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CASE REPORT

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Atypical Expression of Rare Disease Manifested with Cavitary Lung Nodules

Expresión atípica de enfermedad poco frecuente manifestada con nódulos pulmonares cavitados

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ABSTRACT

Lung nodules can vary in nature: solid, ground glass, granulomatous or non-granulomatous, cavitary or non-cavitary. They are a common finding in chest imaging when the patient comes to the consultation. According to their characteristics and the patient's history (medical history, physical examination, and laboratory results), they guide us towards different etiologies: infectious, autoimmune, or oncological. However, many times it is a great clinical challenge.

We present the case of a young patient with no previous medical history who came to the consultation with a cough of 3 months of evolution. The chest CT showed left mediastinal mass displacing the trachea and bilateral cavitary lung nodules. After ruling out more probable etiologies based on frequency for the patient's age and clinical manifestation, the diagnosis was: classic nodular sclerosing Hodgkin lymphoma with extranodal involvement. A very rare disease, and in our case, with an atypical form of presentation.

Key words: Lung nodules; Etiological study; Hodgkin lymphoma

RESUMEN

Los nódulos pulmonares pueden ser variados: sólidos, en vidrio esmerilado; granulomatosos o no granulomatosos; cavitados o no cavitado. Son un hallazgo frecuente en las imágenes de tórax cuando el paciente llega a la consulta. Según sus características y en concordancia con los antecedentes del paciente (historia clínica, exámen físico y resultados de laboratorio) nos orienta a las distintas etiologías: infeccioso, autoinmune u oncológico. Sin embargo, muchas veces es un gran desafío clínico.

Se presenta el caso de una paciente joven sin antecedentes que consulta por tos de 3 meses de evolución. En TC de tórax se evidencia masa mediastinal izquierda que desplaza tráquea y nódulos pulmonares bilaterales cavitados. Luego de descartarse etiologías más probables por frecuencia para su edad y forma de manifestación clínica, se arriba al diagnóstico de Linfoma de Hodgkin clásico Esclero Nodular con compromiso extranodal. Enfermedad poco frecuente y en nuestro caso, modo de presentación atípica.

Palabras claves: Nódulos pulmonares; Estudio etiológico; Linfoma de Hodgki

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INTRODUCTION

Lung nodules are a common finding in chest imaging when the patient comes to the consultation. Basing on their morphological characteristics, size, distribution, and in line with the patient's clinical condition, they usually guide us to consider the most likely cause, and with the help of supplementary studies, we can confirm or discard the diagnostic hypothesis.

The objective of this case is to review the study of lung nodules and their causes; in this particular clinical report due to the atypical manifestation of a rare disease.

CASE REPORT

25-year-old female patient with no relevant medical history (neither personal nor family). The patient presents with persistent cough with occasional whitish expectoration; subsequently she starts to have dyspnea mMRC 2, isolated fever episodes and multiple consultations at the on-call service, where she is indicated bronchodilator treatment. One month before consultation she started having night sweating, and 24 hours before consultation at the on-call service she showed hemoptoic expectoration. The patient claims she didn't lose weight.

She enters the emergency service in a normotensive state, 97% room-air pulse oximetry, no fever, good general condition. Good ventilatory mechanics, bilateral rhonchus and laryngeal rumor. She had palpable axillary and supraclavicular nodes.

Blood tests: HCT (hematocrit) (%) 27%, Hb (hemoglobin): 8.6 g/dl, Leukocytes: 18.600, Neutrophils: 83%. Lymphocytes: 9%, Platelets: 401.000, RIN: 1.52, LDH (lactate dehydrogenase): 314 U/L, CRP: C Reactive protein (polymerase chain reaction): 164 mg/L, GSR (globular sedimentation rate) 140 mm/H. HIV (human immunodeficiency virus): non-reactive.

Chest CT without endovenous contrast (Fig. 1 & 2): at the level of the sternoclavicular articulation, mass on the left side displacing the trachea towards contralateral, appears to infiltrate tracheal wall with reduction of lumen of at least 50%. Mediastinal nodes increased in size at the retrocaval-pretracheal, paratracheal, perihilliary and subcarinal levels, without reaching the adenomegalic range. Multiple node-like bilateral images with increased attenuation in the pulmonary parenchyma, of various sizes, with diameter greater than 10 mm, some of them cavitary, associated with perilesional ground glass opacity.

SPIROMETRY: FEV1/FVC (forced expiratory volume in the first second/forced vital capacity): 0.38 - FEV1: 34% (1.20 L) - FVC: 75% (3.14 L) Flow-volume curve: the expiratory phase shows curve flattening compatible with intrathoracic collapse; inspiratory phase remains unaltered.

Interpreted as tracheal stenosis associated with cavitary lung nodules under study.

For this situation, we suggest three diagnosis-related groups: First: infectious, and due to the prevalence and form of presentation, Tuberculosis, without discarding other common bacterial microorganisms. Secondly: autoimmune, granulomatous vasculitis with polyangiitis. Thirdly: primary or metastatic cancer.

Within the first 24 hours after the patient had been admitted, a bronchoscopy was performed, showing stenosis at the distal third of the trachea by extrinsic compression of left lateral wall infiltrating the entire trachea in a 2 cm path. We performed mechanical resection, dilation, and biopsy and placed a silicone tracheal stent.

Supplementary tests were done according to diagnostic suspicions. Sputum culture for common germs, mycological test, and BAAR (acid-alcohol-resistant bacillus): all negative. Blood test: negative. Koch culture: negative. Immunologic tests: FAN, Anti DNA, PR3 and MPO: negative.

Anatomopathological results of tracheal mucosa: classic nodular sclerosing Hodgkin lymphoma, with extranodal involvement (stage IV).

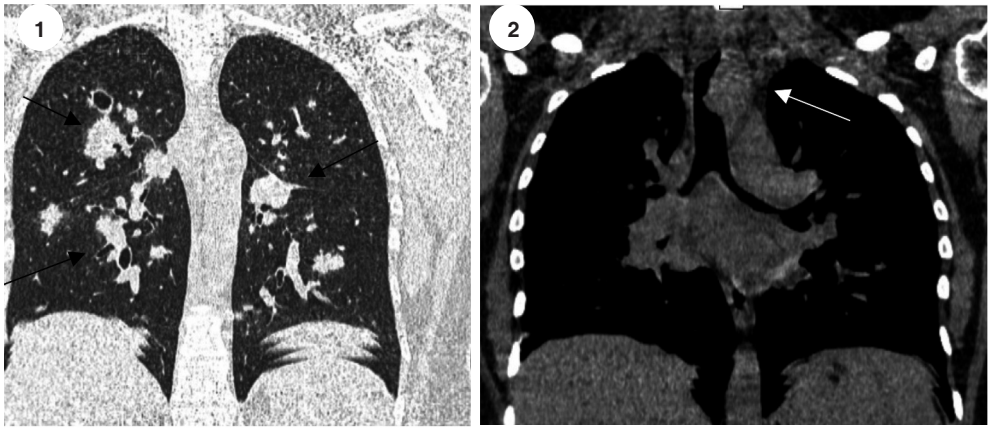

Figure 1 and 2. The black arrows show images with increased attenuation of random distribution, some of them cavitary, of various sizes. The white arrow shows mediastinal mass stenosing and displacing the trachea towards contralateral side

DISCUSSION

The finding of multiple lung nodules, some cavitary, and mediastinal nodes, in young woman without known clinical personal or familiar history makes us think of an infectious cause in the first instance (1-2): bacterial, such as Staphylococcus aureus, Nocardia, Haemophylus influenzae; fungal, such as Aspergillus; and mycobacteria such as Mycobacterium tuberculosis. However, due to the absence of isolates from sputum and blood cultures, and the chronicity of the patient's clinical symptoms, this etiologic suspicion is less probable.

Among the autoimmune causes (1-3-4), the granulomatosis with polyangiitis, necrotizing vasculitis, of very low incidence (5-10/millions of inhabitants) is presented with pulmonary involvement in more than 90% of the cases. It is shown in the chest CT with bilateral cavitary nodules in 50% of the cases. In advanced stages of the disease, the subglottic involvement of the trachea can occur, generating stenosis. The antineutrophil cytoplasmic antibodies (ANCAs) with cytoplasmatic pattern tend to be elevated in 90% of the cases.

Oncologic causes (2): the presence of multiple solid nodular images, some cavitary, of various sizes suggests metastatic lesions. In the particular case of our patient, thyroid, breast and germ cell cancer or lymphoma are the most probable types. Those types of cancer have low prevalence, but if the infectious cause is discarded, the probabilities of a non-infectious cause increase, among them the neoplastic cause._

Thyroid cancer (5) accounts for 1-2% of all types of cancer; in 50% of the cases, they show distant metastasis at the moment of the diagnosis and the lung is the involved organ in 50% of the cases. Malignant breast and germ cell tumors (2) also tend to metastasize to the lungs.

With regard to the Hodgkin lymphoma (6-7-8), it accounts for 0.5% of malignant neoplasms in adults, with an incidence of 2.5 cases every 100,000 inhabitants. The nodular sclerosing form is the most common sub-type, accounting for 2/3 of all the classic variants of lymphoma, with a high survival rate when diagnosed in early stages of the disease. It is mostly presented with intrathoracic involvement, mainly the superior mediastinum, due to the presence of adenopathies that sometimes generate compression of mediastinal organs, thus causing dyspnea, dysphagia or dysphonia, depending on the organ that is involved: the trachea, esophagus or recurrent laryngeal nerve. Lung involvement in lymphoma usually appears throughout the course of the disease and represents failure of response to the oncospecific treatment. As regards the involvement of the pulmonary parenchyma, it is usually manifested through three radiological patterns: unique consolidative mass that many times makes us suspect of an infectious process; due to contiguity from hilar lymph node involvement, or pulmonary nodules. The exception is the presence of cavitary nodules described in less than 1% of the cases post-chemotherapy. At the time of the diagnosis, it is unusual and has been observed in patients younger than 30 years. In those cases, it is extremely important to discard other diseases that also show this type of pulmonary lesions, basing on the patient's inquiry and physical examination.

It is a rare disease of atypical presentation that makes us think and discard multiple differential diagnoses.

Conflict of interest

The authors declare that there is no conflict of interest.

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