Case Report

DOI: 10.7860/JCDR/2022/59275.17326

Pathology Section

Pilomatricoma of Pinna- A Maze that may Misdirect

D SENTHILKUMARAN[1], SANKAPPA P SINHASAN[2], SIMON DASIAH[3]

ABSTRACT

Pilomatricoma is a rare cutaneous epidermal appendageal tumour having differentiation in the direction of hair cortex cells. It usually presents as single, less than 3 cm size, subcutaneous lesion, commonly seen in children, more commonly in head, neck and upper extremities. It can mimic a variety of other lesions clinically, as well as, cytologically, thereby, posing a diagnostic challenge. The ear lobe is a very rare site for it to occur. The authors report a case of a 61-year-old female presented with history of non tender nodule on left ear lobe for six months. The lesion was clinically suspected as keloid, sebaceous cyst and chondroma, later diagnosed as pilomatricoma on fine needle aspiration cytology, confirmed further on histopathological examination. Sebaceous cyst has been ruled out, due to the presence of characteristic findings in both cytology and histopathology. However, due to the inherent disadvantage of cytology, predominance of one component over the other would have mislead the diagnosis. It is being reported for its rare location, unusual age of presentation and to report its mimics, on cytology.

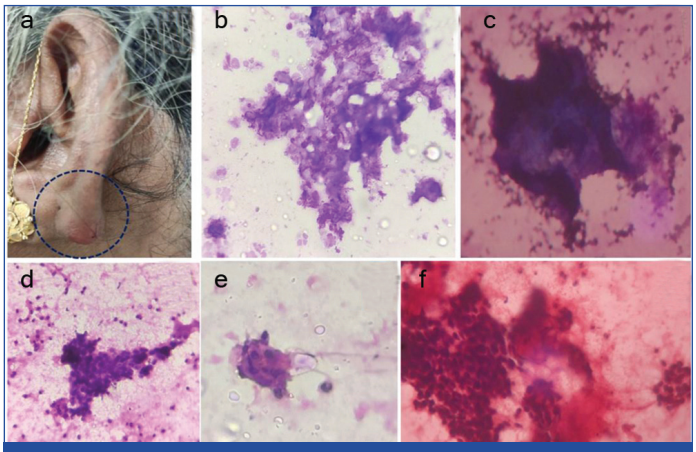
Keywords: Calcifying epithelioma of malherbe, Cytology, Earlobe, Histopathology, Pilomatrixoma

CASE REPORT

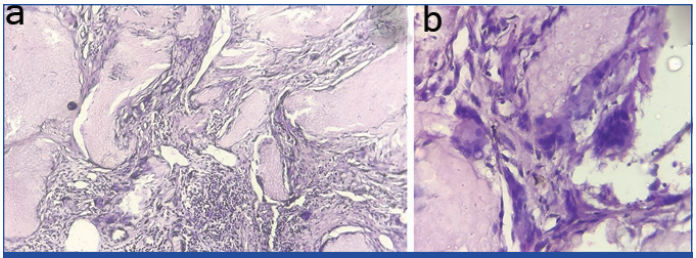
A 61-year-old female presented with an history of non tender skin-coloured nodule on the left ear lobe for six months. It was insidious in onset, gradually increasing in size and its overlying skin was smooth and unremarkable. There was no history of trauma and the patient's past medical history was unremarkable. There was no history of any topical treatments taken for the presenting complaint. On inspection, a partially well-defined nodule of size 0.8×0.5 cm, was seen over the left ear lobe [Table/Fig-1a]. The overlying skin was unremarkable with no atrophic changes or ulceration. No other swellings seen in the patient. On palpation, the nodule was firm in consistency, non tender, subcutaneous in plane, and partially mobile. Otorhinolaryngologist and surgeon's opinion were obtained and keloid, sebaceous cyst and chondroma were considered for differential diagnoses. Fine Needle Aspiration Cytology (FNAC) was suggested.

On FNAC, very scanty blood mixed material was obtained. The Leishman stain was performed on air-dried smears, Haematoxylin and Eosin (H&E) stain and Papanicolaou stain (Pap stain) were performed on alcohol fixed smears. On microscopy, the smears were moderately cellular and showed sheets and sheets of anucleate squames with multinucleate foreign body type of giant cell reaction to keratin admixed with clusters of monotonous population of basaloid cells displaying round regular nucleus. The background showed acute and chronic inflammatory response and foci of amorphous dystrophic calcifications. The Pap stained smears revealed admixture of basaloid monotonous population admixed with anucleate squames [Table/Fig-1b-f]. The differential diagnoses that were considered in FNAC smears were pilomatricoma, epidermal inclusion cyst and foreign body granuloma. If one of the components was predominant over the other, say only squames would favour epidermal inclusion cyst, predominantly giant cells would favour granulomatous disease or giant cell tumour. Since, the characteristic features of pilomatricoma were present, diagnosis of pilomatricoma was offered. The excision biopsy was advised for confirmation.

The biopsy was sent for histopathological examination after excision of lesion under local anaesthesia. On gross examination, there was four soft tissue fragments of which one was skin covered measuring 0.7 cm in greatest dimension. A few tiny chalky gray-white deposits were seen scattered on the cut section of the lesion. Multiple representative bits were taken for microscopic examination and entire gross specimen was submitted for tissue processing. Histopathological examination revealed a benign skin adnexal tumour partly lined by skin composed of sheets of squamous "shadow cells" or "ghost cells" admixed with monotonous population of basaloid cells [Table/Fig-2a]. The stroma showed numerous multinucleated foreign body type of giant cells, as a response to the keratin [Table/Fig-2b] and dense chronic inflammatory cell infiltrate. Focal areas of dystrophic calcification were also seen. There was no evidence of nuclear atypia, increased mitotic activity or ulceration. All these features were correlated with the cytological findings and a diagnosis of pilomatricoma was confirmed. The patient had uneventful postoperative period. Patient did not report for routine follow-up.



[Table/Fig-1]: (a) Clinical image showing swelling in the left ear lobe (circle). (b) Photomicrograph showing sheets of anucleate squames (Leishman stain, 10X). (c) Photomicrograph showing dystrophic calcification foci (Leishman stain, 10X). (d) Presence of sheets of monotonous population of basaloid cells with round regular nucleus (Leishman stain, 10X). (e) Presence of multinucleated foreign body giant cell response (Leishman stain, 40X). (f) Photomicrograph showing monotonous basaloid population of cells admixed with anucleate squames (Pap stain, 10X).



[Table/Fig-2]: (a) Photomicrograph showing sheets of squamous shadow/ghost cells admixed with basaloid population of cells (H&E, 4X). (b) Photomicrograph showing multinucleate giant cell response and chronic inflammatory cell infiltrate (H&E, 40X).

DISCUSSION

Pilomatricoma, a relatively rare benign skin appendageal tumour, was first described by Malherbe in 1880 and hence, it is also known as calcifying epithelioma of Malherbe [1]. However, after electron microscopic studies, the cell of origin was found to be hair cortex cell, hence, the name pilomatrixoma or "Pilomatricoma" was proposed [1]. Immunohistochemistry and various investigations showed that pilomatricoma can have differentiation of hair matrix, hair cortex, follicular infundibulum and hair bulge. Molecular studies reveal that Beta catenin pathway may play major role in this tumour, especially in syndromes like Familial Adenomatous Polyposis (FAP) [2]. Hence, these tumours have diversified differentiation based on their heterogeneity of keratin and filaggrin expression [3].

Pilomatricoma typically presents as a slow growing solitary lesion in head, neck and upper extremities [4,5]. However, pilomatricoma of ear lobe is very rare, only a few cases of such have been reported. The present case is being reported for its rare location, unusual age of presentation and to know its mimics on cytology. The size of this tumour usually ranges from 0.5 to 3 cm. The mean age of presentation is 32 years, however proliferating pilomatricoma and pilomatrix carcinoma are more commonly reported in elderly people [6]. The present case presented with an insidious onset of swelling of size 0.8×0.5 cm, correlating with other studies; however, the location and age of presentation are being rare.

Pilomatricoma is more frequently a deep-seated lesion covered by skin of unremarkable appearance. But, when it clinically presents as a superficial nodule, it may have skin-coloured changes thereby mimicking haemangioma [5]. Multiple lesions can be seen in familial cases and rarely over association with other conditions like sarcoidosis, Churg-Strauss syndrome, Turner's syndrome, myotonic dystrophy [7,8].

In the present case, due to its location, clinical diagnosis of keloid, chondroma, and sebaceous cyst were considered by clinician. Keloid being more common in Indian population, usually followed by trauma due to ear piercings. Auricular chondroma are a rare benign tumour that manifests as solitary slow growing subcutaneous nodule. Sebaceous cysts are the most common type of cysts in the ear, which can occur over earlobe, in the ear canal and behind the pinna. Hence, these three differential diagnoses were offered by clinicians and clinically they were never suspected as pilomatricoma.

Diagnosis of pilomatricoma on FNAC, is tricky, since both clinical and cytological features can mimic a variety of other common lesions. Mostly, it is erroneously diagnosed cytologically as epidermal inclusion cyst, giant cell tumour, sometimes as malignant adnexal tumour [8,9]. The relative scarcity of FNAC material is one of the reasons for their misdiagnosis. Predominance of one component over the other can result in deviation of diagnosis from pilomatricoma. The characteristic cytology triad include presence of primitive appearing basaloid cells, shadow or ghost cells and stroma showing foreign body type of giant cell response with inflammatory background [10]. In addition to these, foci of calcifications can also be seen. Depending on the stage of lesion and the site inside the lesion where FNAC is taken, the predominant population of cells vary from case to case. Pilomatricoma in its early stage may show cystic change rarely [11]. If multinucleate giant cells are predominant, it may be misdiagnosed as a granulomatous inflammation or giant cell tumour. However, the presence of shadow or ghost squamous cells adjacent to these giant cells should be considered suspicious for pilomatricoma. In the present case, clinical diagnosis of chondroma, keloid and sebaceous cyst were ruled out. Chondrocytes were not seen in smears, which ruled out chondroma. Sparsely cellular spindle cells of keloid were absent. Presence of basaloid cell population near the anucleate squames clusters favours pilomatricoma more than epidermal inclusion cyst. Hence, combination of cytology and biopsy are diagnostic to rule out the mimics. In elderly age group, one should search for any presence of atypia/invasion to rule out malignant counterpart of pilomatricoma, since the management and prognosis differs [11]. The main differentiating characteristics include a high mitotic rate with atypical mitosis, central necrosis, infiltration of the surrounding soft tissue, and lymphovascular invasion [12].

Literal meaning of maze is a puzzle through which one has to find a way, similarly correct diagnosis of pilomatricoma remains challenging and interesting on cytology. But at the same time, it is very simple to diagnose pilomatricoma on histopathology biopsy.

CONCLUSION(S)

To conclude, the diagnosis of pilomatricoma on cytology is tricky and presence of diagnostic triad-anucleate squamous ghost cells, monotonous population of basaloid cells and foreign body type of multinucleate giant cells, are not always seen in aspirated samples. Therefore, one should always keep pilomatricoma in differential diagnosis in mind, even, when one of these three diagnostic components may be missing in the smears. It is always advisable to carry out different stains in cytology as shadow cells and foreign body giant cells are better appreciated in Pap stained smears, whereas basaloid population of cells are better appreciated with Giemsa, Leishman and H&E stained smears.

REFERENCES

[1] Darwish, AbdullaAl-Jalahema, Dhiman E, Al-Khalifa AK. Clinicopathological study of pilomatricoma. Saudi Med J. 2001;22(3):268-71.
[2] Plotzke JM, Adams DJ, Harms PW. Molecular pathology of skin adnexal tumours. Histopathology. 2022;80(1):166-183.
[3] Abdeldayem M, Mekhail P, Farag M, Shehata G, Al Sheikh M, Izzidien A, et al. Patient profile and outcome of pilomatrixoma in district general hospital in United Kingdom. J Cutan Aesthet Surg. 2013;6(2):107-10.
[4] Kurokawa I, Yamanaka K, Senba Y, Sugisaki H, Tsubura A, Kimura T, et al. Pilomatricoma can differentiate not only towards hair matrix and hair cortex, but also follicular infundibulum, outer root sheath and hair bulge. Exp Dermatol. 2009;18(8):734-37.
[5] Sinhasan SP, Chaitra RJ, Ramachandra VB, Anandhi. Pilomatrixoma-Presenting as tender hypopigmented lesion. Ind J Dermatol. 2013;58(5):405.
[6] Lan MY, Lan MC, Ho CY, Li WY, Lin CZ. Pilomatrixoma of the head and neck: A retrospective review of 179 cases. Arch Otolaryngol Head Neck Surg. 2003;129:1327-30.
[7] Byun JW, Bang CY, Yang BH, Song HJ, Lee HS, Shin JH, et al. Proliferating pilomatricoma. Am J Dermatopathol. 2011;33(7):754-55.
[8] Do JE, Noh S, Jee HJ, Oh SH. Familial multiple pilomatrico mass howing clinical features of a giant mass without associated diseases. Int J Dermatol. 2013;52(2):250-52.
[9] Bhushan P, Hussain SN. Bullous pilomatricoma: Astagein transition to secondary anetoderma? Indian J Dermatol Venereol Leprol. 2012;78(4):484-87.
[10] Ma KF, Tsui MS, Chan SK. Fine-needle aspiration diagnosis of pilomatrixoma. A monomorphic population of basaloid cells with squamous differentiation not to be mistaken for carcinoma. Acta Cytol. 1991;35:570-74.

[11] Barui GN, Karmakar R, Sinha A, Bhattacharya A. Pilomatrixoma: Misdiagnosed as a round cell tumor of soft tissue on fine needle aspiration cytology. J Cytol. 2009;26:125-26.

[12] Akther SQ, Shahriar S, Lima IJ, Islam MS, Sarker RD, Gupta AS, et al. An unusual diagnosis at neck: Pilomatricoma in a young female: A case report. Journal of Shaheed Suhrawardy Medical College. 2022;13(1):86-88.

PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Student, Department of Pathology, Indira Gandhi Medical College and Research Institute, Puducherry, India.
2. Professor, Department of Pathology, All India Institute of Medical Sciences, Guwahati, Assam, India.
3. Professor, Department of Surgery, Indira Gandhi Medical College and Research Institute, Puducherry, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Sankappa P Sinhasan,
Professor, Department of Pathology, All India Institute of Medical Sciences,
Silbharal Changsari, Guwahati-781101, Assam, India.
E-mail: drspsinhasan@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jul 26, 2022
- Manual Googling: Sep 21, 2022
- iThenticate Software: Oct 11, 2022 (3%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jul 24, 2022**
Date of Peer Review: **Sep 02, 2022**
Date of Acceptance: **Oct 12, 2022**
Date of Publishing: **Dec 01, 2022**