

## Challenges in Diagnosis and Management of External Ear Melanoma Case Report and Literature Review

Erisa Kola<sup>1\*</sup>, Edlira Horjeti<sup>2</sup>, Elmijola Janushaj<sup>3</sup>, Ina Kola<sup>4</sup> and Mehdi Alimehmeti<sup>1</sup>

<sup>1</sup>Department of Pathology “Mother Teresa” University Hospital Center, Tirana, Albania

<sup>2</sup>Family Doctor, Primary Care Center, Nr. 2, Tirana, Albania

<sup>3</sup>Dermatologist, Vlora Polyclinic Dermatology and Venerology, Vlora, Albania

<sup>4</sup>Department of Burns and Plastic Surgery, “Mother Teresa” University Hospital Center, Tirana, Albania

### \*Corresponding author

Erisa Kola, Department of Pathology, “Mother Teresa” University Hospital Center, Tirana, Albania, Ph: +35-5-692869351; E-mail: erisa\_k87@yahoo.com

Submitted: 22 May 2019; Accepted: 27 May 2019; Published: 05 June 2019

### Abstract

Primary external ear melanoma is a rare disease that accounts for only 1% of all cutaneous melanomas and only 14.5% of cutaneous melanomas of the head and neck region [1]. The external ear represents a site with high ultraviolet exposure and predisposed to various cutaneous lesions. The pathophysiology of these tumors is different from other skin lesions because of their specific anatomical site close to cartilage, lymphatic drainage and functional characteristics. Malignant melanoma of the ear was considered to be very aggressive, with a propensity for spreading to both regional lymph nodes and distant sites. Surgical management of external ear melanoma has always been a challenge between avoiding aggressive amputations, achieving an aesthetic pleasant result and adequate oncologic control. According to our literature review a limited number of publications exist regarding malignant melanoma of the external ear.

We present the case of a 69-year-old woman, who was presented at the Department of Dermatology because of an ulcerated lesion in the helix part of the left ear. Clinical, histopathological and immunohistochemical examination confirmed the diagnosis of malignant melanoma of the external ear, nodular type, ulcerated.

**Keywords:** Malignant Melanoma, External Ear Melanoma, Melanoma Multidisciplinary Approach, Excision Margins

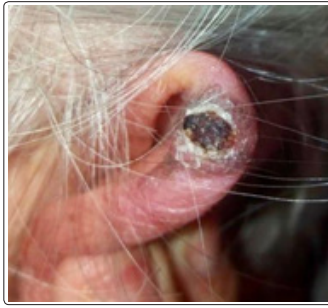
### Introduction

Malignant Melanoma is a malignant tumor arising from melanocytes and comprising one of the most aggressive types of cancers. Melanomas are commonly anatomically found in extremities, but have been seen having a rare localization in the head and neck with 1% of all cases, whereas 14.7% on the external ear [2]. Although melanoma accounts for only 1% of all skin cancer cases, it is responsible for the vast majority of skin cancer deaths. The incidence of cutaneous melanoma in general has risen over the past decade, and this trend has also been noted for melanoma of the external ear [3]. According to our epidemiologic data it typically affects males in the 7<sup>th</sup> decade of life [4, 5]. The difference in incidence by sex has been attributed to the fact that women traditionally have longer hair which serves a protective role [6]. The auricle is a cosmetically sensitive area and plays an important functional role in hearing by transmitting sound into the external acoustic meatus [7]. The earliest reports of melanoma of the external ear appeared in the mid-1900s. Early studies suggested that external ear melanoma carries a worse prognosis compared to other head and neck anatomic sites due to inherently more aggressive tumor biology, the presence of thin

skin in an area of high levels of ultraviolet exposure, the variable lymphatic drainage pathways of the ear [8, 9]. Subsequent literature has indicated that when controlled for other prognostic variables, location on the ear is not associated with a worse prognosis [10, 11].

### Case presentation

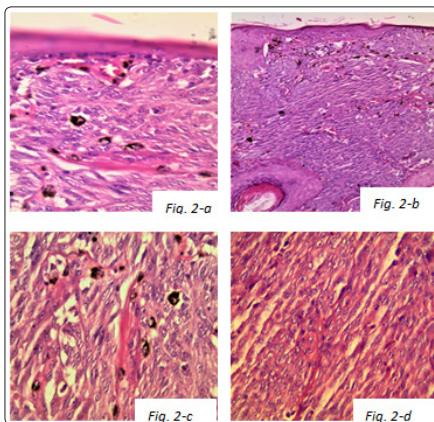
A fair-skinned 69-year-old female was presented at the Department of Dermatology because of concerning signs regarding a nodular lesion on the helix part of the left ear. At the time of presentation she complained of intermittent bleeding. She displayed that she started to notice the appearance of a small beige bump in her left ear one year ago, continuously growing in size, and last 6 months started itching and bleeding. She refers she has been scratching it chronically until it bleeds and sores. Neglecting the situation over a year she has noticed changes in size, shape and color. The patient reveals that she has been working for a long time as an agricultural farmer often carelessly after long-term exposure to the sun. Dermatological examination showed a single dome-shaped lesion measuring 2 cm x 1.5 cm x 0.5 cm, covered with sero-hemorrhagic crusts, with a white color hyperkeratosis collaret-stripped surrounding (like irregular halo sign). The lesion extends onto a melanocytic base with a 3 cm diameter. Dermatoscopy findings weren't helpful because of the type of lesion covered with crusts all over the surface.



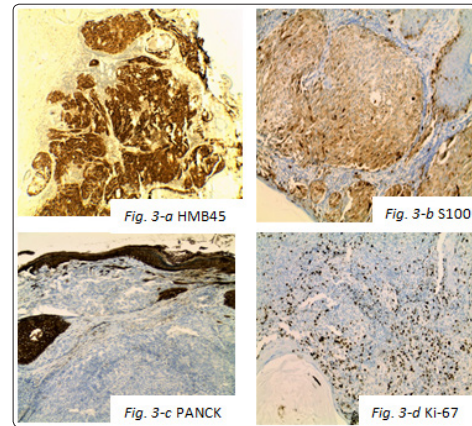
**Figure 1:** Clinical examination showing an ulcerated lesion in the external part of the right ear, appeared first one year ago, covered with crusts all over the surface

She had no familiar history of skin tumors or previous history of pigmented nevus at the site of the lesion. Clinical examination was within the normal limit, except for some suspicious palpable small nodes detected in left cervical submandibular region. Clinical laboratory values were within the normal range except an elevated ESR (76mm/h) and slightly increased LDH. Peripheral blood smear examinations were normal. According to the clinical dermatologic features of the lesion, the clinical suspicion was that of a squamous cell carcinoma of the external ear. Thus, a plan of excision and histopathology examination was made very soon as a final step of diagnosis. The surgical management of the case was an excisional biopsy of the lesion with wide-free margins 1cm laterally to a full depth sufficient to ensure that the lesion was not transected.

Macroscopic examination of the biopsy specimen revealed a exophytic ulcerated lesion measuring 1.8cm in the largest diameter, with ill-circumscribed elevated borders, covered by crusts and hemorrhage. Microscopic examination of the lesion showed a polypoid intradermal tumor with ulceration of the overlying epidermis composed of nests of epithelioid tumor cells which extended down to the dermis. The tumor cells had round to oval nuclei, prominent nucleoli, and abundant eosinophilic cytoplasm with some clearing and a distinct cell border. Some cells were large with pleomorphic nuclei and conspicuous nucleoli. Typical and atypical mitotic figures were observed in the dermal component. There was loss of epidermal matrix, including ulceration and epidermal consumption.

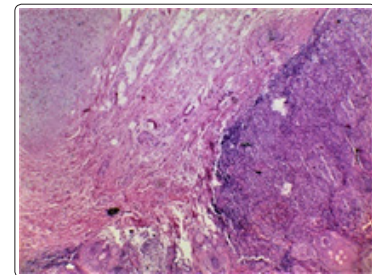


**Figure 2:** Light microscopy findings in the H&E-stained sections revealed an atypical melanocytic proliferation, composed of malignant melanocytes infiltrating the dermis, and dermal mitotic figures present can be seen in the high-power view (c)



**Figure 3:** Immunohistochemical staining for tumor cells showed a strong and diffuse positivity for HMB45 (a) and S-100 (b), while there was no positivity for PanCk in the intradermal neoplastic cells (c), Figure 3-d: Ki-67 shows high proliferation rate (12MF/mm<sup>2</sup>)

The final diagnosis, based on the histopathology and immunohistochemical findings was malignant melanoma, nodular type, ulcerated, with 5mm Breslow thickness and Clark level V. Histopathology examination of the margin representing the erythematous ring surrounding the lesion showed the presence of lymphocytes and plasma cells, suggesting an inflammatory reaction.



**Figure 4:** Light microscopy findings in this H&E-stained section highlights the depth of invasion of the tumor, with a wedge-shaped type infiltrative pattern, very close to the ear cartilage, however there is no invasion through it

## Discussion

Malignant melanomas of the ear are uncommon lesions that require a multidisciplinary approach and careful surgical planning to minimize the cosmetic and functional consequences of surgery [12, 13]. Historically, external ear melanomas have been treated aggressively, due to early perceptions suggesting they had poor prognosis and aggressive biological behavior [14, 15]. Poor outcomes in these reports implied radical surgical management of melanoma of the external ear, which included total amputation of the ear, parotidectomy, and prophylactic dissection of the neck [16, 17]. Particularly, total or partial amputation of the ear could render a patient unable to wear eye glasses or some hearing aids. These aggressive recommendations dominated the literature for over a decade until larger series were reported [18, 19]. Recent studies, have demonstrated that these melanomas can be treated conservatively avoiding aggressive amputations and advocating more moderate treatment strategies including Mohs surgery, composite “wedge” excision of skin and cartilage-sparing surgery [20]. However, due to the unpredictable lymphatic drainage pattern of the ear, which may spread to the pre-auricular or post-auricular sites, parotid gland, or



anterior or posterior cervical chains, sentinel lymph node biopsy is performed to identify the risk of advanced stage and guide further management such as parotidectomy and neck dissection [21-23].

Adequate biopsy is the first step for a definite diagnosis of malignant melanoma. Nodular melanoma is an aggressive subtype of malignant melanoma, the second most common, after superficial spreading melanoma, with a potential to metastasize even in early stages. Bleeding and ulceration are common signs of nodular melanoma according to different clinical studies [24]. The presence of ulceration upstages patients with localized melanoma by both subcategories and stages and is included as an independent prognostic factor defining the T-stage in the American Joint Committee of Cancer's melanoma staging criteria [25]. Various current cutaneous melanoma treatment guidelines recommend excision with 1, 2, or even 3 cm margins depending on the tumor depth. Margins of this size are more difficult to achieve on the ear while preserving structure and contour compared with other head and neck melanomas, since an overaggressive treatment can negatively impact cosmetic and functional outcomes [26-31].

Given the vast histologic diversity of melanoma, diagnosis can be challenging. In our case the presenting clinical features of nodular melanoma posed a real challenge in its earlier suspicion due to the symmetrically elevated borders and quite amelanotic appearance that didn't fit the classic "ugly duckling" radial growth phase of melanoma as defined by the ABCDE rule (asymmetry, border irregularity, color variegation, diameter >6 mm, and evolving).

The patient was subjected to a second excision biopsy to achieve a clear margin and further radiological assessments to detect the presence of metastasis.

## Conclusions

Melanoma of the external ear is a relatively rare and infrequently reported disease, with little consensus on its prognosis or management. Consequently, there is currently a lack of consensus regarding best surgical management. Tumor thickness and depth of invasion are the most important prognostic factors regarding malignant melanoma. Ulceration is an important prognostic factor for patients with melanoma and also a predictive marker for the response of adjuvant immune-stimulating therapy. Abnormal melanogenesis and loss of the functional capacity of tumor cells due to rapid proliferation have been postulated to cause the lack of pigment and amelanosis. Such atypical clinical features may pose a challenge to early suspicion and diagnosis, leading to advance stage diagnosis and poorer prognosis. Hence high suspicion should be attributed to skin lesions over sun-exposed areas along with the patient history of a rapidly growing lesion.

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