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Primary Pulmonary Mucinous Cystadenocarcinoma: Presentation of a Case and a Review of the Literature

Cistoadenocarcinoma gigante mucinoso primario de pulmón: presentación de un caso y revisión de la literatura

To the Editor:

Pulmonary mucinous tumors are an infrequent entity, and the malignant presentations (cystadenocarcinoma and mucoepidermoid) are more frequent than the benign forms (cystadenoma or mucus gland adenoma),² presenting in patients near their fifties, with no clear difference between sexes.² There are only 20 cases reported of the cystadenocarcinoma subtype in the literature.

We present the case of a 67-year-old woman with moderate COPD, smoker of 30 pack-years. After treatment of what seemed to be a respiratory infection, thoracic computed tomography (CT) revealed a solid mass in the right upper lobe (RUL) that was 3.6 cm in diameter, with ipsilateral lymphadenopathies. A study was carried out with bronchoscopy, biopsy and PET, with no conclusive results. No surgical treatment was carried out at this time.

Afterwards, the patient did not attend periodic follow-up visits, and remained asymptomatic. Six years later, she presented with constitutional and respiratory infection symptoms. A new study was initiated of the lung mass with CT, revealing complete occupation of the RUL, persisting lymphadenopathies, mediastinal invasion and pleural effusion (fig. 1). FNAC demonstrated abundant mucinous material; two bronchoscopies showed evidence of complete stenosis of RUL and the bronchial biopsies were reported to be non-atypical bronchial mucus with a slight increase of the seromucous glands, with BAL and bronchial brushing negative for malignancy. Thick-needle lung biopsy showed fibrous tissue with no evidence of tumor. PET gave evidence of peripheral pathological captation of the mass and the lymphadenopathies, with no affectation at other levels. Echoendoscopy and mediastinoscopy were carried out, finding necrotizing granulomas with no signs of malignancy. Given the high suspicion for neoplasm and the lack of diagnostic confirmation, thoracotomy was performed and intraoperative biopsies taken until sufficient tumor tissue was visualized for the diagnosis, confirming the unresectability of the mass. The definitive diagnosis was mucinous primary pulmonary cystadenocarcinoma. The patient died six months after having received chemotherapy.

The spectrum of this type of neoplasm is extensive, having been described with multiple denominations.^{1,2} Perhaps the simplest classification is to group these tumors in accordance with the WHO into three entities: mucinous cystadenoma (localized cystic mass, full of mucus and surrounded by well-defined columnar mucinous epithelial wall), mucinous cystic tumor with atypia (invasive growth of the subjacent tissue with significant atypia and marked pseudo-stratification) and mucinous cystadenocarcinoma (cystic adenocarcinoma with abundant production of mucus that follows a pattern similar to that described in ovarian, breast and pancreatic tumors).¹ In most cases, the findings are incidental, asymptomatic and with no compromised lung function.³ They are generally slow-developing, although cases have been described of progression, and even of metastasis and recurrences.^{1,2} Radiography and thoracic CT are essential for the diagnosis.² Their confirmation is made by means of pathologic anatomy.⁵ The differential diagnosis is extensive, including bronchogenic cyst, mucus gland adenoma, mucoepidermoid carcinoma, mucinous bronchoalveolar carcinoma or mucinous adenocarcinoma metastasis. Treatment is based on complete resection of the tumor. Radiotherapy and chemotherapy seem to play a limited role in non-resectable tumors. The general prognosis is good,^{4,5} with an approximate survival rate of 75% at 5 years and 50% at 10 years.



Figure 1. CT showing complete occupation of the RUL, persistent lymphadenopathies, mediastinal invasion and pleural effusion.

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World Spirometry Day: Experience in a Hospital Centre

Día mundial de la espirometría: experiencia en un centro hospitalario

To the Editor:

World Spirometry Day was celebrated on 14 October 2010, and was a world-wide event promoted by the *European Respiratory Society* (ERS) through its organism the *European Lung Foundation* (ELF).

The objective of this event was for the general public, professionals and authorities to learn more about the most widely used diagnostic tool among pulmonologists: spirometry. Another aim was to increase consciousness about the importance of lung diseases and the preventative measures for avoiding its appearance and/or evolution.

Among the pneumological societies, SEPAR-ALAT performed spirometries at the hospital centers in the public who attended.

From the Hospital de la Santa Creu i Sant Pau in Barcelona, we would like to share our experience with this event at our center. On the day mentioned, from 9am to 2pm, spirometries were carried out on the general public, in accordance with SEPAR procedures.¹ The event was run by 10 nurses with experience in lung function techniques (two per hour), an adjunct doctor and a resident doctor from the Pulmonology Department. The following material was used: two spirometers (Datospir 600, Sibelmed, Barcelona, Spain), stadiometer, scale, disposable mouthpieces, tissues and antiseptic hand gel, while informational material about spirometries² and the dangerous effects of smoking,³ translated in Spanish, were distributed. Prior to the test, a 12-question survey⁴ was administered, as proposed by the ERS and ELF and translated into Spanish, in order to obtain more information on the people interested in doing the test. The physicians in charge of the clinic completed spirometry reports, and those patients' whose results were altered were remitted to their Primary Attention physician with a letter reporting on the degree of alteration. All smokers were given minimal anti-tobacco advice as well as the informational material that was available.³

The media were informed of the event with the aim of their helping to spread knowledge about spirometry among the general

public, as well as the importance of early detection and prevention of some of the most prevalent lung diseases.

Seventy-five spirometries were carried out. Mean (standard deviation) age of the participants was 56.3 (17.8) and forced expiratory volume in one second (FEV₁) was 2.58 (0.92) L [88.3 (20.8)%]. There was a slight predominance of women (n = 39, 52%). The percentage of smokers or ex-smokers was 49%. Obstructive pathology was observed (FEV₁/FVC < 70%) in 20 of the subjects participating (27%), 6 of whom (30%) had never been tested with spirometry before. Out of the 32 active smokers, 8 (25%) presented airway obstruction. Out of all the participants, 29 (39%) manifested having performed the test before, and only 33 (44%) stated having knowledge of the test.

This event had very good acceptance and participation amongst the population, and it allowed us to detect some cases of spirometric alterations that were previously unknown.

The degree of knowledge of this test is still quite low in the general public. It is therefore recommendable to organize future editions of World Spirometry Day in order to share information and spread understanding of this test, as well as possibly detecting individuals with respiratory diseases early on and therefore be able to take categorical steps in controlling risk factors and trigger mechanisms.

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