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Malherbe's calcifying epithelioma (pilomatrixoma) in childhood; Reviewed 105 cases observed over 10 years

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Abstract

Pilomatrixoma is an uncommon benign skin tumour that is usually seen in children and adolescents, and can be treated with surgical excision. The aim of this study was to evaluate the clinical characteristics of these lesions that can cause difficulties in diagnosis, and to discuss the differential diagnosis, histopathological characteristics and follow-up in the context of the literature.

A retrospective examination was made of 105 cases which were treated with excisional biopsy for a subcutaneous mass, which was then pathologically diagnosed as pilomatrixoma between 2008 and 2017. The cases were evaluated with respect to age, gender, lesion diameter, location, recurrence, and histopathological characteristics of the lesions.

A total of 113 lesions were detected in 105 patients comprising 75 females and 30 males. The mean age at diagnosis was 8.9 years. In 3 patients, 2 lesions were determined in separate locations on presentation. The lesions were localized on the head and neck in 48 cases, on the upper extremities in 38, on the trunk in 18, and on the lower extremities in 9. The diameter of the lesions ranged from 0.3 to 3 cm. Within the study period, recurrence developed in the same location after 1 year in 1 patient, and a separate second lesion developed after mean 3.7 years in 4 cases.

In the present study, a realistic comprehensive picture of pilomatrixoma in our central hospital has been provided. It's manifestation is diverse and easily misdiagnosed. Early complete excision is recommended for hard or firm nodules on head, neck and upper extremity.

Keywords: Children and adolescents, pilomatrixoma, a single center experience, ten years

Introduction

Pilomatrixoma, also known as calcifying epithelioma of Malherbe, is a benign tumor of the skin which develops

from matrix cells of the hair follicles located in dermal or subcutaneous tissue (1). Pilomatrixoma occurs within the first two decades of life, and classically presents as a solitary, small (but rarely up to 5 cm), firm subcutaneous nodule, with overlying skin (2). Complete surgical excision of the mass and overlying skin with a wide margin is the recommended treatment for pilomatrixoma.

Pilomatrixoma is slightly more common in females and usually occurs in the head and neck, followed by the extremities and the trunk (1, 3, 4). Histologically, pilomatrixoma shows a characteristic, well-circumscribed border, surrounded by a connective tissue capsule. It is usually located in the lower dermis and is typically composed of two distinct cell populations: (i) small basaloid cells at the periphery of the island, and (ii) the keratinized anucleated cells, so called "ghost or shadow cells", in the central portion. Early lesions show predominantly basaloid cells in islands, and as the lesions mature, the basophilic cells acquire more cytoplasm and lose their nuclei to become ghost cells which may subsequently be seen with calcifications. Squamoid epithelium, amorphous debris, and cyst formation are frequently found in the tumor and sometimes foreign body reaction can be observed. Calcification is common, and ossification may also be seen in the "ghost or shadow" cell area (1, 5-9).

There are only a few studies in literature which have described the characteristic histopathological findings of pilomatrixoma in children (1, 10, 11). The aim of this study was to define the clinical and epidemiological features of pilomatrixoma in children and adolescents of our hospital and to determine and discuss the histopathological characteristics of these lesions.

Material and methods

A retrospective analysis was made of 113 lesions in 105 patients, who were diagnosed with pilomatrixoma in our department between 2008 and 2017 in the Antalya Education and Research Hospital, Department of Pathology. All histological slides were reviewed by two pathologists and the diagnosis of each case was confirmed.

From the hospital archived records, a record was made for each patient including gender, preoperative diagnosis, age at excision, tumor location, and concomittant neoplasms. The patients were followed-up for recurrence or malignant transformation after the excision. The results were compared with the information available in current literature.

The research was reviewed and approved by an institutional review board, and that participation involved informed consent. The study was approved from Antalya Education and Training Hospital Ethical committee. (Number: 18/16, Date: 27/09/2018)

Results

A retrospective review was made of the records of a total of 105 childhood pilomatrixoma patients. The 105 patients comprised 75 (71.5%) females and 30 (28.5%) males with a median age of 8.9 years (range, 0-18 years).

Clinical presentation at diagnosis

A total of 113 lesions were detected in 105 patients, of which 48 (42.5%) were located in the head and neck, 38 (33.7%) in the upper extremities, 18 (15.9%) in the trunk or back, and 9 (7.9%) in the lower extremities. Lesions were described as hard and immobile subcutaneous, usually painless masses on physical examination. The size of the lesions ranged from 0.3-3 cm. No patients had a history or any additional physical examination findings.

In 3 patients, 2 lesions in different locations were determined on presentation. Thus, the incidence of multiple lesions was 2.8%. Patient characteristics are summarized in Table I.

Histopathological examination

Histopathologically, due to differentiation from hair matrix toward the cortex cells, a keratin core in the center of the lesion and peripheral basaloid cells are seen around this core. These are also known as "shadow or ghost" cells and are pathognomonic for pilomatrixoma, occurring as a result of this cell's keratinization (Figure I). The classic appearance of basaloid and "ghost or shadow" cells was seen in all cases. Calcification and ossification were frequently seen. Foreign body reaction was also common (Figure II) (6, 10, 12). The histopathological features of the patients with pilomatrixoma are summarized Table ll.

Treatment and follow-up

All lesions were excised completely. Recurrence rate was determined to be low (4.7%) in the current series. Within the study period, recurrence developed in a different location after mean 3.7 years in 4 cases. Local recurrence in the same area developed after 1 year in 1 patient. To date, no patient has developed malignant transformation.

Discussion

Pilomatrixoma is the second most common benign skin tumor in childhood. This lesion was first described as a calcifying epithelioma of the sebaceous glands by Malherbe and Chenantois in 1880. It was found to originate from the hair follicle in 1942 and the name "pilomatrixoma" that is used today was given by Forbis and Helwig in 1961 (1, 2, 5). Pilomatrixoma shows differentiation from hair matrix toward the cortex cells (3, 6). Some studies have shown that pilomatrixoma is thought to arise from a mutation of the β-catenin gene (1). Although the etiology is not clear, it is thought to be due to the pause in the life cycle of hair follicles caused by trauma and inflammation. Polyoma viruses have also been reported in the etiopathogenesis (2).

Pilomatrixoma is most frequently seen in the first two decades of life, with females more frequently affected than males. The incidence in all pathology specimens has been found to be between 1/500-2.200. Pilomatrixoma most commonly occurs in hair-bearing areas, primarily in the head and neck area, followed by the trunk and extremities (1, 3, 4, 7). In the current study, these tumors were more common in female patients. In addition, the tumor location showed similar results, with head and neck presentation being the most common.

Pilomatrixoma usually presents as a solitary lesion and multiple lesions have been reported to be seen at rates between 2%-3% (2, 5). In literature, rare cases of multiple pilomatrixoma have been reported in patients with systemic diseases, such as Gardner's syndrome, myotonic dystrophy, sarcoidosis and Turner syndrome (1, 13). In the current series, multiple lesions were determined in 3 patients with 2 in each. None of these patients had any concomitant neoplasms.

Pilomatrixoma do not regress or spontaneously recover so complete surgical excision of the lesion with a wide margin is the suggested treatment. When complete resection can be achieved, no recurrence can be seen. In literature, recurrence is rare with reported recurrence rates of 0-3% (1, 2, 7). In the current study, recurrence developed in 5 patients, 1 as local recurrence and 4 as recurrence in another site of the body. Within the study period, the recurrence rate was 4.7%.

The reported incidence of calcification in pilomatrixoma ranges from 69% to 85%, and osseous metaplasia has been described in 15% of cases (7). In the current series, calcification was seen in 82% and ossification in 25%. In addition, other secondary changes such as foreign body reaction are common and have been well documented in literature (5, 7, 12). In the current series, foreign body reaction was detected in 62% of the patients.

Pilomatrixoma presents clinically as a firm, bluish, mobile dermal or subcutaneous nodule, with a lobulated surface. Such a "non-specific" presentation leads to a wide spectrum of clinical differential diagnosis such as epidermal cysts, dermoid cysts, foreign body granulomas, xanthoma, fat necrosis, and even skin cancers, such as adnexal tumour (12, 13). Hence, recognition of the characteristic histopathological features is needed to distinguish pilomatrixoma from similar lesions (1, 8, 13). Like pilomatrixoma, epidermal cyst, trichoblastoma, trichoepithelioma, panfolliculoma, basal cell carcinoma, apocrine mixed tumor, and complex adnexal tumor can also be seen with matrix cell differentiation. Furthermore, the predominance of basaloid cells in islands in early lesions can mimic basal cell carcinoma, and the "ghost or shadow cells" in mature lesions can resemble squamous cell carcinoma (1, 4, 12, 14-16).

Although almost all pilomatrixoma are benign, malignant transformations have been reported as malignant pilomatrixoma and metastasis of this tumor to the lung, bone, brain, and skin. In addition to ulceration, nuclear pleomorphism, atipical mitosis, central necrosis, and presence of infiltration of the skin or adjacent tissues is suggestive of malignancy (1, 2). There was no malignant transformation in any of the current study patients after total excision.

Conclusion

Pilomatrixoma displays a broad spectrum of architectural and cytological features. Therefore, recognition of the characteristic histopathological features is needed for differentiation from other lesions. Clinically, pilomatrixoma may simulate a variety of malignancies, sometimes histologically, especially when "ghost or shadow cells" are dominant. It can be assumed to be squamous cell carcinoma, and when basaloid cells are dominant, it can mimic basal cell carcinoma. Awareness of the characteristic morphological features of pilomatrixoma is needed to diagnose this lesion correctly. Surgical excision is curative.

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Tables

Table I. Demographic Features of the Patients.

	Number	**%**
Age at diagnosis		
Median	8.9	
Range	0-18	
Gender		
Female	75	%71.5
Male	30	%28.5
Lesion locations		
Head and neck	48	%42.5
Upper extremities	38	%33.7
Trunk or back	18	%15.9
Lower extremities	9	%7.9
Lesion number		
Single	102	%97.1
Multiple	3	%2.9

Table II. The histopathological features of pilomatrixoma

Histopathology	N%
Peripheral basophilic (basaloid) cells	%100
“shadow or ghost” cells	%100
Calcification	%82
Foreign body reaction	%62
Ossification	%25

Figures

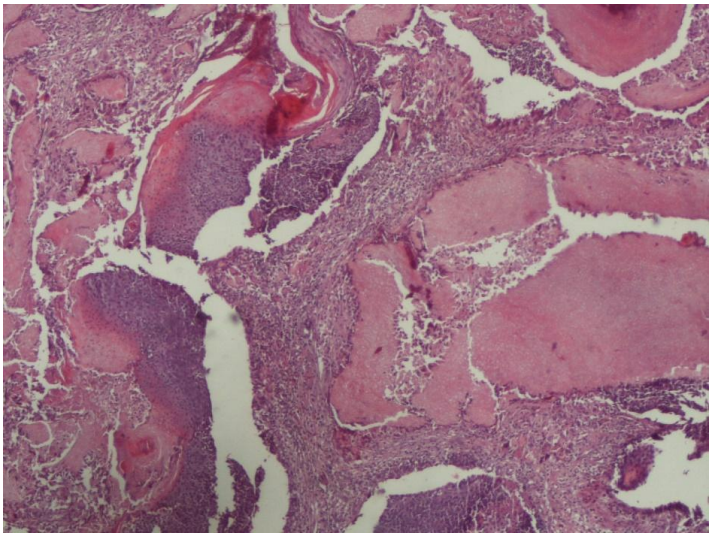

Figure 1: Tumor composed of basaloid cells and “shadow or ghost” cells (H&E X200)

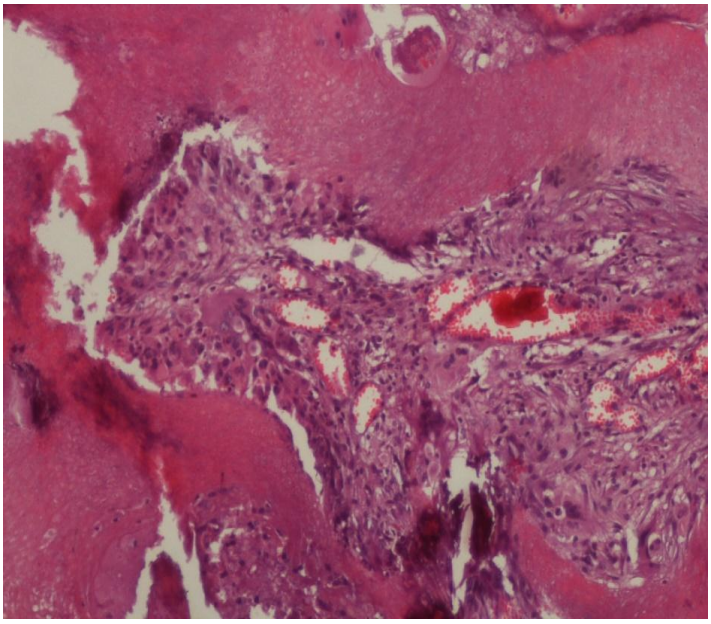

Figure 2: Calcification and foreign body reaction (H&E X400)