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Experiences with Head and Neck Pilomatrixoma

İmdat Yüce[1], Emre Solguntekin[1], Alperen Vural[2], Sedat Çağlı[1], Özlem Canöz[1]

[1]Erciyes University, Faculty of Medicine, Department of Otorhinolaryngology, Kayseri, Turkiye
[2]Istanbul University-Cerrahpaşa, Cerrahpaşa Faculty of Medicine, Department of Otorhinolaryngology, Istanbul, Turkiye

ORCID ID: İ.Y. 0000-0003-0938-4437; E.S. 0000-0003-0611-2823; A.V. 0000-0003-1969-7760; S.Ç. 0000-0003-4913-8687; Ö.C. 0000-0002-0200-6970

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ABSTRACT

Objective: The study aims to provide more information about the clinical features, diagnosis, and treatment of pilomatrixoma, of which little is known.
Materials and Method: The research retrospectively studies pilomatrixoma cases that were operated by a department of otorhinolaryngology between January 2018-October 2021.
Results: Pilomatrixoma is a benign tumor originating from the hair follicle matrix and is observed more frequently in women than in men. It is most commonly seen as a nodule in the head and neck region under the skin. This study examines 7 cases diagnosed as pilomatrixoma.
Discussion: Pilomatrixoma should also be especially considered in palpable superficial or ulcerated lesions in the preauricular region, and differential diagnosis of pilomatrixoma should be made with parotid tumors.

Keywords: Pilomatrixoma, head and neck, benign tumor

INTRODUCTION

Pilomatrixoma is a benign soft tissue tumor originating from the hair follicle matrix. Although Malherbe and Chenantais suggested in 1880 that this benign tumor originates from the sebaceous glands, Forbis and Hellwigshowed showed that this benign tumor arises from the cortex of the hair follicle and named it pilomatrixoma in 1961 (1).

Pilomatrixoma is more common in women than men and usually occurs in the first two years of life. It is most commonly seen as a single nodule under the skin in the head and neck region. However, cases have been found with multiple nodules (2-3).

The treatment for pilomatrixoma is surgical excision. Recurrence is rare, and if it does happen, a malignant pilomatrixoma variant should be suspected (3).

Pilomatrixoma is a rare tumor that is usually seen in the head and neck region and can be confused with malignancy. This study presents head and neck pilomatrixoma cases that have been operated upon and aims to increase awareness of its pathology.

MATERIALS AND METHOD

This article retrospectively studies pilomatrixoma cases that were operated upon by a department of otorhinolaryngology between January 2018-October 2021 and examines the patients' ages, complaints at presentation, location of the lesion, and dimensions, as well as preoperative and postoperative biopsy results. The patients' follow-ups were also recorded. Those who did not follow up were called and invited for a checkup.

RESULTS

The study includes seven patients whose pathology resulted in a pilomatrixoma diagnosis. Six of these patients were women. The age of the patients ranged from 10-64 years (M = 34.8). All of the patients presented with a slow growing mass.

The findings from the patients' physical examinations showed a well-circumscribed mass lesion on palpation. Four of the lesions were located in the parotid region (Figure 1), two were on the sternocleidomastoid muscle, and one was in the frontal region.

Corresponding Author: Emre Solguntekin Bal **E-mail:** esolguntekin@hotmail.com

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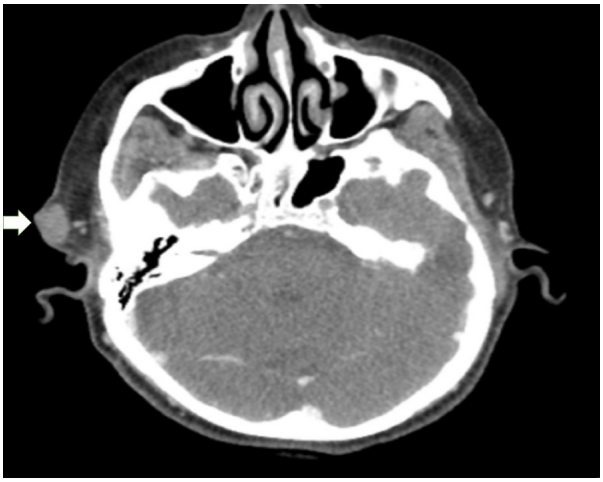

Figure 1: CT image of a 17x12mm contrast-enhancing solid mass lesion (White arrow) in the skin-subcutaneous fatty tissue at the right preauricular level.

All patients had a preoperative radiological examination. More than one imaging method was performed on some of the seven patients. Ultrasonography (USG) was used most frequently due to being non-invasive and easy to apply. However, USG cannot show the relationship of the mass with the parotid as well as computed tomography (CT) and magnetic resonance imaging (MRI) can, and USG may be insufficient regarding masses with a suspected malignant tumor. Therefore, MR and CT were requested for preoperative planning to better understand the relationship of the mass with the parotid gland. USG was run for 5 patients, CT for 3 patients (Figure 1), and an MRI for 1 patient were available. The radiological size of the masses ranged from 1 cm to 3 cm (*M* = 2.1 cm).

When deciding on which biopsy to perform, fine-needle aspiration biopsy (FNAB) was the most common choice due to its ease of application. However, incisional biopsy and Tru-cut biopsy were used in cases where FNAB was insufficient

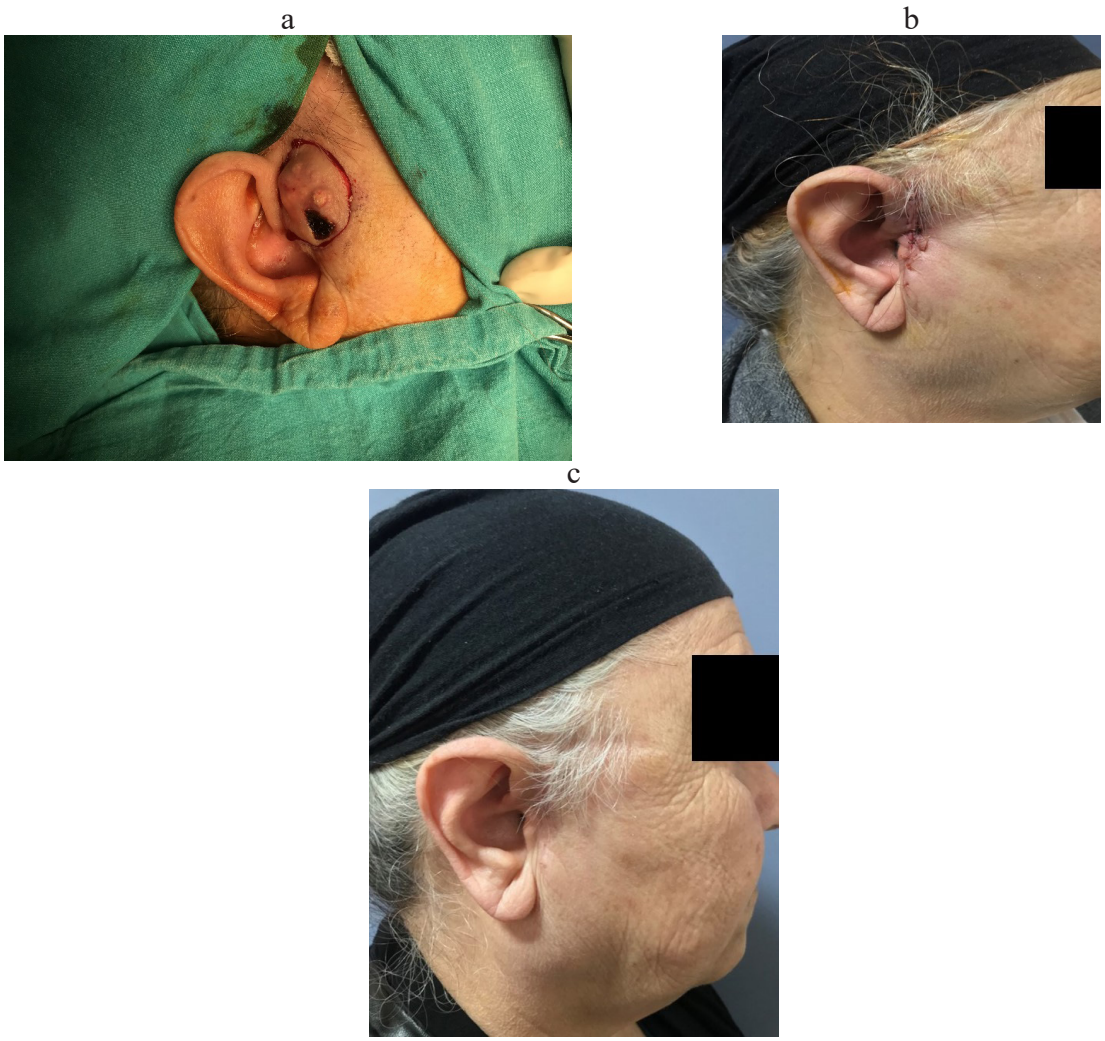

Figure 2: The mass lesion located in the right parotid lodge was excised by taking 2-3 mm from the intact tissue, with primary suturing used to close the defect that had formed. (a) Intraoperative, (b) postoperative 1st-day and (c) postoperative 10th-day images of the patient.

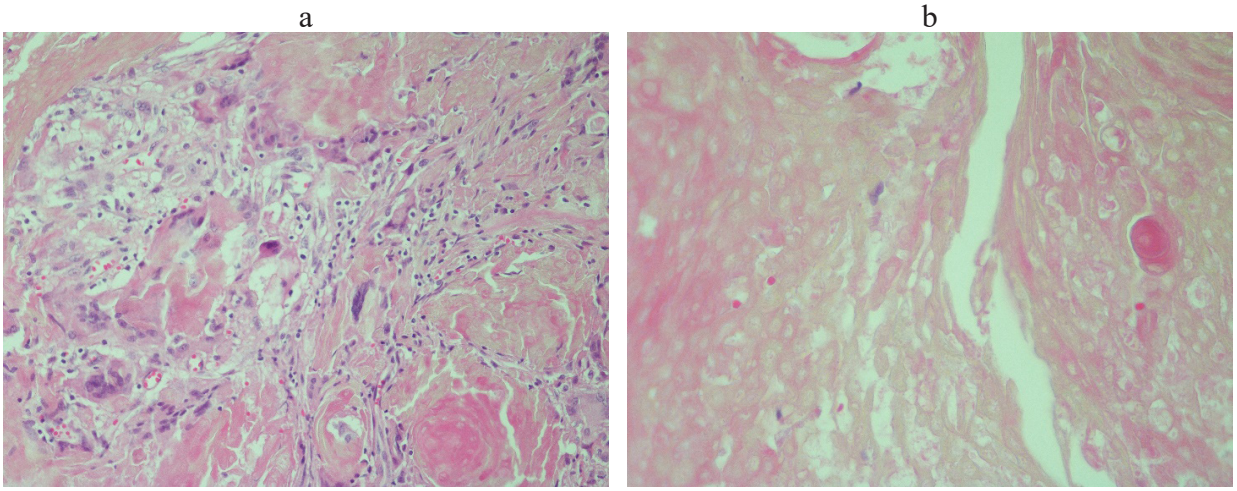

Figure 3: (a) Histopathological image of pilomatrixoma, ghost, and basophilic cells are shown at x20 magnification. (b) Histopathological image of ghost cell shown at x40 magnification.

or the tumor was clinically suspected of malignancy. FNAB was performed in two patients, Tru-cut biopsy was performed on one patient, and incisional biopsy was performed on one patient for diagnostic purposes. In two patients who underwent FNAB, the mass was located in the parotid. One patientis FNAB result was reported as keratinized cells and macrophages, while the other patient's was reported as perhaps compatible with mucoepidermoid carcinoma. The result from the patient who underwent Tru-cut biopsy was reported as benign neoplasia, and the result from the patient who underwent incisional biopsy was reported as pilomatrixoma.

Surgical treatment was applied to all seven patients, and the masses were excised with intact surgical margins and closed with primary suturing (Figure 2). The patient whose FNAB result was compatible with mucoepidermoid carcinoma was presented with a pathology report, and an additional neck dissection was performed. The final pathology reports for all patients were reported as pilomatrixoma. The pathological sizes of the specimens removed during surgery ranged from 0.4 cm to 3 cm (*M* = 1.9 cm).

The histopathological structure of pilomatrixoma involves irregular epithelial cell groups with ghost cells in the center and varying amounts of basophilic cells (Figure 3). The ghost cell is an enlarged eosinophilic epithelial cell with only cytoplasm that has lost its nucleus (4). The number of ghost cells gradually increases with time until the characteristic calcification and osteogenesis of pilomatrixoma occurs (5).

DISCUSSION

Pilomatrixoma is a generally hard and slow-growing benign tumor that develops from the hair follicle matrix and is covered with normal skin tissue. Its etiology is not fully known. This benign tumor has been reported to possibly occur as a result of a disruption in the cycle of the hair follicles (6). As a result of histochemical studies and electron microscopy studies, the belief has formed that these lesions originate from the basal cells of the epidermis. These primitive basal cells transform into hair matrix cells through an uncontrolled proliferation (7).

Although pilomatrixoma usually occurs as a single nodule, it can also occur as multiple nodules (8). Patients' having multiple nodules has been reported to possibly be associated with a familial predisposition to beta catenin gene mutations and to disorders such as myotonic dystrophy, Rubinstein-Taybi syndrome, Turner syndrome, Gardner syndrome, xeroderma pigmentosum, and basal cell nevus syndrome (9-10).In the case series studied in this article, a single nodule was present in all cases. The patients' histories and familial histories revealed no familial predisposition. In most cases, the skin over the tumor may become thinner or even covered with normal skin. In some cases, discoloration of the skin may occur due to ulceration. Patients usually do not have pain complaints. Pilomatrixoma can be located anywhere except the palms and soles of the feet and is especially common in the head and neck region (7, 11). All patients in the current study presented with a palpable mass and no additional symptoms, with one patient also having ulceration of the skin (Figure 2).

The characteristic calcifications of pilomatrixoma can be seen on plain radiographs, but diagnosis is difficult this way (12). USG is the most common imaging method used to aid in diagnosis. It is non-invasive, fast, and easy to apply. USG is important in terms of showing the depth of the lesion, its relationship with neighboring tissues, and calcifications (13). USG has been proposed as an alternative to CT and MRI for imaging preauricular masses in young children, as it can usually be performed without sedation or general anesthesia (12). However, CT and MRI are more helpful in determining the relationship of the preauricular lesion with adjacent structures, especially the parotid gland. Therefore, they provide more benefits in differentiating pilomatrixomas from primary parotid tumors and in preoperative planning. Pilomatrixoma usually appears as a non-contrast-enhancing, well-demarcated, subcutaneous lesion on a CT, whereas on an MRI, it appears as a soft tissue mass with homogeneous moderate signal intensity

on T1-weighted images and as a heterogeneous moderate-to-high signal intensity on T2-weighted images (12-13).

The diagnosis of this benign tumor can be confirmed by histopathological examination. FNAB is useful in the diagnosis of many tumors, as well as in the diagnosis of pilomatrixoma. However, pilomatrixoma aspirates also have properties similar to those in malignant tumors. Therefore, pilomatrixoma can be confused with tumors such as poorly differentiated basaloid cell carcinoma, keratinized squamous cell carcinoma, small cell carcinoma, and Merkel cell carcinoma. The nuclear morphology of pilomatrixoma helps to distinguish it from other tumors (14). The FNAB result of one patient in this study's case series was reported as compatible with mucoepidermoid carcinoma. Therefore, pilomatrixoma should be kept in mind even if a malignant tumor is reported when FNAB is performed due to a mass in the preauricular region.

The treatment for pilomatrixoma is surgery (3). After complete excision of the tumor, recurrence is rare, with malignancy rarely being reported (15). All patients in the current study were treated with surgical excision and cured. No recurrence was observed within at least a 1-year follow-up.

Pilomatrixoma should also be especially considered in palpable superficial or ulcerated lesions in the preauricular region, and a differential diagnosis should be made for it with parotid tumors. As was the case in this study's report, pilomatrixoma and malignancy can be confused pathologically with regard to fine needle aspiration biopsy. In such superficial or ulcerated lesions, a definitive diagnosis can be made with a Tru-cut biopsy or incisional biopsy. In this way, an unnecessary and extensive surgery can be avoided. However, if the lesion is a malignant tumor, care should be taken, as incisional biopsy will affect postoperative survival.

Ethics Committee Approval: This study was approved by Erciyes University Clinical Research Ethics Committee (Date: 14.09.2022, No: 2022/615).

Informed Consent: Written informed consent was obtained.

Peer Review: Externally peer-reviewed.

Author Contributions: Conception/Design of Study- İ.Y., E.S.; Data Acquisition- E.S.; Data Analysis/Interpretation- A.V.; Drafting Manuscript- İ.Y., E.S.; Critical Revision of Manuscript- S.Ç., Ö.C.; Final Approval and Accountability- İ.Y., A.V., E.S.; Material or Technical Support- Ö.C.; Supervision- Ö.C.

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