Pulmonary Malakoplakia Associated with Peripheral Cysts in an Immunocompetent Patient: A Case Report

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Pulmonary malakoplakia is a rare lung lesion more frequently found in immunocompromised patients than in immunocompetent individuals. In this study, we report the challenging case of a young immunocompetent patient with an irregular pulmonary nodule with peripheral cysts who, after undergoing surgery, was diagnosed with malakoplakia. Due to the rarity of the disease and the similarity of this condition to malignant neoplasms, cytopathological or histopathological examinations are necessary for the correct diagnosis. A description of pulmonary malakoplakia with peripheral cysts has not been previously published in the literature.

Keywords: Malakoplakia, Immunocompetence, Granulomatous disease, Lung neoplasm, Pulmonary neoplasms, Case report

Case report

A 22-year-old, previously healthy female patient was consulted in the emergency room after a headache and malaise in April 2021. She denied other complaints or flu-like symptoms. She underwent laboratory tests, which showed no evidence of infection (normal leukogram and inflammatory tests), negative findings on a polymerase chain reaction test for severe acute respiratory syndrome coronavirus 2, and negative serology. She underwent high-resolution computed tomography (HRCT) of the skull, which showed no abnormalities, and HRCT of the chest, with the finding of an irregular 16-mm pulmonary nodule in the left upper lobe with peripheral cysts (Fig. 1A, B). The patient was medicated only with analgesics, and upon improvement of symptoms was instructed to receive medical follow-up due to the incidental finding of lung injury. The patient remained without symptoms or complaints, but underwent a follow-up HRCT scan 3 months later. This scan showed an unaltered lesion, and surgery was indicated.

Fig. 1. Coronal (A) and axial (B) sections demonstrating a solid nodule with spiculated margins located in the medullary region of the apico-posterior segment of the left upper lobe, with peripheral cysts (arrow).
Pleuroscopy, left segmentectomy, and biopsy of mediastinal and pulmonary lymph nodes were performed with histological analysis of the lesion in an intraoperative frozen section of a possible undifferentiated malignant neoplasm. After 1 week, the definitive histopathological results showed a poorly delimited, yellowish-brown, and firm lesion, macroscopically measuring 1.5×1.0×0.8 cm (Fig. 2A). On microscopy, the lesion showed Michaelis-Gutmann bodies, compatible with malakoplakia (Fig. 2B). The material was sent for culture, with no evidence of any isolated germs.

This study was approved by the Institutional Review Board of Irmandade Santa Casa de Misericordia de Porto Alegre (IRB approval no., 5.560.949), and the patient provided written informed consent for the publication of her clinical details and images.

Discussion

The term “malakoplakia” is derived from the Greek malakos (soft) and plakos (plaque) due to the macroscopic characteristics of this disease [1-3]. The pathophysiological mechanism remains unclear, but it may be associated with defective lysosomal activity [2-4]. Inadequate phagocytosis is thought to occur due to low levels of cyclic guanine monophosphate phosphatase causing defective microtubule function, which impairs lysosomal degradation and results in decreased microbial death [1,3,5].

Macroscopically, malakoplakia presents as a yellowish-brown lesion, and microscopically, it is characterized by Michaelis-Gutmann bodies—foamy macrophages showing basophilic intracytoplasmic inclusions, which undergo mineralization by subsequent deposition of calcium and iron associated with lysosome calcification [1,2,4,6].

The genitourinary tract is the site most commonly affected by malakoplakia, but other organs and systems may also be involved, including the lung in relatively rare cases [2,3,7]. Pulmonary lesions can include nodules, masses, and cavitations and can also affect the trachea and pleura [6]. In the lung, malakoplakia has been associated mainly with Rhodococcus equi, an opportunistic pathogen that mainly infects immunosuppressed patients [1-3,5,6,8].

Pulmonary malakoplakia is possibly associated with defective macrophage activity, which leads to the accumulation of granulomatous masses, generating lesions that can be confused with tuberculosis, primary malignancy, or metastatic disease [2-6]. The symptomatology is nonspecific and depends on the extent and type of involvement of the lesions [9]. Furthermore, clinically, the disease is difficult to diagnose, which leads to underdiagnosis [2], and without an experienced pathologist, histological findings can be misinterpreted [5].

Malakoplakia occurs predominantly in adults, and when it occurs outside the urinary tract (where women are more affected), there is no sexual preponderance [5]. Most affected patients present immunosuppression either due to transplantation or human immunodeficiency virus infection [1-3,7]. Patients with decompensated diabetes mellitus [1-3,6] and alcoholism are also more likely to be affected [1,3].

Pulmonary malakoplakia is a rare diagnosis, with few cases described in the literature; it is even less common in immunocompetent patients. Based on an extensive laboratory investigation, our patient did not have immunosuppression or present evidence of concomitant infection. Temporary forms of immunosuppression of various origins (such as post-infectious) may explain the occurrence of such lesions in immunocompetent individuals [3].

Our patient had an irregular lesion with peripheral cysts, a finding not yet described in the literature, according to which there is an apparent predominance of cavitated lesions in pulmonary malakoplakia. Our hypothesis is that a previous infection by an unidentified germ resulted in local necrosis, with consequent formation of perilesional cysts due to scarring.

The rarity of this disease and its similarity with malignant neoplasms makes it necessary to perform a cytopathological or histopathological diagnosis, but the differential diagnosis of malakoplakia should be considered for young patients and those without risk factors for cancer to avoid unnecessary invasive measures. In this context, the presence of perilesional cysts may serve as an indication of pulmonary malakoplakia in future patients, helping clinicians to avoid surgery or more aggressive forms of diagnosis.
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Conflict of interest

No potential conflict of interest relevant to this article was reported.

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