H)** Granuloma with epithelioid cells.

Discussion

The diagnosis of sarcoidosis remains a challenge. Although sarcoidosis has been known for more than a century, its etiological agent is still unknown and its diagnostic construction is still based on situations that can be slippery. The nodular form of presentation, although uncommon, is well described in Sarcoidosis [3]. This can be externalized with multiple or isolated nodules. It is a presentation that always brings us concern with differential diagnosis of neoplastic disease, whether metastatic when multiple or with primary pulmonary tumor when isolated. Furthermore, a certain relationship between sarcoidosis and cancer has long been described in the literature [4]. Sarcoid granulomas may coexist with some neoplasms, either peri-tumoral or even in theirs draining lymph nodes [5]. In these circumstances, the differentiation between sarcoid reaction and sarcoidosis can be very difficult. Neoplasms have been described after some time of the existence of sarcoidosis more frequently than the opposite direction, that is sarcoidosis after the neoplasm [6]. Different types of tumors are shyly pointed out with a certain relationship with sarcoidosis. The lung tumor, however, has a controversial position, either with a higher incidence [5] or reporting less evidence of association and speculating as a reason that sarcoidosis is most frequent in non-smokers [4]. In this patient, the single tumor in the RLL had a negative PET-CT uptake study, making the diagnosis of cancer unlikely. Carcinoid tumors can behave without PET-CT uptake and there are also reports in the literature of an association between these and sarcoidosis [7]. Negative 68Ga DOTATEC PET-CT uptake also greatly decreased the likelihood of carcinoid tumor.

The histopathological study of the lesion revealed a chronic granulomatous inflammatory process with necrosis and the presence of an asteroid body. No histological alteration suggestive of neoplasia or carcinoid tumor were evidenced. The presence of necrosis always leads us to think of infection and tuberculosis among these, especially in the city of Rio de Janeiro, Brazil, where there is a high incidence of this disease. However, the patient had a non-reactive PPD, non-reactive IGRA, all investigations for tuberculosis (direct examination, GeneXpert and culture) performed on the BAL and on the biopsy material were negative. Similarly, the investigation of fungal infection (direct examination and culture), either in BAL or in a biopsy fragment of the material removed, and also the performance of serology for these most frequent infections (Paracoccidioidomycosis, Histoplasmosis, Cryptococcosis and Aspergillosis) were negative. These facts ensure, in a way, the exclusion of these diagnoses (Tuberculosis and fungal infections).

Another disease to be considered in the differential diagnosis would be SNG. It was primarily described by Liebow [8] in 1973. It is a rare disease with findings that lie between sarcoidosis and what was formerly known as Wegner's Granulomatosis, with necrosis and vascular granulomatous involvement. The description of the histopathological material doesn't mention any angiitis with sarcoid reaction or granuloma s in the vessels. On the other hand, there are unicist groups in the literature that believe that SNG and Nodular Sarcoidosis are manifestation of the same disease named Sarcoidosis. Their arguments are mainly based on the existence of more similarities than discrepancies between the two presentation, SNG and Nodular sarcoidosis [9,10]. Liebow himself, in his description, asked the following question that remains current: '*the problem is whether the disease represents necrotizing angiitis with sarcoid reaction, or sarcoidosis with necrosis of the granulomas and of the vessels*.'

In summary, Sarcoidosis continues to be an intriguing disease whose diagnosis still has a certain amount of clinical conviction in the case. However, the basic postulates for its diagnosis must be pursued, that is, a compatible clinical radiological picture, evidence of a granulomatous structure, and ruling out other known causes of granulomatous lesions, especially when faced with an unusual presentation such as isolated nodular

form. Therefore, the diagnosis was concluded as Nodular Sarcoidosis and, for those with unicist thinking, another spectrum of presentation of Sarcoidosis.

References

1. Ma Y, Gal A, Koss MN. (2007). The pathology of pulmonary sarcoidosis: update. *Semin Diagn Pathol*. 24(3):150-161.
2. Rosen Y. (2015). Necrotizing Sarcoid Granulomatosis. *Arch Pathol Lab Med*. 139:252–262.
3. Malaisamy S, Dalal B, Bimenyuy C, Soubani AO. (2009). The Clinical and Radiologic Features of Nodular Pulmonary Sarcoidosis. *Lung* 187:9–15.
4. Chopra A & Judson MA. (2015). How are câncer and connective tissue diseases related to sarcoidosis? *Curr Opin Pulm Med* 21:517–524.
5. Brincker H. (1989). Coexistence of Sarcoidosis and malignant Diseases:Causality or Coincidence? *Sarcoidosism* 6:31-43.
6. Grados A, Ebbo M, Bernit E, et al. (2015). Sarcoidosis Occurring After Solid Cancer: A Nonfortuitous Association. Medicine 94(28):1-9.
7. Álvarez RL, Alvarez II, Fabrellas EF, Olivas RB. (2012). Association of Carcinoid tumor and Sarcoidosis. Arch Bronconeumol. 48(12**)**:469–471.
8. Liebow AA, The J. (1973). Burns Amberson lecture: pulmonar angiitis and granulomatosis. *Amn Rev Respir Dis.* 108(1):1-18.
9. Popper HH, Klemen H, Colby TV, Churg A. (2003). Necrotizing sarcoid granulomatosis; is it different from nodular sarcoidosis? *Pneumologie* 57:268–271.
10. Churg A, Carrington CB, Gupta R. (1979). Necrotizing sarcoid granulomatosis. 76:406–413.

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