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A 12-Year Clinical Experience with Pilomatrixomas in the Head and Neck Region

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ABSTRACT

Background: Pilomatrixoma is a superficial, benign skin tumor differentiating from the hair follicle matrix. More than half of these tumors occur in the head and neck region. This study aims to share the clinical experience with patients diagnosed with pilomatrixoma in the head and neck region through surgical excision, to raise awareness of differential diagnosis, and to contribute to the literature.

Methods: The data of patients diagnosed with pilomatrixoma through surgical excision in the Otorhinolaryngology Clinic of Dokuz Eylül University Hospital between January 2009 and July 2021 were retrospectively analyzed. The demographic data of the patients, presenting complaints, tumor location and pathological size, imaging methods used, surgical procedures performed, and postoperative follow-up characteristics were evaluated.

Results: Between January 2009 and July 2021, twelve patients underwent surgery in the clinic and were diagnosed with pilomatrixoma. Seven of these patients were male (58.3%), and 5 were female (41.7%), with a mean age of 23.75 years, and a median age of 15 years. Seven of the twelve patients (58.3%) were under 20. Nine patients presented with a firm, painless mass covered by normal skin in the preauricular region, 2 in the auricle, and 1 in the postauricular region.

Conclusion: Pilomatrixoma is a rare condition that should be considered in the differential diagnosis of head and neck masses. The treatment is complete excision of the lesion, as failure to excise it may result in a very low risk of malignant transformation. The importance of complete excision in preventing recurrences should not be overlooked.

Keywords: Malherbe epithelioma, pediatric, pilomatrixoma, skin tumor, surgery

Introduction

Pilomatrixoma is a superficial, benign skin tumor that differentiates from the hair follicle matrix.[1] Malherbe and Chenantais first described it in 1880 as a calcified epithelioma originating from sebaceous glands, known as "Malherbe's calcified epithelioma."[2] In 1961, Forbis and Helwig established that it originates from the hair follicle cortex and proposed the term pilomatrixoma.[3]

Pilomatrixoma commonly occurs in childhood. More than 60% of cases appear within the first 20 years of life, with a slightly higher incidence in females than in males.[4] It accounts for approximately 0.12% of skin tumors.[4] The most frequent anatomical location is the head and neck region, with 40%-77% of cases reported in this area.[5] The second most common site is the upper extremities, followed by the trunk and lower extremities.[5] The most common location in the head and neck region is the preauricular area.[6] No cases have been reported in these areas since hair follicles are absent in the palms and soles.[7] Malignant transformation is rare and, when it occurs, is more frequently observed in middle-aged and elderly patients (fifth to seventh decades) and in males.[8,9]

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Clinically, pilomatrixoma presents as a firm, slow-growing, painless, mobile subcutaneous nodule with well-defined borders.[10,11] The overlying skin is usually of normal color, but a bluish hue or telangiectasia may be present.[12] It typically appears as a single lesion measuring less than 3 cm, though larger lesions can occur.[13] Diagnosis can be made via physical examination, imaging, and cytology; however, misdiagnosis is possible due to a lack of awareness among clinicians.[14,15] The definitive diagnosis is based on histological findings, which reveal islands of epithelial cells extending from the dermis into the subcutaneous fat. These islands consist of basophilic cells, ghost cells (enlarged eosinophilic epithelial cells with no nucleus), occasional foreign body giant cells, and calcifications.[16]

This study aims to retrospectively review patients diagnosed with pilomatrixoma who underwent surgery in the clinic, share the clinical experiences, and contribute to the literature.

Material and Methods

The data of patients diagnosed with pilomatrixoma through surgical excision in the Otorhinolaryngology Clinic of Dokuz Eylül University Hospital between January 2009 and July 2021 were retrospectively analyzed. The demographic data of the patients, presenting complaints, tumor location and pathological size, imaging methods used, surgical procedures performed, and postoperative follow-up characteristics were evaluated. Prior to the commencement of the study, ethical approval was obtained from the Non-Interventional Research Ethics Committee of Dokuz Eylül University (Decision No: 2022/07-19, dated February 23, 2022). As the study was of a retrospective methodological nature, informed consent was not required from the patients. However, consent was obtained from those whose surgical images were utilized.

Results

Between January 2009 and July 2021, twelve patients underwent surgery in the clinic and were diagnosed with pilomatrixoma. Seven of these patients were male (58.3%), and 5 were female (41.7%), with a mean age of 23.75 years (19.6 years in females and 28.1 years in males), and a median age of 15 years (ranging from 2 to 62 years). Seven of the twelve patients (58.3%) were under 20 years old. Nine patients presented with a firm, painless mass covered by normal skin in the preauricular region, 2 in the auricle, and 1 in the postauricular region. In 8 cases (66.6%), the lesion was located on the left side, while in 4 cases (33.3%), it was on the right side. Imaging was performed in 8 patients: ultrasonography (USG) in 2, computed tomography (CT) in 3, and magnetic resonance imaging (MRI) in 3, while 4 patients underwent no imaging studies. None of the patients had a preoperative fine-needle aspiration biopsy. Partial superficial parotidectomy was performed in 3 patients due to the proximity of the lesion to the parotid gland. The mean follow-up period was 15 months. No postoperative complications or recurrences were detected, and no cases of pilomatrix carcinoma were observed. The demographic characteristics of the cases, the location of the lesion, and the mass sizes obtained from pathology reports are presented in Table 1.

A study found the lesion on the left side in 8 of the twelve cases (66.6%) and on the right side in 4 cases (33.3%). Regarding the surgical procedure, a simple mass excision was performed in 9 patients, while a wide surgical excision with partial superficial parotidectomy was planned for 3 patients due to the adhesion of the mass to the parotid gland. Figure 1. shows the simple excision of Case 1, who presented with swelling in front of the left ear, as indicated in Table 1, while Figure 2. shows the wide surgical excision of Case 8.

The mean follow-up period of the patients after surgery was 15 months. No complications or recurrences were detected in the patients after the operation, and no patient had pilomatrix carcinoma.

Discussion

Pilomatrixoma is a benign neoplasm that differentiates from the hair follicle matrix cells, more commonly seen in children and adolescents.[10] Data on ethnic variations are limited, but a slight female predominance typically appears in the first and second decades of life.[10] A study by Julian et al[17] reported 2 distinct age peaks: a significant peak occurring between 0 and 20 years and a secondary, less pronounced peak between 50 and 65 years. These lesions typically present as firm, slow-growing, painless masses that develop months or even years before diagnosis.[12] They usually adhere to the overlying skin but are freely mobile over underlying structures with well-defined borders. The overlying skin may exhibit a bluish hue or telangiectasia.[12] Graham and Merwin described the "tent sign," where the stretched skin over the tumor reveals its firm structure and allows palpation.[18] Most lesions measure less than 3 cm in diameter.[5,10] While they are generally solitary nodules, multiple lesions have been reported in 2% to 10% of cases.[16]

Preoperative misdiagnosis of pilomatrixoma is common, with an initial diagnostic accuracy rate often below 50%.[12] The differential diagnosis includes epidermoid cysts, dermoid cysts, branchial remnants, atypical mycobacterial infections, parotid gland lesions, preauricular sinuses, ossified hematomas, giant cell tumors, chondromas, and foreign body reactions.[19] Therefore, when operating on a suspected pilomatrixoma, consideration of alternative diagnoses is essential.

When clinical diagnosis is uncertain, adjunct imaging techniques such as direct radiography, USG, CT, or MRI can be employed.[20] Imaging is

Table 1. Gender, Age, and Tumor Size of Pilomatrixoma Patients

Case	Gender	Age	Tumor Location	Tumor Size
1	Female	2	Left preauricular area	1 × 0.8 × 0.6 cm
2	Female	56	Left preauricular area	1 cm
3	Male	34	Left preauricular area	2.5 × 2 × 1 cm
4	Male	35	Left preauricular area	2 × 1 cm
5	Male	29	Right preauricular area	3 x× 1.6 × 1.8 cm
6	Male	9	Left preauricular area	2,1 × 1.8 × 1 cm
7	Male	13	Left preauricular area	2 × 1,5 cm
8	Female	15	Right preauricular area	2 × 1 × 1 cm
9	Female	6	Left ear lobule	1.2 × 0.8 × 0.5 cm
10	Male	15	Left preauricular area	1.2 × 1 × 1 cm
11	Male	62	Right ear helix	3.5 × 2 × 1.5 cm
12	Female	9	Right postauricular area	1 × 0.6 × 0.4 cm

MAIN POINTS

- *Pilomatrixoma is most commonly seen in the head and neck region.*
- *It should be considered in the differential diagnosis of head and neck masses.*
- *Complete excision of the lesion is important to prevent recurrences.*

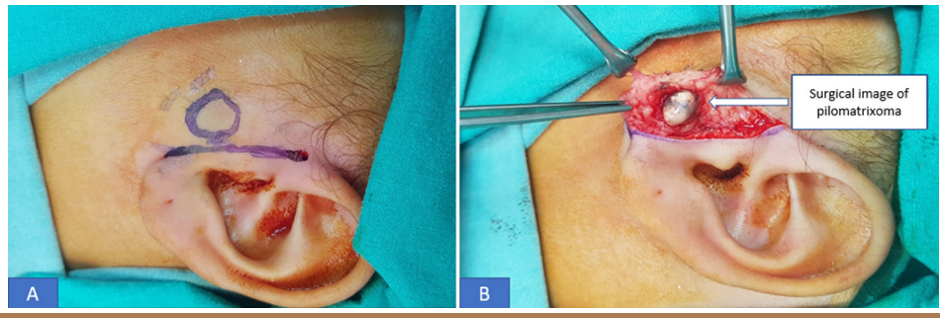

Figure 1. A. Patient image before surgery. B. Image of the mass during surgery marked with a white arrow.

particularly useful in distinguishing preauricular masses from parotid tumors or in assessing large, aggressive lesions. Among these modalities, USG is the most commonly used for pilomatrixoma diagnosis.[10] Studies have demonstrated that ultrasonography is valuable in ruling out the invasion of the parotid gland.[10] Pilomatrixoma typically appears on USG as a well-defined, oval, hypoechoic heterogeneous mass at the dermal-subcutaneous junction, often containing echogenic foci.[21] On CT, pilomatrixomas present as well-circumscribed subcutaneous masses with various degrees of calcification and mild to moderate contrast enhancement.[12] On MRI, they appear as well-defined soft tissue masses with homogeneous intermediate signal intensity on T1-weighted images and heterogeneous intermediate-to-high signal intensity on T2-weighted images.[22]

Due to the lack of spontaneous regression, complete surgical excision is the definitive treatment. Pilomatrixomas are often found in cosmetically sensitive areas, and incomplete excision almost always results in recurrence.[20]

In the study, regarding gender distribution, 5 of the 12 patients were female. Seven were male (female-to-male ratio: 1:1.4). Jones et al[10] reported a slight female predominance (female-to-male ratio: 1.15:1). At the same time, O'Connor et al[20] found no significant difference in incidence between the sexes (female-to-male ratio: 1.1:1). The discrepancy in gender distribution observed in the study needs further validation through a larger case series.

The mean age of the patients was 23.75 years (19.6 years in females, 28.1 years in males), and the median age was 32 years (ranging from 2 to 62 years). Seven of the 12 patients (58.3%) were under the age of 20. Jones et al[10] reported that 55% of cases occur in individuals younger than 20 years. Yencha et al[5] found this proportion to be 60%, whereas O'Connor et al[20] reported 37%, also identifying a secondary peak in incidence between the fifth and seventh decades of life.

Regarding lesion size, 11 out of 12 cases had a maximum lesion diameter of less than 3 cm, with 1 case measuring 3.5 cm. Lesion sizes ranged from 4 mm to 35 mm. All cases were single, solitary nodules, with no instances of multiple nodules. Jones et al[10] reported lesion sizes ranging from 4 to 20 mm, while Yoshimura et al[13] found sizes between 5 and 30 mm. Lin et al[23] reported a range of 4 to 31 mm, with an average lesion size of 13 mm.

In the study, 9 of the 12 lesions were located in the preauricular region, 2 in the auricle, and 1 in the postauricular region. Yoshimura et al[13] identified the preauricular area as the most common site for head and neck pilomatrixomas (58%). Guinot-Moya et al[24] reported that more than 50% of cases occur in the head and orofacial regions, particularly in the parotid region.

It was found that 8 cases involved lesions on the left side, whereas 4 were on the right. Jones et al[10] reported that pilomatrixomas were more frequently located on the right side of the body.

To assess differential diagnoses and tumor characteristics preoperatively, USG was performed in 2 patients, CT in 3, and MRI in 3, while 4 patients underwent no imaging. The choice of imaging modality depended on the evaluating physician's preference. In this study, USG had a diagnostic accuracy of 50%, which aligns with the 47% accuracy rate reported by Jones et al.[10] Computed tomography and MRI findings were consistent with pilomatrixoma in all cases.

All patients in this study underwent complete excision of their lesions, and no recurrences were detected. Three patients required partial superficial parotidectomy due to tumor adhesion to the parotid gland. Studies have consistently emphasized the importance of complete excision to prevent recurrence.[1,25] If the tumor adheres to surrounding tissues or has poorly defined borders, a 1-2 cm margin of excision has been recommended to minimize local recurrence risk.[25] Guinot-Moya et al[24] reported a recurrence rate of 0.48%, while

Figure 2. Surgical images of pilomatrixoma in close relation to the parotid gland.

Danielson-Cohen et al[26] documented a 4% recurrence rate following complete excision.

Malignant transformation (pilomatrix carcinoma) is exceedingly rare but is more frequently reported in head and neck pilomatrixoma, particularly in males and older patients.[9,27] No cases of pilomatrix carcinoma were identified in this study.

Although pilomatrixoma is rare, it tends to occur in the head and neck region and should be considered in the differential diagnosis of head and neck masses. The treatment is surgical excision, and complete excision is crucial to prevent recurrence.

Data Availability Statement: *The data that support the findings of this study are available on request from the corresponding author.*

Ethics Committee Approval: *This study was approved by Ethics Committee of Dokuz Eylül University (Approval No.: 2022/07-19; Date: 23.02.2022).*

Informed Consent: *Written informed consent was obtained from those whose surgical images were utilized.*

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