

Pilomatrixoma: A Clinico - Pathological Study

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ABSTRACT

Introduction: Pilomatrixoma (calcifying epithelioma of Malherbe) is a benign skin tumor with differentiation towards hair follicles. Pilomatrixoma may be mistaken for many other conditions like epidermoid cyst, dermoid cyst, calcified lymph node or a hemangioma. The aim of this study was to describe the clinical and histopathological characteristics of patients presenting with pilomatrixoma.

Material and Methods: The present study was conducted in the Post Graduate department of pathology, govt. medical college Srinagar. 51 cases of pilomatrixoma were included in the study. The final diagnosis was arrived by histopathological examination of the specimens. The clinical features and physical examination including age, sex, site of lesion and size were noted and analyzed.

Results: 51 cases of pilomatrixoma, comprising about 37% cases of all skin adnexal tumors were considered. The most common age group involved was 11-20 years comprising of 33.34% of all cases. 23 (45.10%) were males and 28 (54.90%) were females. There was a slight female predominance. The most common anatomical location for the tumor was found to be head and neck followed by trunk, upper limbs and lower limbs. The histopathological findings observed were: a benign well demarcated lesion surrounded by a capsule in almost all of the cases. The lesions were found to be composed of islands of epithelial cells embedded in a stroma. Two types of cells were identified in these epithelial islands basaloid cells and shadow cells.

Conclusions: Pilomatrixoma should be considered in the differential diagnosis of nodules, especially those on the head and neck. Careful clinical examination and familiarity with the condition may lead to accurate diagnosis and appropriate treatment.

Keywords: Pilomatrixoma, Skin Tumor, Hair Follicle, Adnexal.

INTRODUCTION

Pilomatrixoma is also known as calcifying epithelioma of Malherbe. The lesion was first described by Malherbe and Chenantais in 1880 as a calcified tumor, originating from the sebaceous glands.¹ Later, Forbis and Helwig discovered that the cell of origin for pilomatrixoma is the outer sheath cell of the hair follicle root.² These lesions mostly present as solitary, superficial painless firm swellings. The overlying skin usually depicts bluish discoloration. The lesions grow slowly and are usually asymptomatic. The most common site of involvement is the head and neck with a predilection for face. However the presentation may vary from case to case. These are most commonly seen in children and young adults.³ The pilomatrixoma may be mistaken with many other skin conditions. Notable among these are epidermoid cyst, dermoid cyst, calcified lymph node or a Hemangioma.⁴

Pilomatrixoma is usually diagnosed clinically. Ultrasound may be helpful. The features vary from a partially to totally calcified nodule or a hypo-echoic nodule with internal calcification.⁵

The purpose of this retrospective study was to describe the clinical presentation and histopathological aspects of pilomatrixoma received in the department of pathology over a period of three years in a tertiary care hospital.

MATERIAL AND METHODS

The present study was conducted in the Postgraduate department of Pathology, Government Medical College, Srinagar and its Associated Hospitals over a period of three years from January 2015 to December 2017. The study was partly retrospective from Jan 2015 to December 2015 and partly prospective from January 2016 to December 2017. The material for this study comprised of surgically excised specimens received from the departments of Dermatology and Surgery. In prospective duration of study the clinical history, physical examination and investigations done were noted. The clinical profile included age, sex, site of location and size of lesion. The specimen received was subjected to meticulous gross and microscopic examination. The specimen were fixed in neutral buffered formalin and paraffin embedded. Thin sections were cut, stained by Hematoxylin and Eosin and examined under microscope. For retrospective study the histopathology slides as well as the clinical profile were retrieved from the archive and reviewed.

RESULTS

A total of 138 skin adnexal tumors were diagnosed on histopathological examination during the study period. There were 51 cases of pilomatrixoma, comprising about 37% cases of all skin adnexal tumors. The age of the patients varied from 4-60 years. The highest occurrence was observed in the age group of 11-20 years comprising of 33.34% of all cases followed by age groups 21-30 years comprising of 23.53% and 1-10 years comprising of 13.73% of cases. The distribution according to age is shown in (Table 1). Mean age of presentation of pilomatrixoma was 28.5 years. Among the 51 cases, 23 (45.10%) were males and 28 (54.90%) were females yielding a M:F ratio of 1:1.22.

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S No.	Age	No. of cases	Percentage
1	<10	7	13.72
2	11-20	17	33.34
3	21-30	12	23.53
4	31-40	7	13.72
5	41-50	4	7.84
6	51-60	4	7.84
7	>60	0	0

Table-1: Depicting the distribution of age

Site	No. of cases	Percentage
Head	14	27.45
Neck	7	13.72
Trunk	12	23.52
Upper limb	12	23.52
Lower limb	6	11.76

Table-2: Depicting the Anatomical location of the lesion

There was a slight female predominance. The most common anatomical location for the tumor was found to be head and neck followed by trunk, upper limbs and lower limbs. 21(41.18%) cases were located in head and neck with 8 cases on the face, 7 on the neck and 6 on the scalp (Table 2). The trunk and upper limb was found to be the next common site with 12 (23.53%) cases each followed by lower limb with 6 (11.76%) cases. Most cases in our study presented as the superficial solitary firm nodule. 8 out of 51 were cystic and 5 were multiple. Most of the patients presented with a painless swelling (45 Out of 51) however, 6 cases presented with pain. Ultrasonography was available in 10 cases. The findings ranged from a calcified nodule to a hypoechoic nodule with foci of calcification.

In this study the histopathological findings observed were: a well demarcated lesion surrounded by a capsule in almost all of the cases. The lesions were found to be composed of islands of epithelial cells embedded in a stroma. Two types of cells were seen in the epithelial islands the basaloid cells and the shadow cells. The proportions of these cellular components varied from case to case. The keratinized eosinophilic shadow cells were mostly seen in the form of whorls near the center of the tumor. The basophilic cells were found near the periphery. Granulomatous inflammation with giant cell reaction in areas of keratinization was seen in 4 cases. Foci of dystrophic calcification were identified in 3 cases. Morphological features of aggressiveness or malignancy, such as cytological atypia, infiltrative basaloid nodules, abundant mitosis, and areas of necrosis, were not identified.

DISCUSSION

Pilomatrixoma is a benign appendageal tumor of skin that has differentiation towards hair follicles. This study is an attempt to describe the clinicopathological presentation of pilomatrixoma. Out of a total of 138 adnexal tumors received during the period of 3 years, 51 cases of pilomatrixoma were identified on histopathological examination. The age of patients varied from 4-60 years with 11- 20 as the most common age of presentation. Julian et al described similar results with 5-15 years as the most common presenting age. 6 Schwatz et al described pilomatrixoma as the most common

solid cutaneous tumors in patients younger than 20 years of age.⁷ Among the 51 cases in our study, 23 (45.10%) were males and 28 (54.90%) were females yielding a M:F ratio of 1:1.22 thus showing a slight female preponderance. Most studies describe a slight female preponderance.⁸ The most common site of location in this study was head and neck with highest number of patients presenting with involvement of the face followed by trunk, upper limbs and lower limbs. Studies describe head and neck as the most frequently affected site.⁹

The preoperative diagnosis in majority of the cases was clinical. Most cases in our study presented as the solitary deep seated nodular lesion. 8 percent (4 out of 51) of cases were multiple. Some of the several studies have reported incidences of multiple pilomatrixomas ranging from 2 to 26.2 percent.¹⁰ Most of the cases were solid while a few presented as cystic lesions. Only 6 (11.76percent) cases presented with pain while most of the cases were painless. Duflo et al. reported a 20 percent incidence of pain and inflammation associated with pilomatrixoma.¹¹ Ultrasonography was available in 10 cases. The findings ranged from a calcified nodule to a hypoechoic nodule with foci of calcification consistent with earlier studies.

On gross examination size of the lesion ranged from 0.2 to 2.0 cm with a mean of 0.8 cm. Most studies describe lesions 0.5-3.0 cm in size. 43 out of 51 cases in our study presented with a solid pattern and 8 with cystic pattern which is consistent with many other studies.^{12,13} The histopathological findings in this study were a well demarcated lesion surrounded by a capsule. The lesions were found to be composed of Islands of irregularly shaped epithelial cells embedded in a stroma. Two types of cells were seen in the epithelial islands the basaloid cells and the Shadow cells. The proportions of these cellular components varied from case to case. The keratinized eosinophilic shadow cells were mostly seen in the form of whorls near the center of the tumor. The basophilic cells were found near the periphery. Granulomatous inflammation with giant cell reaction in areas of keratinization was seen in 4 cases. Foci of dystrophic calcification were identified in 3 cases. Morphological features of aggressiveness or malignancy, such as cytological atypia, infiltrative border, abundant mitosis, and areas of necrosis, were not identified. These findings are consistent with the observations reported in other series.^{14,15}

CONCLUSION

Pilomatrixoma, a benign skin adnexal tumor, with differentiation towards hair follicles, should be considered in the clinical differential diagnosis of solitary firm skin nodules. The most common age group involved is the adolescents. The head and neck is the most common region involved. Microscopic examination shows a well demarcated lesion surrounded by a capsule. The lesions composed of Islands of irregularly shaped epithelial cells embedded in a stroma depict two types of cells in the epithelial islands, the basaloid cells and the Shadow cells. Morphological features such as cytological atypia, infiltrative border, abundant mitosis, and areas of necrosis are not seen and should arise suspicion of malignancy and aggressiveness.

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