SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES 2016; 33; 292-296

Spontaneous resolution of a tumor like pulmonary sarcoidosis

Zied Moatemri, Ghassen Soussi, Salsabil Dabboussi, Samira Mhamdi, Chiraz Aichaouia, Mohsen Khadraoui, Rezaik Cheikh
Respiratory department Hôpital militaire de Tunis, Faculté de médecine de Tunis, Université Tunis El Manar

Abstract. We report a case of thoracic sarcoidosis in a 72-year-old female, snuff taker, who presented with multinodular pulmonary lesions on chest x-ray. Clinical and biological findings were poor. Thoracic imaging showed soft tissue density nodules with irregular borders. The diagnosis of 'cannon ball' metastases was suspected. A thorough investigation strategy could not prove malignancy. A complete radiologic clearing was obtained spontaneously within three months. A rereading of pathology slides performed afterwards showed multinucleated giant cells on hemorrhagic background with a lymphocytic alveolitis. The diagnosis of pseudotumoral sarcoidosis was made. The clinical course was favorable with a 6years follow-up. *(Sarcoidosis Vasc Diffuse Lung Dis 2016; 36: 292-296)*

Key words: sarcoidosis, lung masses, transthoracic biopsy

Background

Often labeled 'the great mimic' (1), thoracic sarcoidosis fore shadows an initially broad differential diagnosis with a burden some thought of malignancy beneath. As a matter of fact, physicians are rather prone to retain the latter diagnosis, a fortiori when imaging shows a 'cannonball' aspect, which is obviously uncommon inthoracic sarcoidosis. We report a rarely-seen case of pseudotumoral sarcoidosis with spontaneous resolution within a few weeks.

Case presentation

A 72-year-old female patient, with history of snuff taking, hiatal hernia and postmenopausal vaginal bleeding presented with dry cough, intermittent chest pain, lethargy and loss of appetite for 3 months. There was no associated history of fever, night sweats or extra-thoracic symptoms. Upon admission, the physical examination showed no fever and no chest abnormalities. Chest x-ray revealed scattered multiple pulmonary nodules and a retrocardiac image with an air-fluid level in relation to the hiatal hernia (Figure 1).Electrocardiogram showed no anomalies. Blood tests showed normocytic normochromic anemia with a hemoglobin level of 11.9 g/dL and normal white cell and platelet counts. Tests for hemostasis and blood biochemistry were correct. There were no signs of biological inflammation. The chest x-ray findings urged us on performing a computed tomography (CT) scan which showed multiple soft tissue density nodules with irregular bordersand no evidence of axillary, hilar or mediastinal lymph nodes meeting size criteria for lymphadenopathy (Figure 2). The diagnosis of 'cannon ball' lung metastases was suspected. In order to get histological evidence of the potential tumoral process, fibrobronchoscopy was performed and showed no anomalies. Aspira-

Received: 10 March 2015
Accepted after revision: 15 September 2015
Correspondence: Zied Moatemri
Respiratory department
Military Hospital of Tunis
Montfleuri 1008, Tunis, Tunisia
E-mail: moatemrizied@yahoo.fr

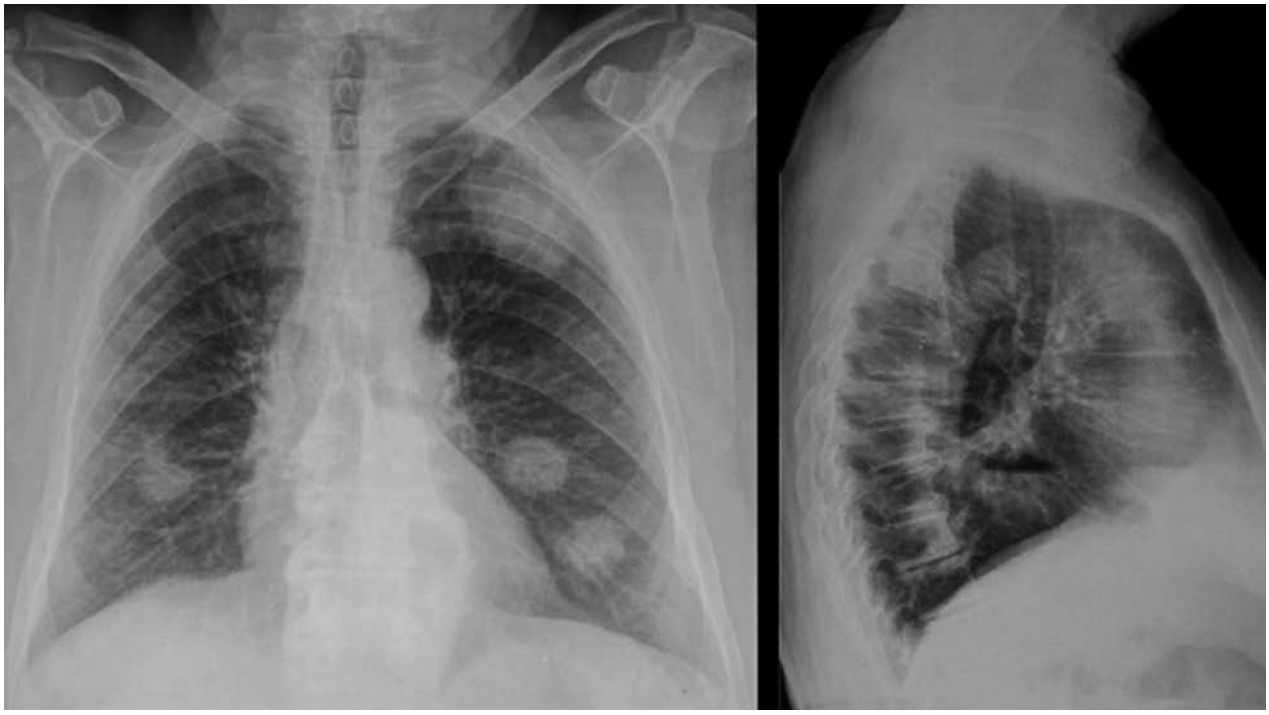

Fig. 1.

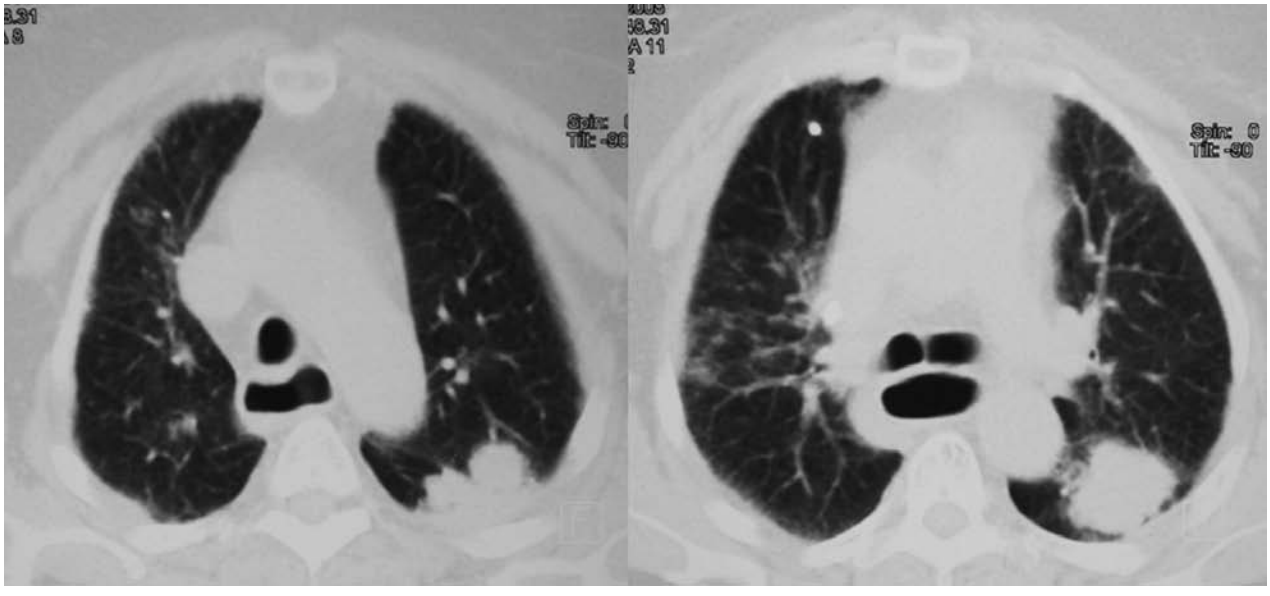

Fig. 2.

tion cytology was benign. CT-guided biopsy of the lung showed necrotic areas with cells originating from either the thyroid gland or the gynecologic tract. On thyroid ultrasound, the right lobe showed a small non suspect nodule of 11 mm occupying nearly the middle third of the lobe. Thyroid function tests were normal. The gynecologic examination was normal too, but pelvic ultrasound imaging

revealed a small uterus with an anechoic lesion of 5.6 x 14.7 cm. Hysteroscopy with endometrial curettage and biopsy under general anesthesia revealed a post-menopausal endometrial atrophy. Echomammography revealed bilateral BI-RADS category 2 lesions. Tumor markers (CEA, AFP and CA125) levels were normal. Bone scintigraphy showed no evidence of osseous metastatic disease. Esophagogastroduodenoscopy (EGD) revealed grade A peptic esophagitis (Los Angeles Classification) and a sliding hiatus hernia. Abdominal CT scan demonstrated a roughly 5.5 cm diameter round, partially calcified mass in the right hepatic lobe, highly suggestive of hydatid cyst. Serological tests for hydatid disease were negative though. Sputum smears examination screening *Mycobacterium tuberculosis* was negative. Mantoux tuberculin skin test (TST) showed a complete anergy. These findings tally with multiple lung hydatidosis but our concern was with the probability of metastatic lesions of unknown primary origin. A few days later, the patient decided to interrupt any further investigations and was discharged from hospital. She presented only three months later, reporting a progressive and spontaneous clinical remission. Checking up on chest x-ray, a complete radiologic clearing was found (Figure 3). On CT scan, inferior lobes nodules have totally disappeared; only one excavated nodule persisted in the upper left lobe with a few micronodules in the right middle lobe (Figure 4). Pseudotumoral sarcoidosis was the most likely diagnosis. We proceeded to a second fibrobronchoscopy which showed an inflammatory mucosa, bleeding easily in contact with the fibroscope. Multiple biopsy sampling showed no tuberculoid granuloma. Bronchoalveolar lavage showed lymphocytic alveolitis with a CD4/CD8 ratio of 0.35. No tumoral cells were found. Angiotensin-converting enzyme blood levels were normal. Pulmonary function testing showed no ventilatory defects. Immunological tests were negative. A control fibrobronchoscopy showed a saber-sheath trachea with a normal bronchial tree. Biopsies were negative. We eventually proceeded to a thorough rereading of the pathology slides obtained by CT-guided biopsy of the lung nodules which showed a few multinucleated giant cells on hemorrhagic background with a lymphocytic alveolitis. The diagnosis of pseudotumoral sarcoidosis was therefore made. A steady follow-up with no specific therapy was decided. A CT scan, performed three years later, showed bilateral interstitial lesions with ground-glass opacities in both lower lobes as well as an enlarged aortopulmonary window lymph node (Figure 5).In six years of hindsight, the patient was feeling well, only complaining of dry cough and mild arthralgia. Thoracic imaging results showed stable lesions.

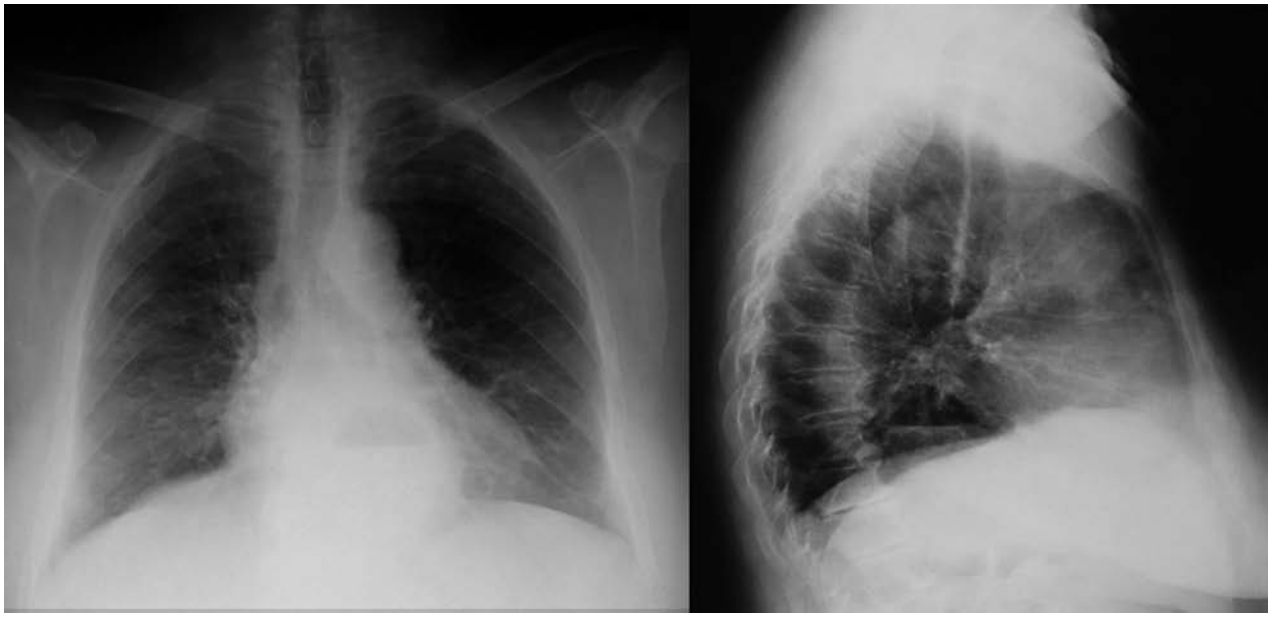

Fig. 3.

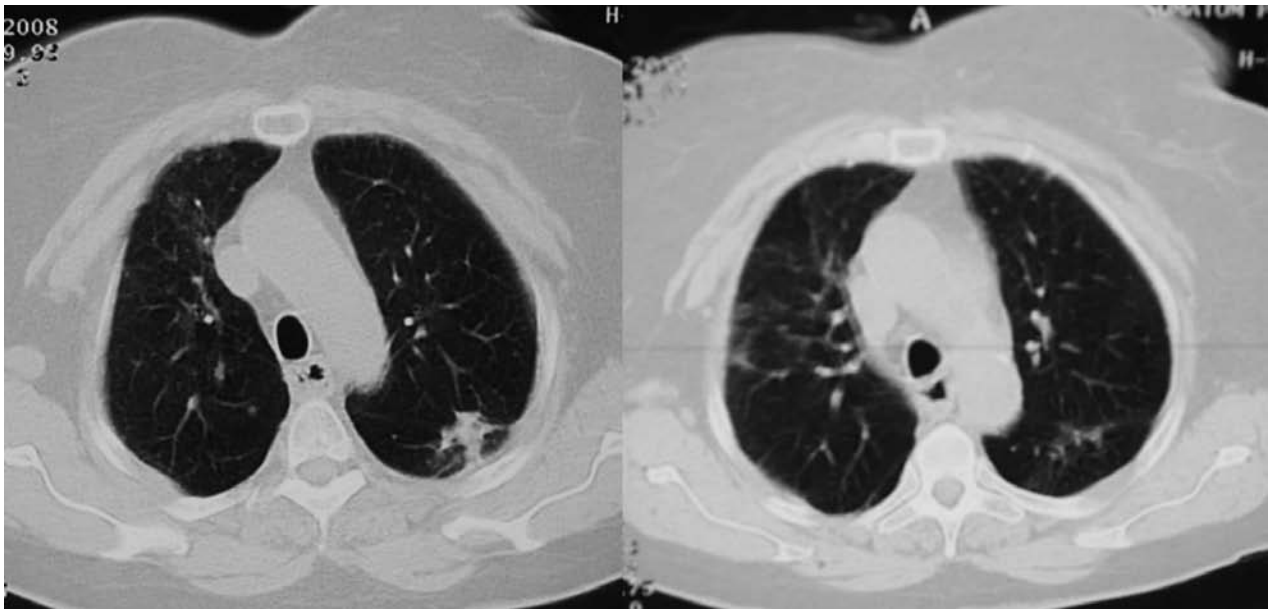

Fig. 4.

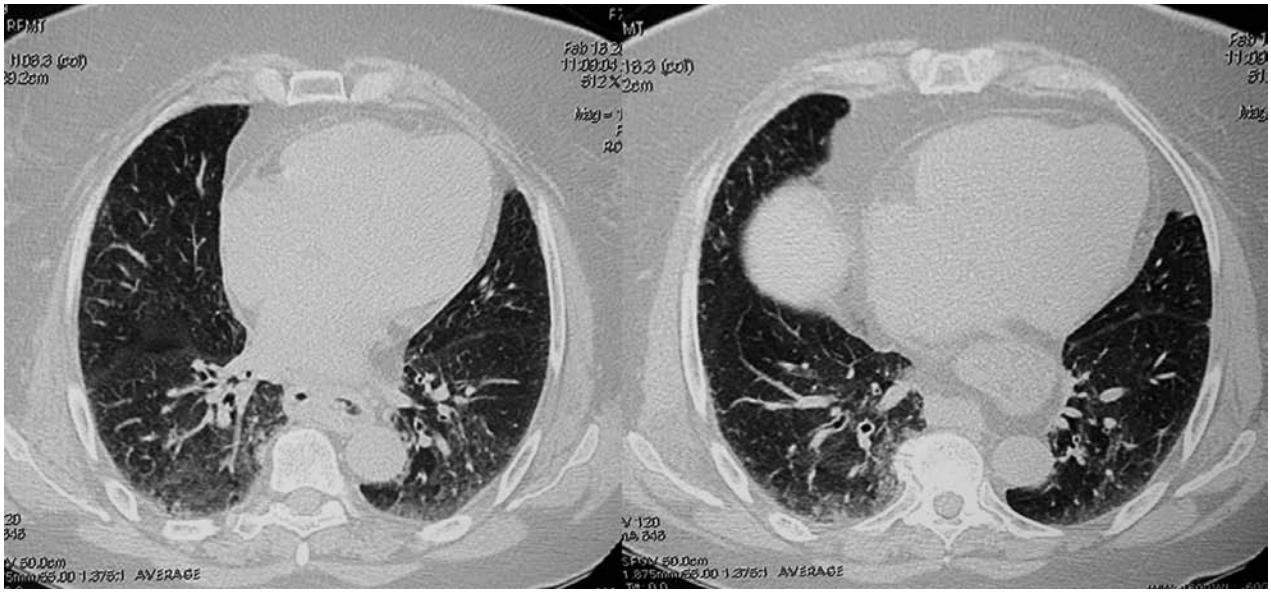

Fig. 5.

Discussion

Sarcoidosis, as reported at the World Congress in Kyoto in 1991, is defined as a multisystem disorder of unknown etiology which usually affects young and middle-aged individuals and presents in most cases with bilateral hilar lymphadenopathy, pulmonary infiltration, ocular and skin lesions. Its pathophysiology revolves around immunologic disorders involving many players of the immune response. An acute onset of the disease with asymptomatic thoracic lesions usually heralds a self-limiting course, whereas an insidious onset, especially with multiple extra-pulmonary involvement, may be followed by relentless

progressive fibrosis of the lungs and other organs (2). That said, the disease may shape into a pseudotumoral form featuring a 'cannon ball' aspect on radiography or computed tomography with an estimated prevalence of 2.4-4% as reported by Hansell (3). As metastatic extrathoracic neoplasms are the main concern to the physician, the importance of histopathology is emphasized within the diagnostic investigations. In our patient, the histological findings were not as conclusive as we expected initially but the diagnosis was somehow made as the course of the disease bolstered our suspicion. Published data suggest that this rare form of sarcoidosis is rather the prerogative of a much younger age group (20– 40 years), African-American ethnicity and female gender (4). This is in contrast with primary pulmonary neoplasms which tend to occur more frequently in males and older age groups. The case reported here stands out of the crowd as our patient is a 72-year-old woman of Caucasian ethnicity. Patients usually report mild general symptoms (such as fever, fatigue…) and clinical examination is poor. Biology shows a slight inflammatory syndrome and angiotensin-converting enzyme levels are usually normal. This form of sarcoidosis belongs - according to Abehsera et al. - to either the linear or the distorted pattern which are associated with the least functional impairment (5). Confluent sarcoid granulomas shape radiologically into nodules formingill-defined opacities with a diameter ranging from 1 to 4 cm usually with a perihilar distribution (1). The 'galaxy sign', initially described as the 'sarcoid galaxy' and representing coalescent granulomas can also be seen in tuberculosis where small satellite nodules are seen to border the periphery of the larger nodules (6). The appearance of a central core with peripheral nodules is reminiscent of a globular cluster galaxy. Central cavitation occurs in12.5% of cases (7) and the lesions may be surrounded by ground-glass opacity. The latter is thought to represent extensive interstitial sarcoid granulomas below the resolution of CT scan rather than alveolitis or haemorrhage (8). Resolution of the nodules, either spontaneously or with corticosteroid treatment, has been largely reported. According to Romer (9), this favorable outcomeis observed within 1 to 2 years which points up how much this case is unique by its swift course (only three months). Corticosteroids are the corner stone of therapy, but methotrexate has recently received significant attention as either a corticosteroid alternative or a corticosteroid-sparing agent (10).

Conclusion

Discarding malignancy in pseudotumoral sarcoidosis is a challenge. Thoughtful reasoning along with meticulous histopathological investigation is a milestone in the diagnostic approach. Further efforts in understanding sarcoidosis pathophysiology may lighten the investigations and dampen thoughts of malignancy earlier in such presentations of the disease. Fortunately, treatment is accessible and prognosis is fair.

References

1. Criado E, Sanchez M, Ramirez J, et al. Pulmonary sarcoidosis: typical and atypical manifestations at high-resolution CT with pathologic correlation. Radiographics 2010; 30(6): 1567-1586.
2. Yamamoto M, Sharma OP, Hosoda Y. Special report: the 1991 descriptive definition of sarcoidosis. Sarcoidosis 1992; 9: 33-34.
3. Hansell DM AP, Lynch DA, Mc Adams HP. Miscellaneous lung diseases: sarcoidosis. In Imaging of diseases of the chest. 4th edition. Philadelphia: Mosby; 2005.
4. Malaisamy S, Dalal B, Bimenyuy C, Soubani AO. The clinical and radiologic features of nodular pulmonary sarcoidosis. Lung 2009; 187(1): 9-15.
5. Abehsera M, Valeyre D, Grenier P, Jaillet H, Battesti JP, Brauner MW. Sarcoidosis with pulmonary fibrosis: CT patterns and correlation with pulmonary function. Am J Roentgenol 2000; 174(6): 1751-1757.
6. Heo JN, Choi YW, Jeon SC, et al. Pulmonary tuberculosis: another disease showing clusters of small nodules. Am J Roentgenol.2005; 184 (2): 639-42.
7. Freundlich IM, Libshitz HI, Glassman LM, Israel HL. Sarcoidosis. Typical and atypical thoracic manifestations and complications. Clin Radiol 1970 Oct; 21(4): 376-83.
8. Nakatsu M, Hatabu H, Morikawa K, et al. Large coalescent parenchymal nodules in pulmonary sarcoidosis: "sarcoid galaxy" sign. Am J Roentgenol 2002; 178 (6): 1389-93.
9. Romer FK. Sarcoidosis with large nodular lesions simulating pulmonary metastases. An analysis of 126 cases of intrathoracic sarcoidosis. Scand J Respir Dis 1977; 58(1): 11-6.
10. Baughman RP, Lower EE. Medical therapy of Sarcoidosis. Semin Respir Crit Care Med 2014; 35(03): 391-406.