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PILOMATRICOMA: A RETROSPECTIVE STUDY

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Objectives: *The purpose of this study is to review the clinical and histologic features of pilomatricomas diagnosed in our department.*
Materials and Methods: *In this study, 32 patients with pilomatricoma diagnosed at Pathology Department of Dicle University Medical Faculty, during the period 1984-1999 are presented.*
Findings: *The mean age was 27.43 years and the majority of the cases occurred in the third decade of life. One of our cases was perforating pilomatricoma. Histopathologically, the most important feature of pilomatricoma was shadow cells (ghost cells). We observed that the most common sites were head, neck and upper limb.*
Conclusions: *Pilomatricoma can be easily confused clinically with other lesions.*

Key words: *Skin, pilomatricoma, calcifying epithelioma of malherbe*

Pilomatrikoma: Bir retrospektif çalışma

Amaç: *Anabilim Dalı'mızda pilomatrikoma tanısı alan 32 olgunun histolojik ve klinik özelliklerini gözden geçirmeyi amaçladık.*
Materyal ve Metot: *Bu çalışmada, 1984-1999 yılları arasında Dicle Üniversitesi Tıp Fakültesi Patoloji Anabilim Dalı'nda pilomatrikoma tanısı alan 32 olgu sunuldu.*
Bulgular: *Ortalama yaşı 27.43 olup, olguların çoğunluğu üçüncü dekaddaydı. Olgulardan birinde perforasyon izlendi. Histopatolojik olarak pilomatrikomanın en önemli özelliği hayalet hücreleridir. Baş, boyun ve üst ekstremitelerin en sık yerleşim yeri olduğunu izledik.*
Sonuç: *Klinik olarak pilomatrikoma diğer lezyonlarla kolaylıkla karışabilmektedir.*

Anahtar kelimeler: *Deri, pilomatrikoma, kalsifiye epitelioma, malherbe*

Pilomatricoma, or calcifying epithelioma of Malherbe, is a tumor with differentiation towards hair cells, particularly hair cortex cells.[1] Pilomatricoma is a benign adnexal tumor.[2] Most commonly, it manifests itself as a firm, deep-seated nodule that is covered by normal skin.[3] Pilomatricoma presents usually as a solid lesion. Their most common site is the head and neck region.[1,4,5] Generally, the tumor varies in diameter from 0.5 to 3 cm. The tumors may arise in patients of any age, but usually appear in first three decades of life.[3,4] Rare familial occurrence and association with myotonic dystrophy has been reported.[3,6,7] It usually presents itself as a hard dermal or subcutaneous tumor.[8] The controversy regarding its histogenesis is indicated by the variety of names given to it in the past;ie., calcifying epithelioma of Malherbe, mummified epidermal cysts, mummified tumor of Malher-be, and trichomatricoma.[9]

The purpose of this study is to review the clinical and histologic features of pilomatricomas diagnosed at our department.

MATERIALS AND METHODS

We reviewed files and histologic sections of 32 patients with pilomatricoma diagnosed at the Department of Pathology, University of Dicle, Diyarbakir, Turkey, between 1984 and 1999. The entire slides stained with Hematoxylin and eosin were analysed using an Olympus BH-2 microscope. All histologic features observed were noted regardless of their intensity. The clinical features and relevant data were also retreived from the files of the patients.

RESULTS

During the period 1984-1999 a total of 32 cases of pilomatricoma were diagnosed. Eighteen cases were female and 14 cases were male. The mean age was 27.43 years. The female to male ratio was 1,29 and 40.62% of cases occurred in the third decade.Of 32 cases, 29 had information about location. The distribution of the lesions was as follows: 12 (41%) on the head and neck, 13 (45%) on the upper limbs and upper trunk, and 4 (14%) on the lower limbs and lower trunk.

The presenting features were stony hard dermal or subcutaneous swellings in twenty-two cases. All cases were initially seen by surgeons. A variety of clinical diagnoses are given in Table 1.

Table 1. Clinical diagnosis prior to biopsy.

Clinical Diagnosis (N=32)	No. of cases	%
Swelling	22	68,75
Sebaceous cyst/epidermoid cyst	7	21.88
Pilomatricoma	2	6.25
Lymph node	1	3.12

The biopsy sent to The Pathology Department were of variable sizes ranging from a tiny fragment to as large as 6 cm x 3 cm x 2 cm. Since the majority of lesions were probably curetted out, skin was attached to the tumor masses in only 12 cases.Ghost cells (shadow cells) were present in all thirty-two cases. Giant cells were seen in the stroma in 20 cases, and other types of inflammatory cells were seen in 23 lesions. Hemosiderin pigment was found in the stroma (5 cases) close to blood vessels. However, only one case had ossification, but not extramedullary hemato-poiesis. All main histopathologic elements are summarized in Table 2.

Table 2. Histologic features of pilomatricoma

Histologic features	No. of cases	%
Ghost cells	32	100
Stromal inflammation	23	71.87
Foreign body giant cell	20	62.5
Calcification	18	56.25
Basaloid cells	15	46.87
Capsule formation	13	40.62
Stromal fibrous	9	28.12
Squamoid cells	6	18.75
Keratin pearl formation	6	18.75
Hemosiderin pigment	5	15.62
Osteoid tissue	1	3.12

We present a photograph of pilomatricoma showing basaloid, foreign body giant and ghost cells (Fig. 1).

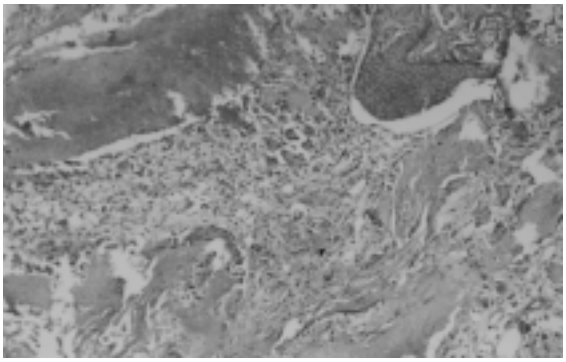

Fig. 1. A photograph of pilomatricoma showing basaloid, foreign body giant and ghost cells (H.EX200).

Capsule formation was present in only thirteen of the 32 cases. 12 cases were found in the dermis tissue, but the cutaneous levels of the others were uncertain. One of our cases presented itself as perforating pilomatricoma. She was a 43 old-age woman, with a history of bleeding mass in her face, which had ulcerated. There was not any patient with a positive familily history or multiple lesions.

DISCUSSION

Pilomatricoma is a benign cutaneous uncommon neoplasm with differentiation toward hair matrix.[3,5,10] There is a general consensus that the tumor cells differentiate towards hair cortex cells especially in light of studies by electron microscopy[11] and immunohisto-chemistry.[12]

Pilomatricomas appear at any age, but usually are seen in first three decades of life;also, have second peak incidance in fifth and sixth decades.[1,5,13] Julian et al. [4] also reported pilomatricomas with peak presentation bimodally in the first and sixth decade. In our series, 40.62% of tumors did occurred in the third decade of life. We did not have any cases in the elderly (>70 years);this is in contrast to the findings of Taaffe et al. [1] who have reported 5,1 % cases in this age group. We compared the age distribution with Ahmad [13] and Taaffe (1) in Fig. 2.

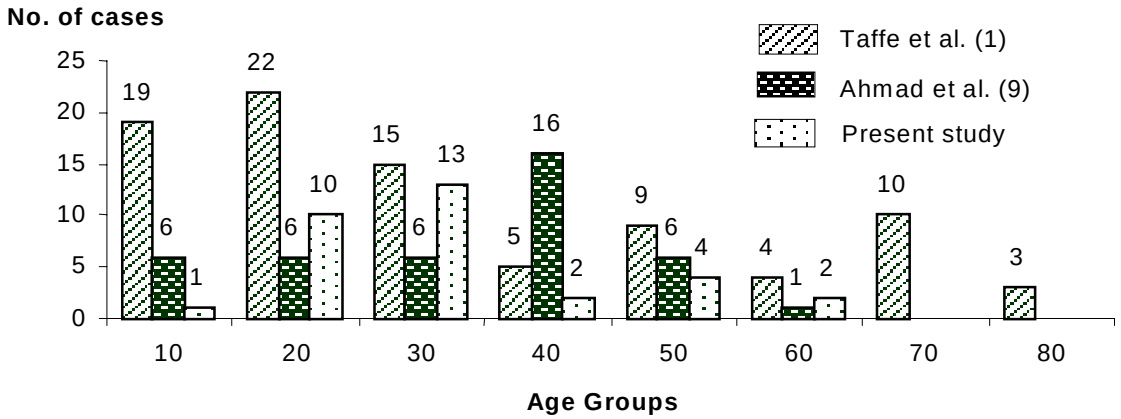

Fig. 2. Distribution of age at onset of pilomatricoma in different studies.

Familial occurrences are usually accepted as an incidental event. In the literature there are only a few instances of familial occurence reported, and in some of these cases the tumor is associated with myotonic dystrophy.[3,6,7,14] Althogh we did not have any cases, Demircan [14] et al. had two patients out of 15 with familial occurrence. Pilomatricoma usually occurs as a stony hard dermal or subcutaneous nodule, mostly on the head, neck and upper extremities [1,3,4,8,9,13,14] In our study, of 32 cases, 29 had information about location. The site distribution of our cases correlates with those described earlier[1,13] (Table 3).

Because pilomatricoma is confused with the other lesions, it is often misdiagnosed clinically.[1] All of the surgical specimens were sent to our Department by surgeons. Only two cases (6.25%) had correct clinical diagnosis. This rate was 3.77% in Ahmad's [13] report. One of our cases was a perforating pilomatricoma that is rare.[13,16] It occurred in a 43 old-age woman, with a history of bleeding mass in her face, which was ulcerated. The estimated incidence for multiple pilomatricoma is 3.5%.[14] However, we did not have any multiple pilomatricoma case. Histopathologic features

Table 3. Comparative analysis of site distribution.

	Head & Neck (%)	Upper Limb & Upper Trunk (%)	Lower Limb & Lower Trunk (%)
Taaffe (1) et al. (n=78)	64	26	10
Ahmad (9) et al. (n=45)	38	44	18
Gündoğdu (15) at el. (n=17)	53	29	18
Present study (n=29)	41	45	14

Table 4. Comparative analysis of the histologic features of pilomatricoma.

	Our study		Ahmad et al. (9)		Taaffe et al. (1)		Gündoğdu et al. (15)	
Histologic features	No. of cases	%	No. of cases	%	No. of cases	%	No. of cases	%
Ghost cells	32	100	53	100	78	100	14	82,4
Stromal inflammation	23	71.87	29	54.71	77	98.71	15	88,2
Foreign body giant cell	20	62.5	28	52.83	75	96.15	13	76,5
Calcification	18	56.25	32	60.37	29	37.17	15	88,2
Basaloid cells	15	46.87	20	37.73	51	65.18	-	-
Capsule formation	13	40.62	10	18.86	25	32.05	-	-
Squamoid cells	6	18.75	29	54.71	40	51.28	2	11,8
Stromal fibrous	9	28.12	-	-	70	89.74	15	88,2
Keratin pearl formation	6	18.75	7	13.20	-	-	-	-
Hemosiderin pigment	5	15.62	-	-	30	38.46	3	17,6
Osteoid tissue	1	3.12	1	1.88	6	7.69	1	5,9
Immature hair follicles	-	-	1	1.88	1	1.28	-	-

of pilomatricoma are typical. Irregularly shaped islands of epithelial cells are present. As a rule, two types of cells, basophilic cells and shadow cells (ghost cells), compose the islands. Histopathologically the most important feature is shadow cells.[3,9] Pilomatricomas have different histologic features from case to case. There are comparative analyses of the histologic features of pilomatricoma with different series including ours in Table 4. The rates of ossification range from 1% to 15% in some studies.[1,2,13]. Kaddu et al. [17] reported on seven adult patients with pilomatricomas showing histopathologic findings of extramedullary hematopoiesis. Only one of our cases had ossification, but not extramedullary hematopoiesis. In conclusion, pilomatricoma presents itself clinically as a stony hard dermal nodule within skin, and correct clinical diagnosis is hardly possible. For this reason, when such a lesion is present in patients, pilomatricoma must be considered clinically. Histopathologically the most important feature for diagnosis is shadow cells (ghost cells).

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