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Case Report

Dermatofibroma Metastasizing to the Lung: Current Treatment

Enrique Bermejo Casero,* David Pérez Alonso, Santiago Quevedo Losada, and Luis López Rivero

Servicio de Cirugía Torácica, Hospital Universitario Insular, Las Palmas de Gran Canaria, Las Palmas, Spain

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ABSTRACT

Dermatofibromas are very common skin tumors. Their typical presentation is as a slow-growing, firm, solitary papule. They have been described histopathologically as a reaction of the connective tissue of the skin or as a benign neoplasm. Cases of these tumors metastasizing to the lung were first reported in 1990. Since then 12 cases with similar characteristics have been reported worldwide. We present the case of a young woman with a recurrent dermatofibroma on the shoulder that metastasized to both lungs. We discuss the histologic characteristics of this tumor that could raise suspicion of unusually aggressive behavior.

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Dermatofibroma y metástasis pulmonares. Tratamiento actual

RESUMEN

El dermatofibroma es una lesión cutánea común en la práctica dermatológica. La manifestación típica es una pápula de crecimiento lento, firme y solitaria. Desde el punto de vista histopatológico, se define en ocasiones como una reacción del tejido conectivo dérmico y en otras como una proliferación neoplásica benigna. En 1990 se publicó por primera vez la existencia de metástasis pulmonares de este tumor. Desde entonces se han recogido en todo el mundo 12 casos de similares características. Presentamos el caso de una mujer joven con un dermatofibroma recidivante en el hombro y metástasis pulmonares bilaterales. Analizamos qué características histológicas del tumor podrían hacer sospechar un comportamiento posterior inusualmente agresivo.

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Introduction

Dermatofibromas are skin tumors that affect mainly women. They are generally located in the lower extremities and their typical presentation is as a slow-growing, solitary papule. Clinical variants include giant, atrophied, and polypoid tumors. Dermatofibromas usually have a benign behavior, though it is not unusual for them to recur even when the surgical borders are tumor-free. However, these tumors may metastasize to remote organs in rare instances; only 12 such cases in the world have been reported in the English-language literature.¹⁻⁸ The biological nature of dermatofibromas is not yet clear in histopathological terms. Some authors claim that they are

reactive tumors, which they describe as fibrous histiocytomas of the skin surrounded by collagen fibers and inflammatory components. Others consider them to be benign neoplasms because the tumors show high cellularity, nuclear pleomorphism, and even a high mitotic index.⁹

We present the case of a 37-year-old woman operated on for a recurrent dermatofibroma on the right shoulder. The chest radiograph showed the presence of a 3×4-cm nodule on each upper lobe. The wedge resection of these nodules using sequential bilateral thoracotomy confirmed that it was a metastasizing dermatofibroma. Surgery also revealed micronodular deposits with the same histologic characteristics as the larger tumors.

Case Description

A 37-year-old woman was referred from the Hospital de Fuerteventura after 2 pulmonary nodules were detected in the

* Corresponding author.

E-mail address: luiss175@hotmail.com (E. Bermejo Casero).

preoperative assessment of a skin tumor on the right shoulder. She worked as an insurance broker. The only medical history of interest was the surgical resection of a skin tumor in the right deltoid region in 2002. This tumor was described macroscopically in the pathology report as a raised, lobulated tumor consisting of 2 main adjacent nodules measuring $3 \times 3 \times 2$ and $2.5 \times 1.8 \times 1.5$ cm. The tumor occupied part of the skin tissue and on unaided visual inspection did not seem to affect the subcutaneous adipose tissue.

Histopathology revealed the presence of a fibrous histiocytoma surrounded by collagen fibers. The tumor was highly cellular, with a high mitotic index and nuclear pleomorphism. Immunohistochemical analysis showed the cells to be negative for CD34 (a marker for dermatofibrosarcoma) and for the S-100 protein. The pathologic diagnosis was dermatofibroma; the surgical borders were tumor-free.

Six years later, a new nodular lesion measuring 2.5 cm in diameter appeared on the scar left by the previous operation; the lesion was pigmented and had an elastic consistency (Figure. 1). The biopsy confirmed that this tumor was again a dermatofibroma. In the preoperative assessment for the resection of this tumor, the chest radiograph revealed pulmonary nodules on both upper lobes. Computed tomography (CT) of the thorax showed the presence of nodules measuring 2.5 cm and 2.6 cm in the right and left upper lobes, respectively. The presence of micronodules in both upper lobes was also reported.

The results of the basic laboratory workup were as follows: white cell count, 7300/mL (neutrophils, 76%; lymphocytes, 16.4%; monocytes, 6%); hemoglobin, 14.4 g/dl; and platelet count, 304 000. The coagulation tests, sedimentation rate, and biochemical workup were normal. The tumor markers also showed values within the normal range (carcinoembryonic antigen, 0.9 ng/mL; alpha-fetoprotein, 4.5 ng/mL).

No important lesions were found during examination by fiberoptic bronchoscopy. Pathologic analysis of the bronchial aspirates, brushings, and transbronchial biopsy material from the posterior apical segment of the left upper lobe showed no malignant cells. The results of microbiological tests of the bronchial aspirates were negative, showing only common microorganisms.

The lung function tests showed a forced expiratory volume in 1 second of 3020 mL (88%) and a forced vital capacity of 3470 mL (88%).

The case was presented to the lung tumor committee, and it was decided to complete the preoperative assessment of the patient with



Figure 1. Dermatofibroma recurring on the scar where an earlier tumor had been removed.

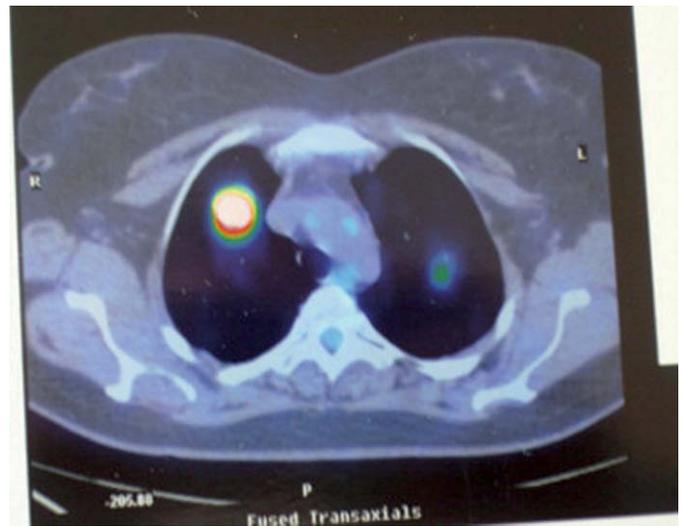


Figure 2. Positron emission tomography: abnormal uptake by tumors in the lung apices.

positron emission CT (Figure 2). In the deltoid muscles of the right shoulder the images revealed the presence of a subcutaneous oval lesion of 2.6 cm with pathologic hypermetabolism (maximum standardized uptake value [SUV], 2). In the lungs it revealed a 2.6-cm nodule in the right upper lobe, with abnormal hypermetabolism (maximum SUV, 11.3), and a 3-cm nodule in the left upper lobe, with abnormal uptake (maximum SUV, 7.4). The presence of micronodules was also detected in all the lobes, without uptake probably due to their small size.

Based on these data, a 2-stage surgical procedure was planned. In the first intervention the skin tumor was resected, and the defect was covered with a full-thickness skin graft taken from the abdominal region. Then, a right lateral thoracotomy was also performed. This procedure revealed a 2.5-cm cystic nodule in the right upper lobe and micronodules distributed throughout the lung. A wedge resection of the pulmonary nodule was performed and the micronodules were biopsied. Pathologic examination of the skin tumor, the pulmonary nodule, and the micronodules revealed similar features. The tumor was thus described as a metastasizing dermatofibroma with similar characteristics to those of the one resected 6 years earlier. At this point the suitability of treating the left hemithorax was considered. There is insufficient experience of such metastasizing tumors to offer clear guidelines. For some unknown reason, the biological behavior of the tumors in the lung apices was different from that of the rest: the metastases in this region grew faster and were more aggressive. Therefore, 1 month later, a wedge resection of the pulmonary nodule in the left apex was performed, confirming that there were also dozens of micronodules distributed throughout that lung. Both postoperative periods were without complications. Nine months later, the patient was still asymptomatic, and a follow-up CT scan of the chest showed that the micronodules had not varied in size and were practically imperceptible. No adjuvant therapy was prescribed, and the patient's quality of life was excellent.

Discussion

The 12 reported cases of dermatofibroma that subsequently metastasized to the lungs show certain clinical similarities: the initial skin tumor appeared when the patients were young and the lung tumors were only recognized after a long latency period.

The question of whether dermatofibromas are reactive or neoplastic lesions still generates debate today. Clinical differentiation is difficult. Tumors that do not involute spontaneously can easily be

assigned to the neoplastic group and vice versa. However, this simplistic view is unsatisfactory. Lesions with a high collagen content and the clearly inflammatory ones could be considered to be reactive. Tumors in this subgroup are characterized by low cellularity, a predominance of extracellular matrix, and fibrous histiocytosis. The neoplastic group is characterized by extensive cell proliferation, nuclear pleomorphism, and a high mitotic index. These were the pathologic characteristics detected in the initial tumor of our patient and they were seen once again in the recurrence. Gillou et al⁴ described a set of histologic characteristics of dermatofibroma that should be considered as risk factors for metastasization: large tumors, high cellularity, clear pleomorphism, a high mitotic index, tumoral necrosis, and local recurrence. Like ours, most cases of dermatofibroma metastasizing to the lungs showed these risk factors.

Pulmonary metastases of this tumor have generally been described as cystic lesions. In the differential diagnosis of metastatic pulmonary lesions, one must consider firstly metastases from a benign uterine leiomyoma or from uterine sarcoma.^{10,11} These tumors also affect middle-aged women and produce cystic nodules in the lung when they metastasize. Furthermore, they tend to have a similar hemorrhagic content to that of a chocolate cyst of the ovary.

The use of adjuvant chemotherapy after surgery has so far shown poor results. Ifosfamide followed by Adriamycin (doxorubicin) monotherapy have been used,⁵ but in no cases did they halt disease progression. However, we believe that as yet too few cases have been studied to draw conclusions regarding this practice.

Finally, it seems that some histologic characteristics of dermatofibroma can increase the threat of subsequent metastasis. These cases should probably receive closer monitoring with serial chest radiographs.

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