Case Report / Olgu Sunumu

Non-familial Multiple Trichoepithelioma: A Case Report

Non-familyal Multipl Trikoepitelyoma: Olgu Sunumu

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Abstract

Trichoepithelioma is a well-differentiated benign follicular tumour. Clinically, it may either be solitary or multiple. Multiple trichoepitheliomas are often located on the face and its vicinity. Here, we present this case due to the increasing number of lesions over ten years, particularly with the lesions blocking the external auditory canal. This case is presented with clinical, histopathological and immunohistochemical features, with differential diagnoses. The patient was a 70-year-old female with localised papules on the external auditory canal, ear lobule, tragus, neck and lower lip. The microscopic examination of the excisional biopsy from the external auditory canal revealed a tumoural formation surrounded with stromal fibrosis and mononuclear infiltration, which is composed of basaloid cells showing peripheral palisading and keratinocytes with infundibular keratinisation. Upon immunohistochemical studies, the surrounding stroma of the tumour showed diffuse cytoplasmic positivity for CD34, and focal cytoplasmic positivity for BerEP4. There was no significant family history in this case. This pathology must be kept in mind in the differential diagnosis of tumours of the epidermis and skin appendages which occur on the face and its vicinity.

Keywords
Non-familial, multiple, trichoepithelioma, external ear canal

Öz

Trikoepitelyoma, iyi diferansiye benign folliküler tümördür. Klinik olarak tek olabileceği gibi multipl olarak da izlenebilir. Multipl trikoepitelyoma olguları sıklıkla yüz ve yüz çevresinde yerleşim gösterir. Bu çalışmada, 10 yıldır sayısı giderek artan lezyonları, özellikle dış kulak yolunu tamamen kaplaması ile ortaya çıkan klinik bulguları nedeniyle klinik, histopatolojik, immünohistokimyasal ve ayırıcı tanı özellikleri ile bir olgu sunulmaktadır. Yetmiş yaşında kadın hastanın lezyonları papüller şeklinde olup dış kulak yolu, kulak lobulü, tragus, boyun, alt dudakta lokalizedir. Olgunun dış kulak yolundan alınan eksizyonel biyopsilerin mikroskopik incelemesinde; stromal fibrozis ve mononükleer enfiltrasyonun çevrelediği, periferde palizad yapan bazaloid hücreler ve enfundibular keratinizasyon gösteren keratinositlerden oluşan tümöral oluşum görülmüştür. Olguya uygulanan immünohistokimyasal çalışmada; tümörü çevreleyen stromada CD34 ile yaygın sitoplazmik boyanma, BerEP4 ile fokal sitoplazmik boyanma görülmüştür. Aile öyküsü bulunmayan bu hastada lezyonların çok sayıda olması ve klinik bulguları nedeniyle trikoepitelyoma olgusu sunulmuştur. Yüz ve yüz çevresi yerleşimli epidermis ve deri eki kaynaklı tümörlerde bu antite ayırıcı tanıda düşünülmelidir.

Anahtar Kelimeler
Non-familyal, multipl, trikoepitelyoma, dış kulak yolu

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Introduction

Trichoepithelioma was first described in 1892 by Brooke and Fordyce under the name ‘‘epithelioma adenoides kystique’’ (1,2). Trichoepithelioma is a rare hamartomatous skin tumor that develops from the germinative cells of the folliculo-sebaceous-apocrine unit and shows follicular differentiation (3). It is a type of tumor that occurs as papules with the same color as the skin, especially in the face area. They are clinically seen as solitary or multiple (4-6). Multiple trichoepithelioma shows autosomal dominant inheritance (7). The histological features of these two types are the same. In the current case, a patient with multiple trichoepithelioma obstructing hearing due to completely covering the outer ear canal around the face is presented.

Case Report

A 70-year-old female patient was admitted to the clinic due to the increasing number of skin lesions which were localized in the the outer ear canal, the ear lobule, tragus, neck, lower lip and periorbital area within the 10-year-period. The tumor completely covered the outer ear canal which also caused hearing problems (Figure 1,2). Several excisional biopsies were taken from this area. In the microscopic evaluation, a tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with keratinocytes showing infundibular keratinization have been observed. Horn cysts were common (Figure 3-5).

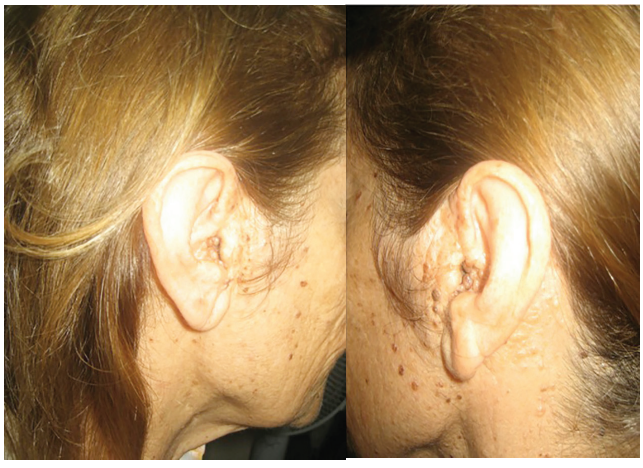

Figure 1,2. Multiple papules that completely fill the outer ear canal in the right and left ear

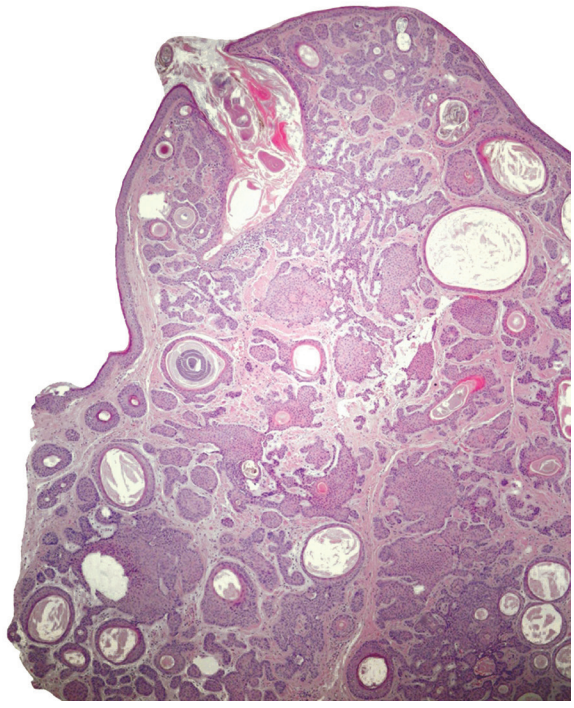

Figure 3. Tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with multiple horn cysts, H&E, X100

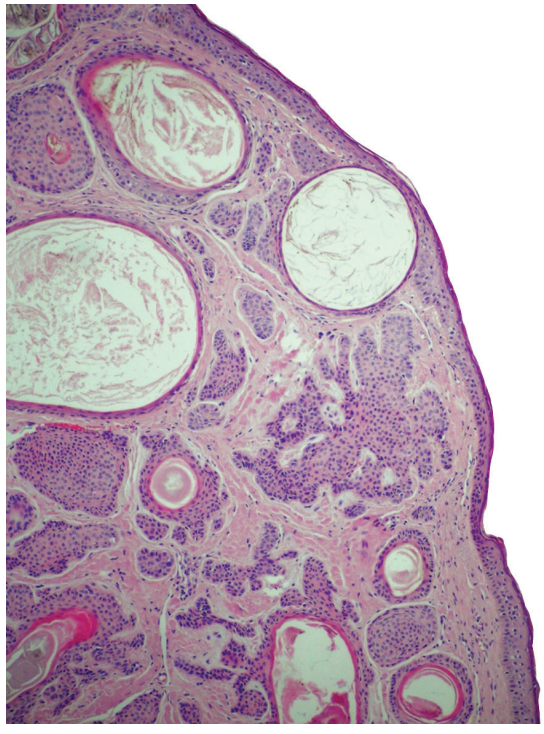

Figure 4. Tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with multiple horn cysts, H&E, X200

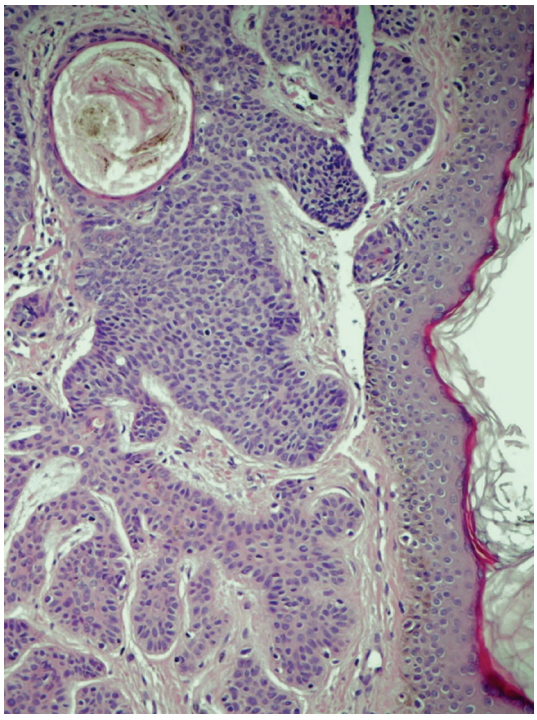

Figure 5. Tumoral formation consisting of mainly basaloid cells that made peripheral palisading of nuclei with multiple horn cysts, H&E, X400

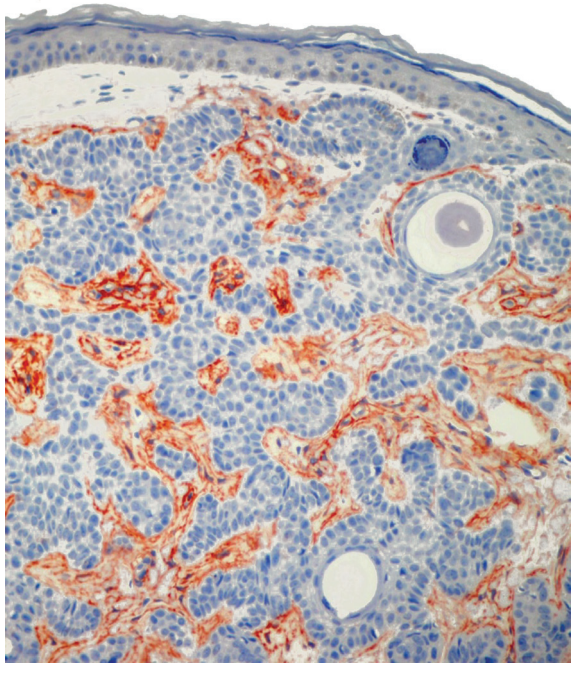

Figure 6. Diffuse cytoplasmic staining with CD34 in the stroma surrounding the tumor, X200

In the immunohistochemical study applied to the case; widespread cytoplasmic staining was observed with CD34 in the stroma surrounding the tumor which supported the diagnosis of trichoepithelioma (Figure 6) wheras BerEp-4 which favors basal cell carcinoma was focally stained in a very limited area. There is no family history in the case. During the clinical follow-up, new lesions have been occurring in the similar area.

Discussion

Trichoepithelioma is a well differentiated benign tumor originating from the hair follicle (3-8). It can be clinically solitary or multiple (4-6). Lesions are most often located in the form of flesh-colored papules located around the face. In this case, skin papules were seen around the face, especially the periorbital area, lower lip, and tragus. The number of the lesions gradually increased in 10 years. However, atypically it caused hearing problems due to the large number of lesions in the outer ear canal. Trichoepithelioma cases show symmetrical location (9). In our case, the lesions were also located within a certain symmetry.

Trichoepithelioma may be familial or may occur sporadically (4,6). There was no family history in this case.

Although there is no genetic difference between women and men, trichoepithelioma frequently occurs in women (10). Lesions complete the formation processes between the ages of 50-70. Our patient was a 70-year-old female patient, and her lesions increased especially in the last 10 years. In terms of diagnosis, it is necessary to take a biopsy and evaluate it histopathologically (11). The most characteristic features of trichoepithelioma are keratin cysts and basaloid cell groups in the form of solid and adenoid formations surrounding them (6). Similar histopathological findings were observed in our case.

Basal cell carcinoma and trichofolliculoma are important in the differential diagnosis of trichoepithelioma. In adults, trichofolliculoma is developing from hair follicles, which are mostly dome-shaped lesions that appear as solitary in the facial region (5). In this study, although our case is similarly located around the face, it differs from trichofolliculoma by being in the form of multiple papules.

It is difficult to differentiate with basal cell carcinoma in immature trichoepithelioma cases where keratin cysts are not seen (3). At this point, immunohistochemical studies are useful in reaching the diagnosis. The stroma between basaloid areas are CD34 positive in trichoepithelioma is positive, while it is negative in basal cell carcinoma (11). In our case, a similar staining profile was observed. Widespread cytoplasmic staining was observed with CD34 in the stroma surrounding the tumor. BerEp-4 was stained in a very focal area.

Malign transformation is rare in trichoepithelioma (12,13). In our case, although the lesions increased in the last 10 years, there was no evidence in favor of malignancy in multiple biopsies taken from the lesions.

In our case, the lesions were seen as multiple papules at different points of the face, especially the outer ear canal and periorbital area. It caused hearing loss due to the large number of occurrence in the outer ear canal.

Local excision is the most significant treatment method, if the lesion is solitary. The surgical approach is not meaningful in the presence of multiple tumors (5). It has been observed that lesions located in the facial region show recurrence after surgical correction by dermabration or laser treatment (14). In our case, the lesions in the outer ear canal were excised locally. During the follow-up, new lesions occured in the similar area in the meantime.

Our case was presented because of the high number of lesions and the clinical appearance that these lesions caused especially in the outer ear canal. The absence of a family history makes it difficult to reach the diagnosis in such sporadic cases, but it is recommended to keep in mind in the differential diagnosis.

Ethics

Informed Consent: Informed consent was obtained.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Concept: D.T., B.Y.Ö., Design: D.T., B.Y.Ö., Materials: A.K.Y., Data Collection or Processing: A.G.S., Analysis or Interpretation: D.T., Literature Search: A.G.S., Writing: D.T., F.K., K.G.E.

Conflict of Interest: No conflict of interest was declared by the authors.

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