International Journal of Medical Research Professionals

Original Article

Clinicomorphological Review of Pilomatrixoma: A Study of 7 Cases.

Rajat Gupta[1*], Deepika Dewan[2], Paramjeet Singh[3]
[1]Consultant Pathologist, [3]Senior Consultant Surgeon, Government Hospital, Gandhi Nagar, Jammu, J&K, INDIA.
[2]Senior Resident, Department of Community Medicine, GMC, Jammu, J&K, INDIA.

Article History
Received: 25 Mar 2016
Revised: 26 Mar 2016
Accepted: 28 Mar 2016

***Correspondence to:**
Dr. Rajat Gupta
H.No 18, Lower Laxmi Nagar, Sarwal, Jammu, J&K, INDIA.
deepika.nity@gmail.com

ABSTRACT

Background: Pilomatrixoma (Calcifying Epithelioma of Malherbe) is a benign appendageal tumor with differentiation towards hair cells most commonly seen in the first and second decades of life and more common in females. The lesion is typically found in head and neck region. Only few studies with preoperative aspiration cytology have been documented in literature.

Aim: To study the clinico-morphological spectrum of patients diagnosed with pilomatrixoma on Fine Needle Aspiration Cytology (FNAC).

Material and Methods: Fine Needle Aspiration cytology was done on patients presenting with palpable dermal and subcutaneous nodules at various sites in Pathology department of Government Provincial Hospital Gandhi Nagar subsequent to referral from clinicians for further evaluation of these lesions. Detailed clinical history was taken and the character of aspirate was noted. In each case, three alcohol fixed smears were prepared, first smear was stained with Papanicolaou stain, second with Giemsa stain and third one was kept unstained for any further required stain. Subsequent excision biopsy was also evaluated.

Results: Over a period of two years, 07 cases of Pilomatrixoma were diagnosed on FNAC. Out of them 4 were females and 3 were males. Most of the patients were less than 25 years of age. The sites involved were neck, head and upper extremity. Basaloid cells accompanied with shadow cells and multinucleated giant cells were the characteristic findings on FNAC.

Conclusion: FNAC is an important, cheaper and rapid diagnostic tool for preoperative diagnosis of pilomatrixoma.

KEYWORDS: Appendageal tumor, Calcifying Epithelioma of Malherbe, Fine Needle Aspiration Cytology (FNAC), Pilomatrixoma.

INTRODUCTION

Pilomatrixoma, also known as Calcifying Epithelioma of Malherbe, first described by Malherbe and Chenantais in 1880[1] is a benign appendageal neoplasm with differentiation towards hair cells, particularly hair matrix cells.[2] It is most commonly seen in the first and second decades of life and more common in females as compared to males. The lesion is typically found in head and neck region, but can also occur in upper extremities and rarely at other sites[3].

Usually because of lack of knowledge, preoperative diagnosis of pilomatrixoma is rarely considered by clinicians. The discussion of the lesion is usually limited to dermatology, pathology and to some extent otolaryngology. Histological diagnosis of pilomatrixoma is straight forward, however the same is not true for cytology. FNAC can be used as an important tool in preoperative diagnosis of the lesion, though sometimes misdiagnosed especially in rare sites and if cytopathologist is not well versed with the variable cellular morphology of the lesion.[4] The present study highlights the importance of FNAC in considering pilomatrixoma as one of the differential diagnosis of nodular subcutaneous lesions especially in rare sites. An accurate diagnosis of this benign lesion on FNAC is important considering that excision is curative.

OBJECTIVE OF STUDY

To study the clinico-morphological spectrum of patients diagnosed with pilomatrixoma on Fine Needle Aspiration Cytology (FNAC).

MATERIALS AND METHODS

The present study was observational descriptive cross sectional hospital based study done on all the patients coming in Outpatient department of Government hospital Gandhi Nagar, Jammu with nodular lesions of head, neck and extremities referred to the cytology section over a period of two years i.e. January 2014 to December 2015. FNAC was done in all such cases for further evaluation. Detailed history and relevant clinical examination was done in all cases. FNAC was performed using 22 gauge needle and 10 ml plastic disposable syringe with a detachable syringe holder (Franzen Handle). The character of aspirate was noted. In each case, three alcohol fixed smears were prepared, first smear was stained with Papanicolaou stain, second with Giemsa stain and third one was kept unstained for any further required stain. Subsequent excision biopsy was also evaluated in all cases diagnosed with pilomatrixoma on FNAC.

RESULTS

A total of seven cases were diagnosed as Pilomatrixoma. The age of these patients ranged from 07 to 53 years. The mean age at diagnosis was 23.4 years. Out of the 07 cases 04 (57.1%) were females and 03 (42.9%) were males with Male:Female ratio of 1:1.3. The most common site of involvement was neck (03) followed by head (02) and upper extremity (02) (Table 1).

Maximum number of cases presented as non-tender nodules. The duration of lesion ranged from 01 month to 13 months. In none of the 7 cases, pilomatrixoma was the clinical diagnosis. 02 cases presented with provisional diagnosis of lipoma and reactive lymphadenitis each, while neurofibroma, inclusion cyst and ganglion were the provisional diagnosis in one case each. The aspirate varied from necrotic, particulate to blood mixed (Table 2). Subsequent histopathological examination confirmed these cases as pilomatrixoma.

Cytomorphologically, all the cases showed cellular smears comprising of aggregates of anucleated squamous cells (Ghost cells) along with clusters of basaloid cells having round basophilic nuclei with vesicular chromatin, prominent nucleoli at places and scant cytoplasm (Fig 1,2). The background population comprised of numerous multinucleated giant cells along with few lymphocytes admixed with areas of necrosis in some cases (Fig 3). In some cases characteristic pink fibrillary material was seen enveloping the basaloid cells (Fig 4,5). Calcification was not appreciated in cytological smears. All the cases were subjected to excisional biopsy and histopathological examination confirmed the diagnosis of pilomatrixoma in all cases.

Table 1: Age, Gender and Site Presentation of 7 Cases of Pilomatrixoma

Case no.	Age (in Completed years)	Sex	Site
01	22	M	RIGHT SIDE NECK
02	53	F	LEFT SIDE NECK
03	16	F	RIGHT ELBOW
04	10	M	POSTERIOR TRIANGLE NECK
05	25	F	SCALP (LEFT TEMPORAL AREA)
06	07	M	FOREHEAD
07	31	F	LEFT FOREARM

Table 2: Clinicomorphological Presentation of 7 Cases of Pilomatrixoma

Case no.	Duration of lesion (in months)	Clinical presentation	Provisional diagnosis	Nature of aspirate	FNAC Diagnosis
01	02	2.5x 2.0 cms, NON TENDER	REACTIVE LYMPHADENITIS	NECROTIC	PILOMATRIXOMA
02	11	2.0x1.0 cms TENDER	NEUROFIBROMA	BLOOD MIXED, PARTICULATE	PILOMATRIXOMA
03	08	3.0x1.0 cms NON TENDER	GANGLION	PARTICULATE	PILOMATRIXOMA
04	13	2.5x1.5 cms NON TENDER	REACTIVE LYMPHADENITIS	BLOOD MIXED	PILOMATRIXOMA
05	05	1.5x1.5 cms TENDER	EPIDERMAL INCLUSION CYST	NECROTIC	PILOMATRIXOMA
06	01	1.0x1.0 cms NON TENDER	LIPOMA	NECROTIC	PILOMATRIXOMA
07	10	2.0x2.0 cms NON TENDER	LIPOMA	PARTICULATE	PILOMATRIXOMA

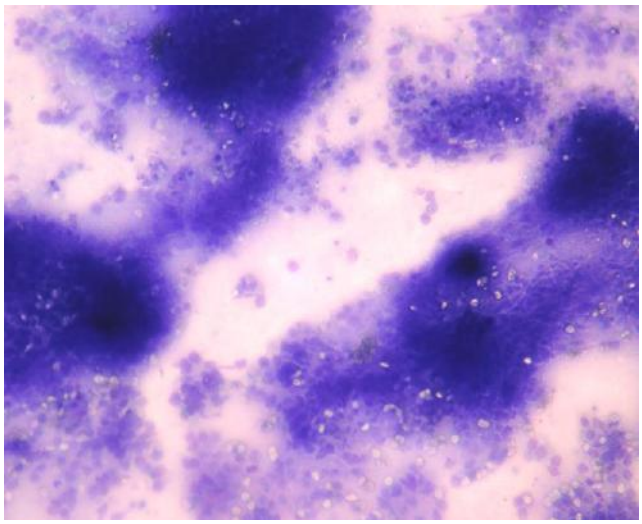

Fig 1: Clusters of basaloid cells with few ghost cells (40x, Giemsa)

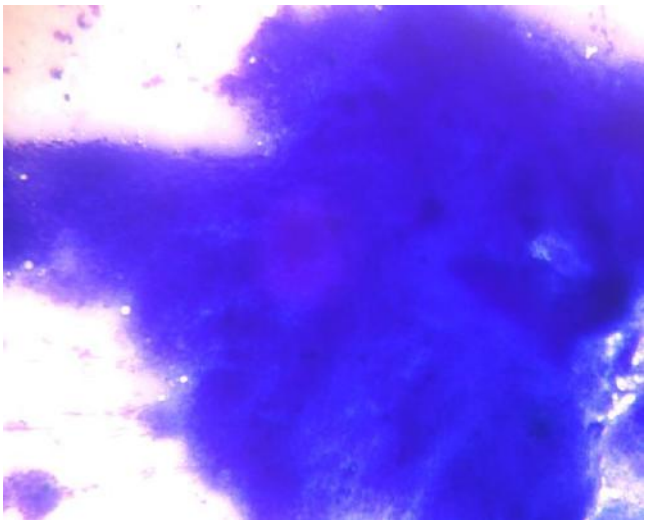

Fig 2: Cluster of ghost cells with basaloid cells on margins (40x,Giemsa)

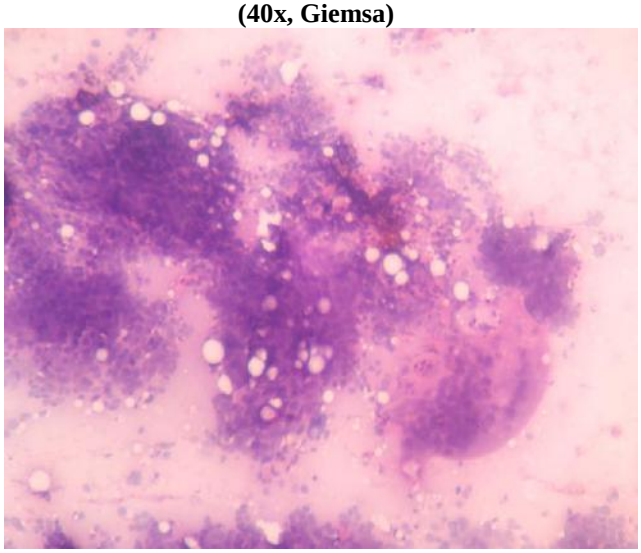

Fig 3: A multinucleated giant cell admixed with basaloid cell cluster (80x,PAP)

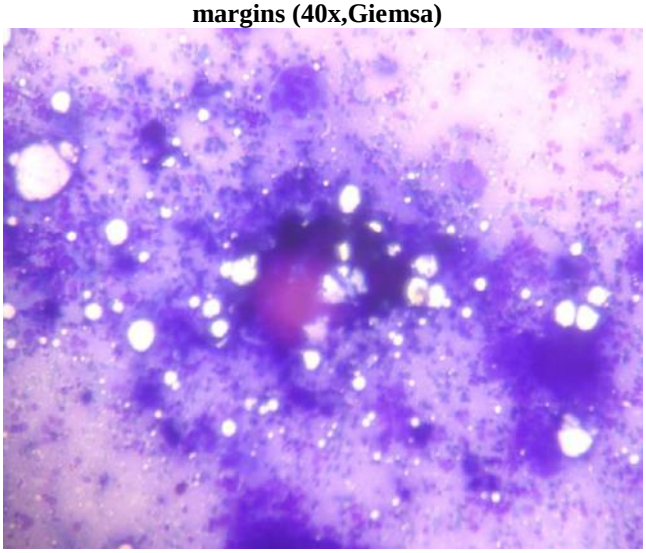

Fig 4: Pink fibrillary material with multinucleated giant cells and basaloid cells (20x,Giemsa)

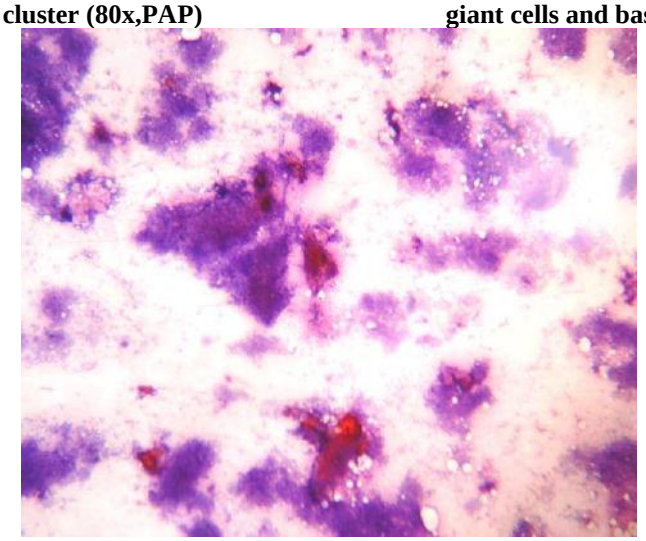

Fig 5: Basaloid cell clusters admixed with pink fibrillary material in a chronic inflammatory background (20x,PAP)

DISCUSSION

In 1922, Dureuilh and Cazenave described the histopathologic characteristics of this neoplasm, including islands of epithelial cells and shadow cells.[5] In 1961, Forbis and Helwig reviewed 228 such tumors with histochemistry and electron microscopy and established the outer sheath cell of the hair follicle as the cell of origin. They coined the term pilomatrixoma to avoid overlapping nomenclature with malignancy as suggested by the previously used term "Epithelioma" [6]

Pilomatrixoma is a benign cutaneous adnexal tumor with a predilection for head and neck region followed by upper extremities, trunk and lower extremities in decreasing order of frequency. The neck is the most commonly affected followed by the cheek, scalp, preauricular and periorbital areas in the head and neck region.[7] In our study also, neck was the most common site followed by head and upper extremity. Similar findings were seen in study conducted by Agarwal R et al.[8] Single isolated lesion in head and neck region was also reported by Dwivedi G et al[9] and Chuang CC et al[10]. One case each from pinna, right hand and neck were reported by Nigam JS et al[11].

Pilomatrixoma can occur in any age, demonstrating bimodal peak presentation during the first and sixth decades of life. The lesion primarily affects children and adolescents, with 40% of cases occurring before age 10 and 60% before age of 20[12]. In our study, most of the cases were less than 25 years of age with one case in sixth decade of life.

Similar findings were seen in study by Dwivedi G et al[9], Chuang CC et al[10] and Nigam JS et al[11]. Birman MV et al in their report presented a case of pilomatrixoma in 52 years old female.[13] In a study by Agarwal R et al[8], all five cases were below 40 years of age with 2 below 20 years. In our study, females were more commonly involved as compared to males. Similar findings were observed by Agarwal R et al[8] Dwivedi G et al [9] and Chuang CC et al[10]. However Nigam JS et al[11] reported more cases in males.

The pilomatrixomas usually appear as a firm, solitary, painless, slow growing nodules with diameter ranging from 0.5 to 3.0 cm in most cases, often with discolouration of overlying skin.[14] This has been attributed to attachment of the tumor to the overlying epidermis and growth of blood vessels into the overlying skin. Although pilomatrixomas are usually solitary, multiple lesions have been reported in association with genetic disorders like Myotonic dystrophy, Gardner's syndrome, Xeroderma pigmentosum and Basal cell nevus syndrome.[15] In our study all the cases were slow growing, solitary in presentation, with tenderness in only 2 cases. Similar findings were seen in study by Agarwal R et al[8], Dwivedi G et al[9], Chuang CC et al[10] and Birman MV et al.[13]

Diagnosis of pilomatrixoma can often be made solely on the basis of clinical features. Differential diagnosis of head and neck pilomatrixoma includes dermoid cyst, branchial cleft remnant, preauricular sinus, adenopathy, sebaceous cyst, giant cell tumor, chondroma, foreign body reaction or malignant soft tissue tumor.[16] In our study, lesions were clinically diagnosed as lipoma, reactive lymphadenitis, ganglion, inclusion cyst and neurofibroma. In the case report by Dwivedi G et al[9], pilomatrixoma was clinically mistaken for round cell tumor.

The accuracy rate of preoperative diagnosis of pilomatrixoma reported ranges from 0-30%, probably due to the lack of familiarity with this tumor. Diagnostic tests and radiological imaging studies are often unnecessary in the workup of a superficial, benign skin lesion like pilomatrixoma, however sometimes may be done to determine the depth of lesion[17]. FNAC, the most favoured diagnostic tool in superficial masses, provides a fairly accurate diagnosis of pilomatrixoma, especially if the concerned cytopathologist is well versed with the morphologically characteristic findings. These include basaloid cell clusters, shadow (ghost) cells, calcification and anucleated squamous cells. Multinucleated giant cells may be seen in response to keratin. Wang et al. noted that 45 percent of cases of pilomatrixoma were incorrectly diagnosed by fine needle aspiration cytology based on their review of multiple case reports and series. Nevertheless, in their study as well as other more recent studies, fine needle aspiration has been found to be quite accurate when two key components, basaloid cells and ghost cells, are visualized, as this has been found to be specific for pilomatrixoma[18]. Hostologically, pilomatrixoma is sharply demarcated and contains basaloid cells and eosinophilic keratinized (shadow) cells. The proportions of these cellular components vary but the basaloid cells generally constitute the smaller component; in some cases, no basophilic cells are noted. These basaloid cells are fairly uniform in size, with rounded nuclei, small nucleoli, fine granular/vesicular chromatin, and delicate nuclear membranes. The keratinized eosinophilic shadow cells are located toward the center of the tumor and form masses, whorls, or stacks. The shadow cells have distinctive cell borders and contain central unstained areas corresponding to the lost nuclei that are characteristic of pilomatrixomas.

As spontaneous regression of pilomatrixoma never occurs and malignant transformation is rare, the standard treatment of the lesion is complete surgical excision[4].

CONCLUSION

The present study highlights the importance of FNAC in considering pilomatrixoma in the preoperative diagnosis of dermal and subcutaneous nodules, especially in sites other than head and neck. Dermatologists and otolaryngologists should be aware of this entity and familiar with the clinical presentation of pilomatrixoma. FNAC can be used as a cheaper and effective preoperative diagnostic tool in pilomatrixoma, so that surgical excision can be done at the earliest.

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Source of Support: Nil.

Conflict of Interest: None Declared.

Cite this article as: Rajat Gupta, Deepika Dewan, Paramjeet Singh. Clinicomorphological Review of Pilomatrixoma: A Study of 7 Cases. Int J Med Res Prof. 2016, 2(2); 234-38.