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Lymphoepithelioma-Like Carcinoma: A Diagnosis to Suspect in Suspicious Skin Lesions

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Abstract

Case Report

Primary lymphoepithelioma-like carcinoma of the skin is an uncommon condition with limited likelihood of spreading to other parts of the body. Its appearance and characteristics resemble those of lymphoepithelial carcinoma found in the nasopharynx. We report a case of a rare tumor involving a 68-year-old man with a small, pearl-like bump below the ear helix, about 1.2 cm wide. Initially suspected to be basal cell carcinoma or squamous cell carcinoma, the lesion was surgically removed. Examination under the microscope revealed a cancerous growth featuring clustered cells with large nuclei, surrounded by dense infiltration of lymphocytes. Immunohistochemical testing showed the presence of cytokeratin-positive cells (AE1/AE3) and, indicating the tumor's origin from epithelial cells with squamous differentiation. This highlights the importance of considering primary lymphoepithelioma-like carcinoma when encountering skin lesions.

Keywords: Primary lymphoepithelioma-like carcinoma – good prognosis – excisional biopsy - Immunohistochemical testing.

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INTRODUCTION

The lymphoepithelioma-like carcinoma is a rare type of poorly differentiated cancer characterized by significant infiltration of lymphocytes and plasma cells. While it's commonly found in the nasopharyngeal region, it can also occur in other areas such as the salivary glands, thymus, tonsils, and cervix. The occurrence of lymphoepithelioma-like carcinoma on the skin was first documented in 1988 by Swanson *et al.*, in a series involving five patients. Recently, Gille *et al.*, conducted a review covering around 60 cases. This article presents a case of a Moroccan patient diagnosed with this particular type of cancer.

CASE REPORT

We report a 68-year-old male patient with an erythematous crusty papule located superiorly in the helix of the left ear measuring 1,2 cm in diameter (Figure 1). He underwent excisional biopsy with clinical suspicion of squamous cell carcinoma or basal cell carcinoma.



Figure 1: Erythematous crusty papule, located in the helix of the left ear. Erythematous papule located inferiorly to the left eyelid measuring 1,2cm in diameter. Our clinical suspicion was squamous cell carcinoma or basal cell carcinoma

Excisional biopsy was carried out in our department with 5mm margins under local anesthetia (Figure 2), It consisted on an amputation of the superolateral part of his left ear, followed by skin closure.

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Figure 2: Per-operative photos of the excisional biopsy

Histological examination revealed a skin The epidermis is normoacanthosic, covering. surmounted by orthokeratotic keratosis. The dermis is the site of an infiltrating tumor proliferation arranged in clusters, trabeculae and isolated cells. Tumor cells are medium to large in size, with irregularly contoured, anisokaryotic nuclei that are hyperchromatic in places, with vesicular chromatin, and nucleolated elsewhere, the site of abnormal mitosis. Cytoplasm is moderately abundant and basoohilic. The stroma reaction is fibroinflammatory. No vascular or lymphatic emboli was seen.

An immunohistochemical study was conducted to confirm the epithelial origin of the lesion. Positive staining for cytokeratins (AE1/AE3) indicated a diagnosis of carcinoma. Squamous differentiation was further confirmed by positive staining for p63. The significant inflammatory infiltrate tested positive for CD45, while the analyzed neoplastic cells showed negative results for the same marker.

Morphological and immunophenotypic findings supported the diagnosis of lymphoepitheliomalike carcinoma of the skin. The patient was screened for possible primary lesion of the nasopharynx. Based on the negative results for nasopharyngeal cancer or other types of tumors, a primary cutaneous carcinoma was confirmed. The patient had a favorable evolution, without relapses or lymph node involvement.

DISCUSSION

Squamous cell carcinoma of the skin is a cancerous growth characterized by a range of clinical behaviors, from slow-growing to aggressive tumors with an average potential to spread to other parts of the body. According to a classification suggested by Cassarino *et*

al., lymphoepithelioma-like carcinoma represents a variant with intermediate malignant potential, carrying an estimated 5-10% risk of metastasis.

It is a rare skin cancer that typically affects individuals over the age of 50, with no specific gender preference. The exact cause of this variant of squamous cell carcinoma remains unclear. Ho *et al.*, proposed two theories to explain its origin. One theory suggests an epidermal origin, but this is contradicted by the lack of connection between the lesion and the epidermis in reported cases. The other theory proposes an adnexal origin, supported by the presence of areas showing adnexal differentiations in some previously described tumors.

Clinically, lymphoepithelioma-like carcinoma lesions are commonly found on the scalp, neck, trunk, and extremities, often in areas exposed to sunlight. They typically manifest as erythematous nodules, plaques, or papules that evolve over several months. The patient in this case had an erythematous papule for seven months, consistent with the typical presentation described in the literature.

Morphologically, these neoplasms resemble those observed in other locations such as the nasopharynx. They usually appear as well-defined dermal nodules composed of syncytial-like epithelial clusters surrounded by inflammatory lymphocytic infiltrates, sometimes accompanied by plasma cells. The syncytial appearance of epithelial clusters is a characteristic feature of this type of neoplasm. The cytological pattern observed in our case is typical, with large vesicular nuclei and prominent nucleoli. The inflammatory infiltrate in these neoplasms exhibits a reactive behavior and is primarily composed of T lymphocytes, similar to healthy skin samples.

The diagnosis of lymphoepithelioma-like carcinoma of the skin typically involves clinical investigation of the primary lesion, particularly in the nasopharynx, through imaging and laryngoscopy. If no lesions are found in other locations, the case is considered primary cutaneous. The primary treatment approach is excisional biopsy, which often leads to a good prognosis with low recurrence and metastasis rates. However, cases of local aggressiveness and metastasis, although uncommon, have been reported.

The role of Epstein-Barr virus (EBV) in the pathogenesis of this skin neoplasm has not been established. Unlike in the nasopharynx, studies on cutaneous neoplasms have not demonstrated an association with the virus. However, such an association has been established in other anatomical sites like the lung, stomach, and salivary gland.

While cases like this one may not be rare, they are unusual and may be overlooked by pathologists and dermatologists due to their infrequency. Sharing experiences with this tumor helps in understanding its various manifestations, which are not always typical, and contributes to knowledge about the disease.

CONCLUSION

Lymphoepithelioma-like carcinoma of the skin is a rare neoplasm with variable clinical behavior and uncertain etiology. It typically affects individuals over 50 years of age, with no gender predilection, and commonly presents as erythematous nodules, plaques, or papules, often in sun-exposed areas. Diagnosis requires thorough clinical investigation, including examination of the nasopharynx, and treatment primarily involves excisional biopsy. Despite its potential for local aggressiveness and metastasis, most patients have a good prognosis. While the role of Epstein-Barr virus in the pathogenesis of this neoplasm remains unclear, sharing experiences with this tumor helps broaden understanding and recognition among pathologists and dermatologists, contributing to improved management of the disease.

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