

Searching for the Primary Source: Syringocystadenocarcinoma as Differential Diagnosis of Metastatic Lung Adenocarcinoma

Andres Rivera*, Gorka Ibarra and Ignacio Capdevila

Department of Plastic surgery, Gregorio Marañón General University Hospital, Spain

*Corresponding author: Andrés Rivera Muñoz, Department of Plastic surgery, Gregorio Marañón General University Hospital, Merida Street, 22, 1st, Madrid Postal code: 28030, Spain



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ABSTRACT

Skin adnexal carcinomas are a heterogeneous group of rare malignancies with hard differential diagnosis against skin adenocarcinoma metastasis. In many cases the primary origin of the lesion is unclear, making it difficult to decide on the best treatment. The aim of the present report is to set out the key points of pathological analysis and tumor evolution for appropriate differential diagnosis. Here we present the case of a 64 year-old patient, a smoker with a scalp tumor informed as syringocystadenocarcinoma papilliferum (SCACP) that developed a second cutaneous lesion, cervical adenopathies and a solitary pulmonary nodule, which was almost indistinguishable from a primary lung adenocarcinoma in simple anatomopathological analysis. The combined analysis of the various lesions finally made an adequate differential diagnosis possible. Literature about SCACP and its differential diagnosis with lung adenocarcinoma is reviewed and discussed.

Keywords: Case Report; Diagnosis; Differential; Carcinoma; Skin Appendage; Adenocarcinoma of Lung; Skin Neoplasms

Introduction

Adnexal malignant tumors are very rare lesions, with an incidence of 5.1 cases per million people per year [1] and tend to be underestimated in the clinical diagnosis because they are generally not suspected [2]. In this regard, syringocystadenocarcinoma papilliferum (SCACP) is a little-known skin apocrine carcinoma whose diagnosis is based on pathological analysis [3,4]. Only 43 cases have been described worldwide. Lung carcinoma is one of the most prevalent neoplasms in the general population, with an incidence rate of 62 per 100,000 people per year, being the leading cause of cancer mortality [5]. Cutaneous metastases from primary pulmonary cancers are the first sign of extranodal disease in 1% of patients with lung carcinoma [6]. SCACP is a low grade skin adnexal adenocarcinoma [1,2] that rarely presents both regional or

distant metastases. The appearance of these metastases is similar to other glandular carcinomas, presenting a diagnostic challenge where most frequent adeno-carcinoma (lung, breast or colon) may be considered as the primary source, misleading the real diagnosis [7]. We present the first case of SCACP with a pulmonary node in its clinical course, which pretends a primary metastatic lung adenocarcinoma. Clues for histopathological and clinical differential diagnosis of these lesions are examined.

Case Report

A 64 year old male patient with smoking habits presented a nodular subcutaneous tumor on the parietal region of the scalp with an accelerated growth during the last 3 months. It showed cutaneous telangiectasias with no skin ulceration. Patient reported a previous

multinodular plaque lesion in the same location lasting more than 10 years. There were no other symptoms. For appropriate analysis, it was treated by wide excision with 1 cm margins and two scalp flaps for coverage (Figure 1). Pathological examination showed a nodular lesion of about 2 cm affecting the entire dermis; consisting of a glandular and tubular pattern with pseudopapillae and papillae, which exhibited a perilesional permeative pattern, signs of

perineural infiltration and a high rate of mitosis. Immunoreactivity was positive for p63/CK7/34-betae12/EMA, being negative for CEA/S100 (Figure 2). The previous description corresponded to a SCACP that respected surgical margins of resection arising on a preexisting nevus sebaceous of Jadassohn. Four months after surgery, the patient consulted because of a painful nodule on the left scapular region and marked constitutional syndrome.



Figure 1: Syringocystadenocarcinoma papiliferum:

- A. Scalp lesion at surgery room and
- B. Second month postoperative aspect.

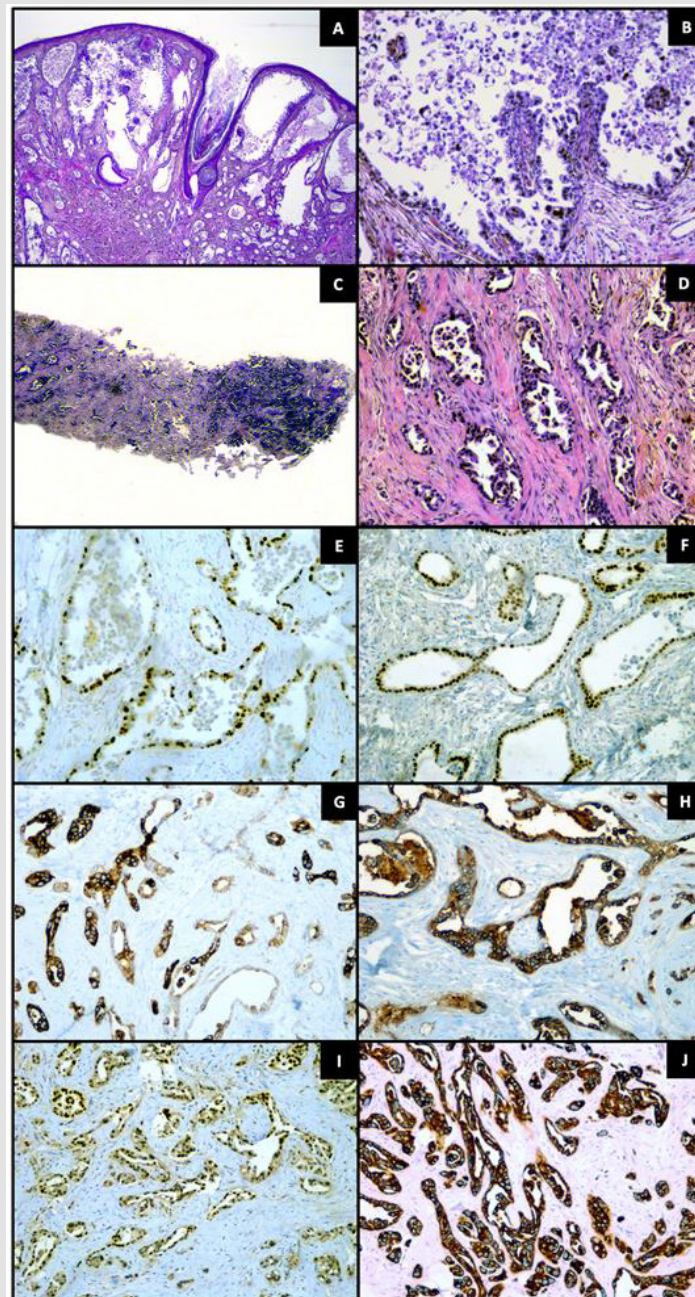


Figure 2: Syringocytadenocarcinoma:

- A. Primary scalp lesion (H/E) [100x],
- B. [400x] and
- C. Scapular lesion [100x],
- D. [400x].
- E. Immunohistochemic stain of p63,
- F. TTF1,
- G. CK-7,
- H. EMA
- I. [400x] of scalp lesion and immunohistochemic stain of TTF1
- J. CK7 [400x] at scapular lesion.

Complementary image studies showed concomitant cervical level IV adenopathies, multifocal skeletal affectation, bilateral hilar adenopathies and a solitary pulmonary nodule (Figure 3). Incisional biopsy of the scapular lesion exhibited an unspecific adenocarcinoma pattern with immunoreactivity for CK7, TTF-1 and negativity for CK20/CDX2/PSA/PAX8. Since differential diagnosis of metastatic lung adenocarcinoma was considered it was decided to perform a CT-guided core needle biopsy of the pulmonary nodule. This time, histopathological analysis showed a moderately

differentiated glandular malignancy focally positive for TTF-1, which was not specific for adnexal tumors nor for lung adenocarcinoma. In order to elucidate diagnosis, the first pathological sample (scalp) was reviewed. Because of the permeative and gradual malignancy pattern observed there, as well as its arising over a preexisting nevus sebaceous of Jadassohn, lung adenocarcinoma was rejected as the primary origin of the clinical chart. The patient refused adjuvant treatment and died eight months after surgical excision.



Figure 3: Syringocystadenocarcinoma metastasis: Computerized tomography that shows nodular 2,4 cms-leght pulmonary lesion at fourth month of intervention.

Discussion

Primary lung neoplasms constitute 13% of all detected cancers every year. Among 90% of the cases are supposed to be tobacco related, especially in adenocarcinomas [8]. At time of diagnosis 55% of total pulmonary neoplasms are staged as metastatic disease [6] nearly 1% of these metastases will be cutaneous. On the other hand, skin adnexal malignancies are very scarce tumors, particularly SCACP, with 43 cases reported worldwide [7]. Mortality is usually low in located lesions with a 5-year survival rate of 99% [1]. Since they are little known, they tend to be under-estimated in clinical practice, which occasionally can lead to regional or distant metastases at debut, rising mortality rates to 43%. Management of these lesions usually implies local wide excision, even though Mohs' surgery has shown the same effectiveness. There is little evidence regarding their response to systemic oncological treatments [2,4]. SCACP is diagnosed mainly by histological sampling, in which a nonspecific glandular pattern is observed. The malignant component of this entity is variable and comprises SCACP in situ

as well as invasive adenocarcinoma or invasive cell squamous carcinoma.

Complementary immunochemical studies are usually performed for differential diagnosis and proper identification between skin metastasis, especially when poorly differentiated or anaplastic. Expression of p63 is associated with the invasive component of basal cells, whereas CK7 is usually expressed in the superficial epithelial cells. Positivity for p63 and CK20 negativity have been described as useful markers to differentiate malign adnexal tumors from metastatic lesions [9]. CK7 and TTF1 expressions are more characteristic from lung or thyroid tumor, but does not refuse the primary origin in our case, where morphology of the lesion was very well differentiated [6,9]. In addition to the previous, the progressive malign histological pattern observed can also aid the diagnosis as SCACP is thought to arise over a previous benign syringocystadenoma papilliferum, often in the sinus of a sebaceous nevus [10]. Here we present a complex clinical case of a malignant glandular tumor that poses, in the its temporal

evolution, a controversy in the differential diagnosis with a lung adenocarcinoma.

What makes this case interesting is the almost synchronous presence of the pulmonary lesion and the adnexal glandular neoplasia of the skin, both with similar histological characteristics. There are no cases described of metachronic presence of SCACP and lung carcinoma given the low incidence of this rare adnexal tumor; also, this is the first report of a SCACP coursing with a metastatic solitary pulmonary nodule. Currently, there is no standardized approach to evaluate patients affected by skin lesions that are compatible with either cutaneous metastasis or primary skin carcinomas. Therefore, several studies have emphasized the serious difficulties in elucidating the primary origin of the tumor when multiple lesions in different organs are present at diagnosis [4]. This aspect is very important with regard to the therapeutic approach especially since metastasis implies a worse prognosis. In the present report, the first lesion (scalp) exhibited progressive peripheral malignancy, a fact that practically excludes the possibility of being a metastatic lesion. Furthermore, an underlying sebaceous nevus support previous diagnosis.

In conclusion, structural description of the surgical piece in malign adnexal tumors allows a more precise identification of the tumor's source.

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