Clinicopathological study of pilomatricoma

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ABSTRACT

Objective: To study the clinical and histopathologic spectrum of pilomatricoma, the benign tumor of hair matrix.

Methods: Retrospective review of 27 cases of pilomatricoma (Calcifying epithelioma of Malherbe) reported at Bahrain Defence Force Hospital from 1993 – 1999.

Results: Most of the cases were confused clinically with sebaceous cysts. Seventy eight per cent of the cases occurred below the age of 30 years. Female to male ratio was 5:4. Head, neck and upper limb were the most common sites for pilomatricoma. The size of the tumors ranged from between 4-35 mm in diameter. Tumors were encapsulated and solid composed of shadow and basophilic cells, and stroma containing varying amounts of calcification, ossification and inflammatory cells.

Conclusion: Pilomatricomas have a wide variety of clinical characteristics and are often misdiagnosed with other skin conditions. They should be considered along with other benign and malignant conditions in the clinical differential diagnosis of solitary firm skin nodules especially those, which occur in the head, neck and upper limb.

Keywords: Pilomatricoma, skin appendage tumors.

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Pilomatricoma is a benign tumor of the epithelial hair matrix. The tumor was first described by Malherbe and Chenantais in 1880 as calcified epithelioma of sebaceous glands,[1] but later in 1942 it was recognized by Turhan and Krainer that the cells of the tumor differentiate in the direction of hair cortex cells.[2] This finding was subsequently confirmed by electron microscopic studies and immunohistochemistry examination. Pilomatricoma are found mainly on the head and the upper extremities, but may occur at any site. Although they tend to occur in children and young adults, pilomatricoma shows increase its incidence in older age group. Although pilomatricoma is not hereditary, there are a few reports of familial occurrence, and in some of these cases, the tumors are associated with myotonic dystrophy.[3] We report in this paper our experience along with the clinicopathological findings of 27 cases of pilomatricoma diagnosed in our institution.

Methods. The surgical pathology files in the histopathology laboratory in our institute were searched for pilomatricoma cases during the period 1993-1999. A total of 27 cases were found. Patients' data such as age and sex was recorded along with the clinical diagnosis. All cases were classified according to the site of the tumors in the different parts of the body. The size of the tumors was also categorized into three grades: Small 10 mm or less in diameter, medium size between 11-20 cm and large tumors above 20 mm in diameter.

Results. The clinicopatholgoical findings of all cases are summarized in Table 1. The age and sex distribution of the tumors are shown in Table 2. The female:male ratio was 5:4. The youngest patient was a one year old female and the oldest one was 47 years old male. Seventy eight per cent of the patients were below the age of 30 years. The most common site

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Table 1 - Clinicopathological findings of pilomatricoma.

No.	Age	Sex	Site	Clinical Diagnosis	Size
1	23	Male	Right pinna	Sebaceous Cyst	0.6 cm
2	21	Female	Right arm	Calcified Cyst	1.6 cm
3	2	Female	Right forearm	Sebaceous Cyst	1.5 cm
4	27	Female	Left arm	Lipoma	3.5 cm
5	4	Male	Retroauricle	Epidermal Cyst	0.6 cm
6	24	Male	Neck	Sebaceous Cyst	2.3 cm
7	32	Female	Forearm	Sebaceous Cyst	1 cm
8	13	Female	Right arm	Lump	0.4 cm
9	6	Female	Right eyebrow	Sebaceous Cyst	0.6 cm
10	32	Male	Upper back	Lump	1.3 cm
11	26	Male	Right thigh	Cyst	1 cm
12	12	Female	Left arm	Sebaceous Cyst	1.5 cm
13	34	Male	Left thigh	Sebaceous Cyst	3.5 cm
14	35	Male	Left arm	Cyst	1 cm
15	47	Male	Neck	Sebaceous Cyst	1 cm
16	9	Male	Neck	Lump	1 cm
17	13	Male	Left arm	Sebaceous Cyst	2 cm
18	13	Female	Left knee	Sebaceous Cyst	1.9 cm
19	29	Male	Right thigh	Lump	2 cm
20	4	Male	Left foot	Granuloma	1 cm
21	11	Female	Neck	Lump	1.2 cm
22	34	Female	Chest wall	Lump	0.7 cm
23	7	Female	Left cheek	Lump	2 cm
24	14	Female	Left arm	Sebaceous Cyst	1.5 cm
25	1	Female	Right ear	Inclusion Cyst	1 cm
26	12	Female	Left eye	Lump	1.7 cm
27	13	Female	Left leg	Sebaceous Cyst	0.4 cm

for the tumors was head and neck (10), followed by upper limb (9), lower limb (6) then the trunk (2). The size of the tumors was variable, 13 cases with 10 mm or less, 11 cases were 11-20 mm, 3 cases were over 20 mm in diameter. Most of the cases (93%) were less than 2 cm in diameter. Table 1 shows that most of the cases were diagnosed clinically as sebaceous cysts. Other diagnoses included lipoma, granuloma or unspecified lump.

Histopathology. The tumor is usually well defined and often surrounded by fibrous capsule. It is located in the dermis and extends into the subcutaneous fat. The tumor consists of two types of cells, basophilic cells and shadow cells (Figure 1). In some of the tumors the basophilic cells are absent. They are arranged either at one side or along the periphery of the tumor islands. The shadow cells have a distinct border and possess a central unstained area as a shadow of the lost nucleus. Small and large areas of calcification are sometimes seen in the stroma of the tumors. The stroma usually shows a considerable foreign body giant cell reaction adjacent to the shadow cells (Figure 2).

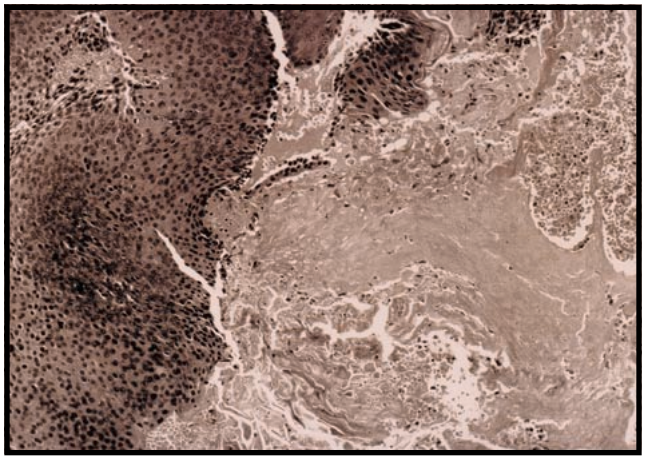

Figure 1 - The tumor consists of basophilic cells and shadow cells. the basophilic cells have deeply basophilic stained nuclei and scanty cytoplasm. The shadow cells in the center of the picture show shadow islands with unstained nuclei (hematoxylin-eosin).

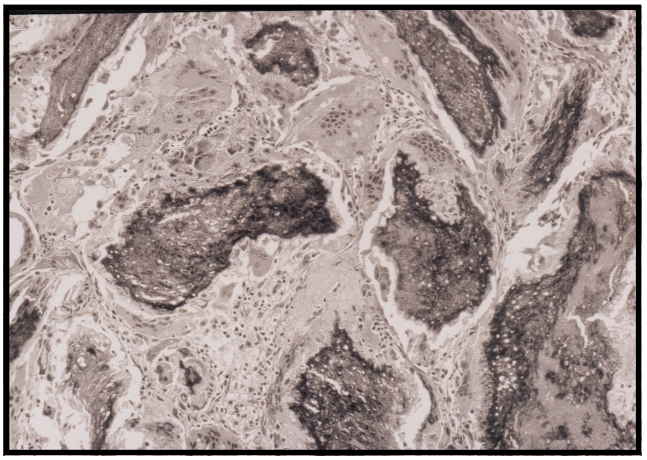

Figure 2 - Areas of calcification present within the lobules of tumor cells. Note the presence of inflammatory cells with multinucleated giant cells surrounding tumor islands (hematoxylin-eosin).

Discussion. Pilomatricoma was first described in 1880 by Malherbe and Chenantis as calcifying epithelioma of the sebaceous glands. The origin from the epithelial cells of hair matrix was confirmed later by electron microscopic and molecular biological studies. Expression of human hair keratin basic 1 (hHb1) in pilomatricoma provides evidence that these tumors can differentiate towards hair shaft keratinocytes.[4,5] Immunohistochemical localization of cytokeratins and involucrin in pilomatricoma was carried out by Watanabe et al from Japan who proved that the tumor cells were differentiated from basophilic cells.[6] Comparative studies with normal skin showed that the shadow and transitional cells corresponded to hair cortex cells, the squamoid cells to the outer root sheath, the basophilic cells adjacent to the stroma to the outermost cell layer of the outer root sheath between the lower permanent portion and the upper transient portion of the follicles, and the basophilic cells adjacent to the transitional cells to the hair matrix. Pilomatricoma is considered to occur mostly in children and in young adults.[7-9] The female:male ratio is roughly between 3:2 and 1:1. Our study showed that 78% of the tumors arise before the age of 30 years with female:male ratio 5:4. Although pilomatricoma was believed to be rare in elderly people, there were cases described in adults and elderly. In a clinical review of 209 pilomatricomas by Julian and Bowers from United Kingdom, the tumors appeared at any age with peak presentation bimodally in the first and sixth decade.[10] This result supported the previous findings, which were carried out by Kaddu et al from Austria who described 118 cases of pilomatricoma, 58 of whom were above 45 years of age.[11] The association of pilomatricomas and myotonic dystrophy has been described. The association seems to involve the development of pilomatricomas before signs of myotonic dystrophy.[3]

From 1993-1999, 27 cases of pilomatricoma have been diagnosed in our department. A review of the literature showed that nearly 75% of the pilomatricomas were located in the head, neck and upper limb.[12] None of our cases has been diagnosed clinically as pilomatricoma. The most common clinical diagnosis was sebaceous cyst. Other diagnoses included lipoma, granuloma or unspecified lump. In older patients, the clinical diagnosis of basal cell carcinoma can be included as in Behnke et al series that described 4 cases of pilomatricoma in elderly individuals.[13] Pilomatricoma was also described in other sites like testis, ovary and breast.[14-16] Generally, the tumor varies in size from 0.5-3 cm, but it may be as large as 5 cm in diameter. The largest tumor in our series was 3.5 cm in diameter. Pilomatricoma occurs usually as a solitary lesion.

Table 2 - Distribution of tumors by age and sex.

Age	Male	Female	Total
0-10	3	4	7
11-20	1	7	8
21-30	4	2	6
31-40	33	2	5
41-50	1	0	1
Total	12	15	27

Most commonly, it manifests itself as a firm deep-seated nodule that is covered by normal skin. Superficial tumor usually has bluish red discoloration. Multiple occurrence of pilomatricoma is rare.[17] There were a few reports describing fine needle aspiration (FNA) diagnosis of pilomatricoma.[18-21] The cytologic presentation can be mistaken for squamous cell carcinoma because of high cellularity, high nuclear/cytoplasmic ratio and presence of anucleate squames. However, a combination of basaloid cells, ghost cells and foreign body giant cells appeared to be necessary in FNA smear for a confident cytodiagnosis of pilomatricoma. The presence of ghost cells seems to be the key to recognizing this type of tumor.[20] However, awareness of the entity leads to the correct diagnosis of pilomatricoma. Malignant pilomatricoma has been reported in different literature, but not in our study.[22-23] They are rare tumors and have been called invasive pilomatricoma, metrical carcinoma and trichomatrical carcinoma. Most cases were located in the head, neck and back. Most of the affected patients were middle-aged men. Aggressive surgical excision is recommended because local recurrence is frequent and pulmonary metastasis has been reported.[24,25] Based upon the histopathological findings, Kaddu et al proposed that pilomatricomas may be categorized into four distinct morphological stages and that these stages reflect the life of a pilomatricoma.[26] The four distinct and chronological stages are early, fully developed, early regressive and late regressive. The lesion begins as an infundibular matrix cyst and ends up as a calcified and ossified nodule with no visible epithelial component.

Pilomatricoma has a wide variety of clinical characteristics and are often misdiagnosed.[27] They appear at any age, with a peak presentation in the first and sixth decades. Accurate diagnosis and appropriate treatment can be obtained through careful clinical examination.

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