CASE REPORT

Pulmonary Sclerosing Hemangioma in a Patient With Cowden Syndrome

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Introduction

Cowden syndrome is a rare disease with an autosomal dominant inheritance pattern that usually manifests as benign hamartomas—mainly colonic polyps. It is therefore classified as a form of colonic polyposis. Pulmonary sclerosing hemangioma is an unusual lung neoplasm which typically presents as a solitary peripheral nodule in asymptomatic women. Although the histology of this entity is well defined, its origin and treatment is debated. One of the main diagnostic problems is to histologically differentiate a pulmonary sclerosing hemangioma from a papillary lung carcinoma.

Key words: Sclerosing hemangioma. Lung tumor. Cowden syndrome.

We describe the case of an 18-year-old female with Cowden syndrome in whom a simple x-ray detected a solitary pulmonary nodule that was identified as a sclerosing hemangioma. Pulmonary sclerosing hemangioma is an unusual lung neoplasm which typically presents as a solitary peripheral nodule in asymptomatic women. Although its histology is well defined, its histogenesis and treatment are the subject of debate. One of the main diagnostic problems is to histologically differentiate a pulmonary sclerosing hemangioma from a papillary lung carcinoma.

Hemangioma esclerosante pulmonar en un paciente con síndrome de Cowden

Describimos el caso de una paciente de 18 años con síndrome de Cowden, en quien una radiografía simple objetivó un nódulo pulmonary solitario que resultó ser un hemangioma esclerosante. El hemangioma esclerosante de pulmón es una neoplasia pulmonary poco frecuente, que suele presentarse como un nódulo solitario periférico en mujeres asintomáticas. En la actualidad, aunque la histología del hemangioma esclerosante está bien definida, son objeto de discusión la histogénesis de esta entidad y su tratamiento. Uno de los principales problemas diagnósticos es diferenciar histológicamente el hemangioma esclerosante del adenocarcinoma pulmonar con patrón papilar.

Palabras clave: Hemangioma esclerosante. Tumores de pulmón. Síndrome de Cowden.

Case Description

We report the case of an 18-year-old woman who was referred to our department with a solitary pulmonary nodule that had been detected by chance in a routine x-ray. The medical history included multiple nodular hyperplasia of the thyroids, high-flow venous vascular malformations in the distal third of the right thigh and back that had been removed surgically, and perirenal, retroauricular and supraclavicular lipomas that had also been treated surgically. She had also been diagnosed with an ovarian cyst and breast fibroadenomas. A simple chest x-ray performed as part of regular follow-up ordered by an endocrinologist revealed a solitary pulmonary nodule—approximately 2 cm in diameter—in the right upper lobe; the patient was consequently referred to the pneumology department.

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She had no history of substance abuse or allergies to medication, and had no fever, cough, expectoration, asthenia, anorexia, or weight loss. A physical examination revealed no enlargement of the adrenal glands. Cardiopulmonary auscultation was normal and analytical data (blood count, biochemistry, and coagulation factors) were normal except for signs of anemia. Immunological and tumor markers were within normal limits. Given the presence of the solitary pulmonary nodule, fiberoptic bronchoscopy was performed with bronchial aspiration and bronchoalveolar lavage, both of which were negative for tumor and inflammatory cells. Computed tomography (CT) revealed a solid nodule in the right upper lobe, approximately 2 cm in diameter and with lobulated edges; there also appeared to be cavitation around the edges, although no uptake of the intravenous contrast could be observed. Nor was lymph node involvement evident within a relevant distance from the nodule. A routine scan 3 months later confirmed no change in nodule size or shape (Figure 1).

CT-guided fine-needle aspiration was performed on the lung nodule; although results indicated epithelial proliferation with papillary formations and isolated atypia, there were no signs of malignancy. Given these inconclusive results, a right thoracotomy was performed to biopsy the nodule (Figure 2); the diagnosis was sclerosing hemangioma. Given the patient’s age and multiple malformations, a genetic study was ordered and Cowden syndrome was diagnosed. The patient was referred to the digestive system department in order to rule out colonic hamartomatous polyps.

No specific treatment was prescribed. The patient is currently being monitored and has presented no further complications related to her underlying disease.

Discussion

Sclerosing hemangioma—which is also referred to as benign sclerosing pneumocytoma, benign pulmonary histiocytoma, sclerosing angioma or xanthomatous pseudotumor—is a rare disease. Although it was initially described as a vascular tumor with cellular infiltration and sclerotic and hemorrhaging areas, its histogenesis...
continues to be debated. The mesothelial origin of sclerosing hemangioma has been attributed to undifferentiated mesenchymal and epithelial alveolar cells; however, immunohistochemical and electron microscopy studies support a pathway, which is regulated by the adenomatosis polyposis coli protein.11 Pulmonary sclerosing hemangioma has also been found in association with myomas and with thyroid and kidney cysts.12 This is the first reported case of pulmonary sclerosing hemangioma in Cowden syndrome, and it would seem that sclerosing hemangioma should be added to the list of the many lesions associated with this entity. Nonetheless, the low prevalence of both diseases has meant that it has not been possible to explore the association between them in depth. This case highlights the fact that the lesions associated with Cowden syndrome tend to be quite similar. Even though some of these lesions—for example, pulmonary sclerosing hemangioma—may have a low prevalence, they should be included in the differential diagnosis of solitary lung nodules.

REFERENCES