ORIGINAL ARTICLE

Clinical Diagnostic Challenges of Pilomatricoma: Retrospective Study From Southern Region of Saudi Arabia

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ABSTRACT

Objective: To analyze the clinicopathological features of PMC, including the patient age at clinical presentation, gender, anatomical location, tumor size, preoperative clinical and histopathologic diagnosis. **Methods:** This is a retrospective study to analyze all cases of PMC diagnosed in two hospitals from August 2008 to January 2017 in Southern Region, Saudi Arabia. Detailed medical records and histopathologic material review were done for all cases of pilomatricoma. Cases were identified using the search engine of each hospital electronic filing system. **Results:** 23 cases with an age range from 8 to 68 years (mean 28.8). The majority of patients were in their second (39%) and fourth (26%) decades of life with a female to male ratio of 1.2 to 1.1. Majority of the cases were noted in the neck (43.5%), followed by head region (30%). The tumor size ranged from 0.3 to 4.0 cm (mean 1.6). The most common preoperative clinical misdiagnosis was sebaceous cyst (34.8%). All cases were confirmed histopathologically as pilomatricoma. All tumors were excised surgically. **Conclusions:** PMC has a wide variety of clinical presentations and is commonly misdiagnosed with other dermatological conditions. PMC should be included in the differential diagnosis of nodules, particularly in the head and neck region. Increasing the clinical awareness and familiarity with the tumor may lead to accurate clinical diagnosis.

Keywords: Pilomatricoma, benign, adnexal skin, neoplasm

INTRODUCTION

Pilomatricoma is a benign adnexal skin tumor of follicular origin. It was first described in 1880 by Malherbe and Chenantais,[1] who proposed that this tumor was derived from subcutaneous sebaceous glands; therefore the lesion was termed as" calcifying epithelioma of Malherbe". Later in 1922, Dubreuilh & Cazenave,[2] described the morphological features of this lesion, including shadow cells and nests of epithelial cells. In 1961, Forbis and Helwig[3] modified the term epithelioma into pilomatrixoma which was modified later due to phonetic reasons to pilomatricoma.[4] The lineage of differentiation was first recognized by Turhan and Krainer[5] to be derived from hair cortex cells, this finding was confirmed in 1966 by Hashimoto[6] through histochemical and electron microscopic studies.

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Pilomatricoma often present clinically as solitary firm slow-growing and painless subcutaneous nodule, the overlying skin can appear normal or bluish; however skin ulceration may occur due epidermal thinning.[7] They tend to occur in children and young adults, show predilection for the head and neck[8] and are rarely reported in the maxillofacial literature.[9] The diagnosis can be rendered initially by cytological examination of fine needle aspiration;[10] however, the definite diagnosis is confirmed by histopathologic examination. The clinical awareness about PMC is variable, and the published national data of PMC is scant. The purpose of this study is to analyze the clinicopathological data from the author's experience with pilomatricoma from the two regional hospitals including Armed Forces Hospital in the Southern Region, and Abha International Private Hospital and to compare our data to the previously published studies.

MATERIALS AND METHODS

The author retrospectively reviewed the surgical pathology files in the Department of Pathology of Armed Forces Hospital Southern Region (AFH-SR) and the Division of Surgical Pathology in Abha International Private Hospital (AIPH), Southern Region, Saudi Arabia, and searched for all cases of pilomatricoma diagnosed during the period of August 2008 to January 2017. The records of the study group were reviewed to determine the age, gender, anatomical location and clinical presentation. The morphological findings were reviewed.

RESULTS

A total of 23 cases were included in our series. The age at the time of diagnosis ranged from 8- 68 years (mean 28.8). The majority of patients were their second (39%) and fourth (26%) decades of life with a male to female ration of 1.1 to 1.2. Most of the cases were noted in the neck (43.5%), followed by head region (30%), upper limb (13%), then lower limb, chest and back (4.3%) each, (Table 1).

The tumor size clinically ranged from 0.3 to 4.0 cm in greatest dimension (mean 1.6). Majority of the cases presented clinically as a mass nodular lesion in (47.8%), asymptomatic lesion in (39%), painful lesion in (8.7%) and repeated infected lesion in (4.3%), (Table 2). Preoperative radiological studies were done in five cases (21.7%), and calcification was detected in two cases (8.7%) only.

Table 1: Accuracy of Clinical Diagnosis of Pilomatricoma

Clinica Diagnosis	Overall No. (%)
Sebaceous Cyst	**8** (34.7)
Dermoid Cyst	**2** (8.7)
Lymphadenopathy	**2** (8.7)
Parotid gland tumor	**1** (4.3)
Lipoma	**1** (4.3)
Calcified Mass	**1** (4.3)
Foreign-body Granuloma	**1** (4.3)
Atypical Hemangioma	**1** (4.3)
Pilomatricoma	**1** (4.3)
No Clinical Diagnosis	**5** (21.7)
Total	**23**

Table 2: Anatomical Distribution of Pilomatricoma

Anatomical Region	Overall No. (%)
Head Region	**10** (43.5)
Postauricular	**3**
Periauricular	**2**
Eyebrow	**2**
Scalp	**2**
Infraorbital	**1**
Neck Region	**6** (26)
Arm	**3** (4.3)
Back	**2** (8.7)
Chest Wall	**1** (4.3)
Calf	**1** (4.3)
Total	**23** (100)

The most common preoperative clinical misdiagnosis was sebaceous cyst (34.8%), followed by dermoid cyst (8.7%), lymphadenopathy (8.7%), parotid tumor (4.3%), lipoma (4.3%), foreign body granuloma (4.3%), atypical hemangioma (4.3%), and calcified mass (4.3%). No preoperative clinical diagnosis was documented in (21.7%).

The correct preoperative clinical diagnosis of pilomatricoma was documented only in one case

(4.3%). None of the cases in our series were investigated by fine needle aspiration (FNA).

Microscopic examination showed well-defined tumor composed of solid nests of basaloid cells at the periphery, and trichilemmal-type keratinization in the central zone, and associated with ghost cells and foreign body reaction (Figure 1). Calcification was noted in (13%) of cases and ossification was observed in (6%) of cases.

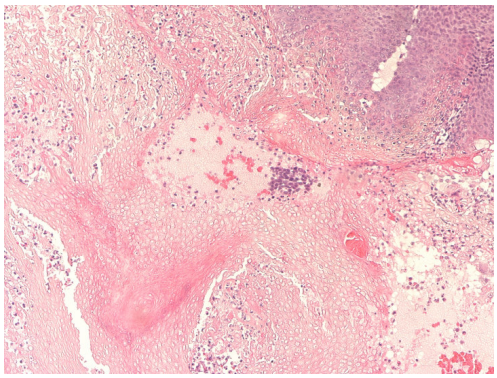

Figure 1: Photomicrograph showing pilomatricoma, with peripheral basaloid cells right upper corner, and central ghost cells. (Original magnification, hematoxylin and eosin stain, ×40).

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All cases underwent surgical excision with minimal margins. The closest resection margin was 1.0 mm. The follow up ranged from 8 to 85 months.

DISCUSSIONS

Pilomatricoma is an adnexal skin neoplasm, which manifests clinically as asymptomatic slowly growing mobile hard subcutaneous nodule.[9] The overlying skin is often normal; however, in some cases ulceration and bluish discoloration can occur. In our series pain was found in two cases (8.7%), and ulceration was found in one case (4.3 %). No clinical diagnosis was provided by the clinical physicians in five cases (21.7%), however eight cases (34.7%) were misdiagnosed clinically as sebaceous cyst, two cases (8.7%) were misdiagnosed as dermoid cysts, two cases (8.7%) were misdiagnosed as lymph node lesions, and four cases (21.7%) were misdiagnosed as other clinical entities (Table 3). Only one case (4.3%) was correctly diagnosed preoperatively as pilomatricoma.

The low index of clinical suspicion of PMC could be related to the varying clinical presentation, low incidence, and possible lack of awareness of the neoplasm by the clinicians. Moehlenbeck[11] studied 140.000 cases of skin tumors, and found that pilomatricoma represented for only 0.1%. The correct clinical preoperative diagnosis could be made in (28%), as reported by O' Connor et al.[9] Kumaran N et al.[12] noted that accuracy of the preoperative clinical diagnosis could reach up to 46 %. In our series the correct preoperative clinical diagnosis was (4.3%) only.

Table 3 Clinical Presentations of Pilomatricoma

Clinical Findings	Overall No. (%)
Pain	**2** (8.7)
Mass	**11** (47.8)
Repeated infections	**1** (4.3)
Asymptomatic	**9** (39.1)
Total	**23** (100)

Though PMC could occur at any age, our study demonstrated bimodal peak at the second decade (34.7%), and in fourth decade patients (26%). These ages of presentation were different from the previous published reports,[9,13] which showed that the PMC commonly presented in the first and second decades.

The most common site of these tumors was head and neck (56.5%) which was comparable to the reported studies.[7-9] The female to male ratio of PMC in our study was 1.2 to 1.1 which was slightly different from that in other reports.[13-15]

Majority of these tumors are solitary. Multiple tumors were reported in 2-10% of previous studies[13,14,16] and were found to be associated with Gardner syndrome, Steinert disease, myotonic dystrophy, and sarcoidosis. All cases included in our study were solitary and none of which was associated with any syndrome.

The imaging studies such as ultrasound, computerized tomography (CT) scan, magnetic resonance imaging (MRI) were rarely used in the routine preoperative workup of PMC. They could be helpful to detect the calcification if present and could increase the accuracy of preoperative clinical diagnosis. In our series, preoperative imaging studies were requested by the clinicians in five cases (21.7 %), and calcification was reported in two cases (8.7%) only (Figure 2).

The utilization of FNA could variably permit to diagnose PMC; however the inherited pitfall for FNA was overinterpretation of PMC as malignant tumor[10,17,18] due to nuclear pleomorphism, increased nuclear cytoplasmic (N:C) ratio, and prominent nucleoli. The typical cytological findings of PMC included presence of basaloid cells, ghost or shadow cells, squamoid cells, foreign body multinucleated giant cells, calcium deposit and mixed inflammatory cells. No FNA was performed in our series.

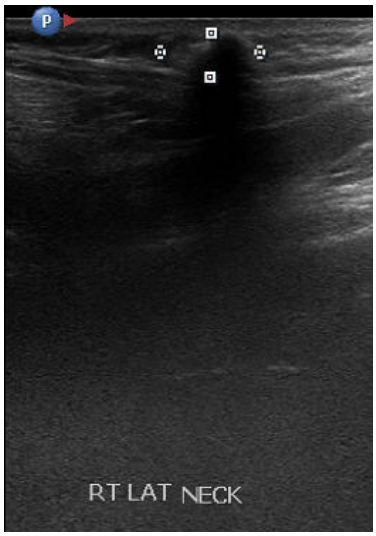

Figure 2: Ultrasound image of the right postauricular region showing superficial echogenic 1.4 x 6.0 mm nodular soft tissue lesion with calcification.

The diagnosis is confirmed by histopathologic examination which showed well defined tumor in the deep dermis and subcutaneus tissue and composed of nucleated basaloid cells arranged peripherally and enucleated shadow or ghost cells centrally. Cystic change usually occurs in early lesions, while older lesions became solid with shadow cells, multinucleated cells, keratin derbies and dystrophic calcification which was reported in 69 % to 85% of cases.[16,19] On microscopic examination of the cases in our series, we detected dystrophic calcification in (8.7%), osseous metaplasia in (6%), and foreign body giant reaction in (4.3 %). The osseous metaplasia was due to the metaplasia of fibroblast into osteoblast.[20] Junctional benign melanocytic nevus was observed as an additional pathologic finding in one case.

Malignant transformation of PMC is rarely reported in the literature, with approximately 80 cases reported.[9] Pilomatrix carcinoma was reported to occur most commonly in elderly men in head and neck regions. None of the cases in our series was malignant. No local recurrence was found in our series.

CONCLUSION

PMC has a wide variety of clinical presentations and is commonly misdiagnosed with other dermatological conditions. PMC should be included in the differential diagnosis of nodules, particularly in the head and neck region. Increasing the clinical awareness and familiarity with the tumor may lead to accurate clinical diagnosis.

ACKNOWLEDGMENT

We acknowledge the Medical Records, Radiology & Pathology Departments for their cooperation to conduct this study.

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