Siringoma Condroide Maligno: a Propósito de un Caso

Malignant chondroid syringoma: Case Report


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Resumen

Introducción: El siringoma condroide maligno es un tumor cutáneo muy infrecuente pero que debemos tener en cuenta en el diagnóstico diferencial de los tumores axilares.

Objetivos: Se realiza el análisis y la presentación del siguiente caso clínico con el objetivo de considerar esta entidad como diagnóstico diferencial de los tumores axilares.

Presentación de Caso: Paciente de sexo femenino de 82 años que refiere la presencia de una tumoración cutánea en axila derecha de 3 años de evolución, con realización de biopsia (PUNCH) que informa infiltración por carcinoma moderadamente diferenciado. Se decide su exéresis quirúrgica.

Discusión: El siringoma condroide maligno es un tumor infrecuente, pero puede dar metástasis, pero con prolongados tiempos de sobrevida. Prefiere las extremidades y el tronco. Su pronóstico es bueno cuando la exéresis es completa, aunque es necesario realizar un seguimiento completo de los pacientes.

Palabras clave: Siringoma Condroide Maligno. Tratamiento quirúrgico

Abstract

Introduction: Although the malignant chondroid syringoma is a very rare skin tumor, it must be considered in the differential diagnosis of axillary tumors.

Objectives: The following clinical case is analyzed and presented in order to consider this entity as differential diagnosis of axillary tumors.

Case Report: 82 year-old, female patient who presents with a skin tumor of 3 years evolution in the right axilla. Biopsy (PUNCH) reports infiltration due to moderately differentiated carcinoma. Surgical extirpation is decided.

Discussion: The malignant chondroid syringoma is a rare tumor, but can metastasize, with extended survival times. It prefers limbs and trunk.

Its prognosis is good when the extirpation is complete, but patients must be followed-up anyway.

Keywords: Malignant chondroid syringoma. Surgical treatment.
Introduction

The usually called “chondroid syringoma” is considered a rare pathological entity affecting mainly glandular processes\(^1,2,6\). It is extremely malignant. As regards its location, it is most commonly found, in 80% of cases, on the head and neck (nose, cheek, upper lip, hairy skin and forehead); this location is followed, in 10% of cases, by the trunk and limbs and, less frequently, it can affect the feet, axillary region, abdomen, penis, vulva and scrotum. It presents as a slow-growing asymptomatic nodule which may develop metastasis over time.\(^2,8,9\)

It is difficult to diagnose the chondroid syringoma before performing the biopsy, and its result may not be definitive since this is a tumor made up of two very different parts. The final diagnosis is provided by the histopathological and immunohistochemical examination of the complete tumor piece and differential diagnosis must be performed against similar entities such as mioepithelium, chondroid choristoma or fibromyxoid tumor.\(^3,4,6\)

Objectives

To highlight the finding of this rare tumor including it within the differential diagnoses of axillary tumors.

Case Report

82-year-old female patient who makes reference to the presence of a cutaneous tumor of 3 years evolution in her right axilla. Biopsie (PUNCH) reports infiltration due to moderately differentiated carcinoma. Surgical extirpation is decided. Several complementary studies are carried out (breast ultrasound, mammography, thoracoabdominal tomography). Both the tomography and the ultrasound show images compatible with adenopathies in the right under arm area.

The surgical procedure is carried out (tumor resection plus right axillary emptying). The results of the pathological anatomy showed skin mixed tumor (malignant chondroid syringoma) with axillary nodal metastases (17/17).

It is submitted to the tumor committee where progress monitoring and a new assessment to start radiation therapy and chemotherapy are decided.
Discussion

The names “chondroid syringoma” and “mixed tumor” have been used as synonyms; whenever it is located in the salivary gland it is called pleomorphic adenoma.\(^1\)\(^2\)\(^5\)\(^7\)

This type of tumor is rarely found outside the salivary glans, but that may sometimes happen.\(^3\)\(^4\)

Although the benign mixed tumor of the skin is relatively frequent, the chondroid syringoma is extremely rare. It is more common in women (women: men 3:2); it is more frequently found in trunk and limbs than in head and neck. Often, it is bigger than the benign form and presents as a firm, unique, subcutaneous, slow-growing, simple or multiple asymptomatic nodule. It may develop metastasis but has extended survival times. Unlike the benign variety, it prefers limbs and trunk. It is difficult to diagnose the chondroid syringoma before performing the biopsy, and its result may not be definitive since this is a tumor made up of two very different parts. The final diagnosis is provided by the histopathological and immunohistochemical examination of the complete tumor piece and it is performed through exclusion against other entities such as mioepithelium, chondroid choristoma, fibromyxoid tumor, sebaceous cyst, implantation dermoid, neurofibroma, dermatofibroma, basal cell carcinoma, squamous cell carcinoma, pilomatrixoma or histiocytoma.

The malignant chondroid syringoma is a very unfrequent cutaneous tumor with a very low incidence; however, it must be considered in the differential diagnosis of axillary tumors since it has a good prognosis when the “complete excision” is performed. In spite of that, the patient must be strictly followed.\(^1\)\(^2\)\(^6\)\(^7\)\(^8\).

References