

# Mixed Skin and Ecrino Component Cutaneous Carcinoma: Literature Review and Clinical Case Presentation

Mijares A Avenida El Hatillo\*, Franco A, Suarez C, Hurtado Y, Aranguren C, Sánchez A, Pérez A, and Aquiles S

Centro Médico Docente La Trinidad, Caracas, Venezuela

## Abstract

**Introduction:** Tumors of the skin adnexal are lesions that arise from, or differ towards, structures in normal skin, such as regions of hair follicles, ducts, sweat glands, sebaceous glands, or a combination of these elements. Corresponding to 0.0005% of all malignant neoplasms of the skin. The present work aims to review the literature on skin adnexal carcinomas, their behavior and management, as well as to present a clinical case from the head and neck surgery service of a female patient with skin adnexal carcinoma.

**Methods:** The search was in the PubMed and Cochrane Library databases, the key words were: trichilemmal carcinoma of skin adnexal and eccrine carcinoma of skin adnexal, a descriptive review was made. The patient was a 31-year-old female, evaluated in April 2021, with a nodular lesion involving the nasal vestibule, columella and jugal mucosa of the upper lip, with 1 year of evolution, with a biopsy that reported: basal cell carcinoma.

**Results:** 11 publications were identified. A descriptive review of the anatomical and epidemiological aspects, clinical findings and treatment was carried out. The patient deserved micrographic oncological resection with immediate reconstruction, whose definitive biopsy reported: Neoplastic lesion of skin attachments with an infiltrative pattern, consisting of covering the outer layer of the hair root and exhibiting pillar keratinization. Immunohistochemistry reported CEA: weak cytoplasmic positivity in tumor areas with a ductal appearance. The definitive report suggested: Adnexal microcystic carcinoma.

**Conclusions:** Investigations of skin adnexal tumors such as trichilemmal carcinoma and eccrine carcinomas are rare, the behavior and prognosis are difficult to predict. Its diagnosis must be supported by biopsy and immunohistochemistry. Mohs surgery or margin-free surgery is ideal to achieve tumor-free margins.

**Keywords:** Skin Adnexal Tumors; Trichilemmal Carcinoma; Eccrine Carcinoma; Mohs Surgery

\*Correspondence to: Mijares A Avenida El Hatillo, Centro Médico Docente La Trinidad, Caracas, Venezuela; E-mail: [mibuenasalud@gmail.com](mailto:mibuenasalud@gmail.com)

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## Introduction

The tumors of the skin annexes are lesions that arise from, or are differentiated, normal skin structures, such as rich follicles, ducts, sweat glands, sebaceous glands or a combination of these elements [1]. They are rare, correspond to 0.0005% of all malignant skin neoplasms [1].

As a purpose, review the literature of carcinomas of cutaneous annexes, their behavior and handling, as well as present a clinical case of the head and neck surgery service of a female patient with carcinoma of skin annexes.

## Methods

The search was in the PubMed and Cochrane Library database, the keywords were: Trilemmal carcinoma of cutaneous annexes and eccrine carcinoma of cutaneous annexes, a descriptive review was made. The patient was a 31-year-old female, evaluated in April 2021, with a nodular lesion involved in nasal hall, columela and upper lip jugal mucosa, with 1 year of evolution, with biopsy that reported: basal cell carcinoma.

## Surgical procedure

The team was made up of head and neck service surgeons, plastic and reconstructive surgery and pathological anatomy service. In the first instance, the lesion demarcation with oncological resection margins in nasal lobby, columela and yugal mucosa.



Figure 1: Nodular lesion in nasal lobby.



**Figure 2:** Demarcation of the lesion in nasal lobby and



**Figure 3:** Columella that extended to yugal mucosa.

Skin incision with scalpel was made in the right nasal lobby, part of the columen, filtrum and yugal mucosa, until the resection of the lesion was carried out, the micrographic margins were resected with histopathological verification until it is achieved free margins of injury.



**Figure 4:** Oncological resection culminated.

After the resection guaranteeing free margins, together with the reconstructive equipment, immediate reconstruction was made with double advance flap in V-Y with Nasogenian extension.



**Figure 5:** Immediate reconstruction with bilateral V-Y advance flap with Nasogenian extension.



**Figure 6:** Patient 4 months after surgery.

The patient evolved satisfactorily in her recovery.

## Results

11 publications were identified. Descriptive review of anatomical, epidemiological aspects, clinical findings and treatment were carried out. The patient meriterite micrographic resection with immediate reconstruction whose definitive biopsy reported: Neoplastic lesion of skin annexes of infiltrative pattern, constituted by cords and cell trabeculae that cover the outer layer of the hairline and exhibit pilar keratinization.

Immunohistochemistry Report: S-100, Androgen Receiver: Negative in tumor cells and CEA: Weak cytoplasmic positivity in ductal-looking tumor areas. Concluding: microcystic annexial carcinoma. It concludes Diagnosis of Mixed Cutaneous Annexes of Trihachemal and Ecrino component.

## Anatomical aspects

The skin has an area of around 2m<sup>2</sup> and a weight of 4 kg, which represents approximately 6% of the total body weight. It consists of outside in, by three layers: epidermis, dermis and hypodermis. The formation of the hair follicle involves a series of complex signs between the dermis and the epidermis [2,3]. The hair follicle with the sebaceous and sweaty glands apocrine form the Pilosebácea unit. The sebaceous glands are composed of acinos attached to an excretory duct, the apocrine sweat glands lead to the hair follicle and the ecrine sweats are developed in the superficial epidermis [4].

The Ecrine glands express in the secretory portion, cytoqueratin, AME and ACE. Myoepithelial cells exhibit S-100 [5-7]. The hair follicle is composed of three segments: the infundibulum it includes from the mouth of the sebaceous duct to the follicular hole, the isthmus that includes from the mouth of the sebaceous duct to the insertion of the erector muscle of the erector Hair and the lower end ranges from the insertion of the erector muscle to the hair bulb and is formed by the internal and external root pods [2,3].

## Characteristics of cutaneous annexes tumors

The tumors of the skin annexes are lesions that arise from, or differ, normal leather structures, such as pile follicles regions, ducts, sweat glands, sebaceous glands or a combination of these elements [1]. They are rare, correspond to 0.0005% of all malignant skin neoplasms. It is a group that has several limitations for diagnosis, such as: high degree of difficulty in differentiating them from their benign counterpart and to differentiate them between themselves and metastases: salivary gland, breast, digestive tract, ovary, lung, kidney, vulva, vulva, etc., [5]. This kind of neoplasms includes benign tumors and carcinomas. Annexial tumors are often presented as solitary sporadic lesions, but in some



cases, they can presage the presence of a hereditary tumor syndrome [1,5]. Multiple Hamartom with characteristics mucocutaneous lesions that include, among other diagnoses, many triharilemmas [8].

Cutaneous annexes can be tumors with sebaceous differentiation or with follicular differentiation (pylomatricoma and pylomatric carcinoma, tricoblastoma, triquemoma and tri-samemal carcinoma), the tri-samemal carcinoma, was first reported by Headington in 1976 [8,6]. Trihachemic carcinomas are rare malignant annexial tumors that derive from the keratinocytes of the outer root sheath of the hair follicles [7,8]. Other tumors of cutaneous annexes are the tumors of the Ecrine sweat glands (Sclerosing duct carcinoma, siringoid carcinoma, syringes Anaplastic, Ecrino epithelioma) and the apocrine sweat glands, it is noteworthy that annexial tumors with follicular differentiation and mixed glandular have historically designated themselves as apocrine, even in the absence of specific apocrine morphology (cylindrical, expiration and expiration Digital papillary adenocarcinoma that is an endocrine carcinoma of sweat glands [1]. South-gland tumors have also been described such as: hydracnoma and hydradenocarcinoma and annexial microquistic carcinoma. The poromas and atocarcinomas that also show differentiation of the dermal sweat ducts [6].

## Epidemiology

Although the incidence rate of carcinomas of cutaneous annexes is not clear, it is known that it is very rare. They are five times more common in women than in men [7,8]. Microquistic ecrino carcinoma, it is a malignant neoplasia of rare cutaneous annexes, the age of presentation is at 40, predominates in men (3: 1). For the trihylamal carcinoma, the type of skin or ethnicity did not comment frequently, but there are many cases published in the Korean, Japanese and China literature, as well as a case reported in an African-American [9,10]. For the trikylamal carcinoma according to the review according to the review from Hamman MS, et al. (2014) [9], only 103 cases have been notified until 2014 [9].

## Clinical aspects

They arise in areas of the scalp in more than 90% of patients, but can occur on the back, vulva, nose, pubis, buttocks, dolls or elbow. Most of the malignant neoplasms of cutaneous annexes have no characteristic clinical marker that leads us to elaborate the correct diagnosis, the clinical diagnosis cannot go beyond proposing to the lesion as probable malignant neoplasia of annexes [6]. It is presented as an exophic or asymptomatic polypoid mass, sometimes with ulceration and with telangiectasia [9,10, and 11]. tend to develop as Solitarian nodules in the skin exposed to the sun and risk factors have been identified that include ionizing radiation, trauma or previous scar and genetic disorders [9]. Many originate from pre-existing benign neoplasms. Microquistic ecrino carcinoma occurs with predilection in the face, the upper lip and the nasolabial groove (85%). They are plaques or very slow growth nodules (years), are asymptomatic or give the feeling of restarting the skin and are of ecrine differentiation and pillar [1].

## Histopathology

In ecrino carcinomas, clear cells are diffuse, its proliferation index is low: a mythosis in 10 40x fields, Ki67 less than 5%. There are training of concentric structures around nerve trunks and neural and vascular invasion, positive for s 100 on tubular periphery [5,6]. Mutations in DNA repair gene tri-samemal carcinoma, sample keratinization and a peripheral pattern in palisade indicating the origin of the follicular root sheath, consists of cytologically atypical glycogen cells with

lobular, infiltrative growth and, often, it focuses on a Pilosebacea unit, the tumor cells of the center are large, polygonal and clear and clear With glycogen rich cytoplasm, sensitive to diastase, they have atypical nuclei and a high mitotic index. The profile reveals a frequent TP53 mutation, with additional alterations variables, it also shows a hot spot for mutations in Alpk1 [1,6].

Kurokawa examined the expression of cytoqueratin (CK) using 9 different markers and found the presence of CK1, which suggests a differentiation of follicular infundibulum. The CK17 is found in the external root sheath and some authors have used it as a marker to support the diagnosis. Positive staining with Ki67 has been shown. The UEA-1 lectin is strongly positive in the dod cells of the outer root and in the trikylamal carcinoma [6,9].

## Treatment

Mohs micrographic surgery (MMS) has proven to be an effective therapy for a series of rare malignant skin tumors. It is a technique originally described in 1932 by Dr. Frederic Edward Mohs to treat malignant skin lesions. This technique allows the extension of the tumor to safely delimit, as well as conserve as much tissue as possible of healthy tissue, obtaining smaller defects with healing rates greater than other surgical modalities [12]. It is a complex; highly specialized surgical technique and its denomination combines Latin roots related to the microscopic evaluation of tumor margins ("micro") and the mapping Detailed tumor orientation ("graphic"). The distinctive characteristic of this technique is the horizontal section of 100% of the tumor margins to be examined in the microscope, unlike conventional surgery that uses the vertical section of margins [13].

## Discussion

The case presented represented a diagnostic and reconstructive challenge. Trikymic carcinomas are locally invasive, they are tumors of the outer sheath of the hair follicle, which represent 1% of annexial carcinomas and it was described by Headington in 1976, who suggested several criteria for his diagnosis: continuity of the epithelium of the external sheath of a hair follicle, epithelium rich in glycogen, trihylamal keratinization and immunohistochemical details similar to the epithelium of external sheath [10]. It is located in Areas exposed to the sun such as the face and scalp, but it is rarely in nasal lobby, as is the case of the patient presented. In a review by Hamman MS, et al. (2014) [9], of the 82 cases that reported, 42 cases (51%) were found in the face. The Middle Ages was 70 years. It is presented as an exophytic, nodular or mass tumor that can be accompanied by teangiectasias [6-8]. Tri-while carcinomas show malignant cells with atypia, abnormal mitosis, high mitotic rate, aneuploidia, complete loss of the P53 tumor suppressor gene due to the Loss allelic of the short arm of the 17P chromosome, form immature Pilosebacea units, with keratinization and the cells can display negative ver-EP4 unlike the basic carcinoma that is usually negative.

The PTEN mutation in the germinal line can be found in cases of codwen syndrome, however, the patient did not have this syndrome [8]. Microquistic ecrino carcinom Malignant neoplasia of rare cutaneous annexes, of low grade, predominates in men (3: 1), the presentation sites are the head and neck, with predilection on the face, the upper lip and the nasolabial groove (85%), The patient's injury to the present study was in nasal lobby in relation to upper lip [1]. They are very slow growth plates or nodules (years) and are asymptomatic, characteristics similar to the patient injury presented and usually these tumors these tumors They exhibit Ecrina and Pilar differentiation. Clear cells are



identified diffuse, their proliferation index is low: a mytosis in 10 40x fields. There are formation of concentric structures around the nerve trunks and neural and vascular and s 100 positive invasion in the tubular periphery [5,6]. The immunohistochemistry of the patient's sample negative in the patient presented and CEA with weak cytoplasmic positivity in areas of ductal appearance tumor. The clinical, anatomopathological and immunohistochemical characteristics coincide in this lesion, it is a carcinoma of mixed cutaneous annexes of pyloso component (triquemal) and Ecrino.

Surgery with free margins in curative in most cases. It has been shown that Mohs micrographic surgery has higher rates of absence of recurrence, becoming the standard gold treatment. During the patient's surgery described, the totality of the surgical margins were histologically analyzed until they were negative [12]. Maya, They reported that despite the wide resection there was recurrence of the tumor in two cases, but 24 of 26 Patients were operated with satisfactory results [10]. Hamman showed that surgery is an effective therapy using it successfully in their cases. Postoperative radiotherapy should only be considered in patients with high-risk characteristics of local recurrence [6-8]. Maya-Rico AM, et al. (2017) [10], reported 1 patient with distance metastasis and 1 patient with regional metastases, who died after surgery, radiotherapy and chemotherapy [10].

## Conclusions

The investigations of the tumors of cutaneous annexes such as trikylamal carcinoma and ecrino carcinomas, are rare, behavior and prognosis, are difficult to predict. Most of the malignant neoplasms of cutaneous annexes have no characteristic clinical marker. They are located in areas exposed to the sun such as the face and scalp. They tend to develop as solitary nodules and risk factors have been identified that include ionizing radiation, trauma or previous scar and genetic disorders. Its diagnosis must be sustained with biopsy and immunohistochemistry. Mohs micrographic surgery is ideal and it is important to get tumor -free margins as a whole. For this, an important diagnostic methodology is required, which aims to locate the tumor, the technique of its approach and the procedures to achieve a tumor resection with free margins with the histological verification. The margins must be enough, equidistant and free. There is still no consensus on optimal treatment with radiotherapy or chemotherapy.

## Declarations

The authors declare that they have no conflicts of interest, that the work has been approved by the ethics committee responsible in the workplace, and do not declare means of financing of the work carried out.

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