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Case Report

PILOMATRICOMA- A BENIGN HAIR MATRICAL TUMOR WITH A DIVERSITY OF DIFFERENTIATION

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ABSTRACT

Pilomatricoma, a benign neoplasm with hair matrical differentiation was earlier referred to as 'Calcifying epithelioma of Malherbe' because of the associated extensive calcification. More recently, it has been aptly rechristened as "Pilomatricoma" due to the predominant hair matrical differentiation that it exhibits. Besides its matrix, pilomatricoma may show diverse differentiation along the other constituents of the hair follicle like the inner sheath and infundibulum because of which it may present some diagnostic difficulties to the Pathologist. Barring these exceptional features, histopathology is the gold standard in the ultimate diagnosis of pilomatricoma. Histopathological features of trichelemmal cyst and basal cell carcinoma with basaloid differentiation closely resemble those of pilomatricoma. Distinction from basal cell carcinoma is extremely important because of the prognostic implications and the differences in management.

We present a case of pilomatricoma with classic histopathological features but with exceptional presenting features like occurrence at 70 years of age which is very unusual and absent calcification.

Keywords: Pilomatricoma, Matrical Differentiation, Hair Cortex, Benign, Skin Adnexal Tumor.

INTRODUCTION

Pilomatricoma is a benign, slow growing, asymptomatic tumor with differentiation towards hair cells particularly of the hair cortex. It occurs most commonly in the face and upper extremities. It may occur at any age but majority (60%) occurs in the first two decades of life and the rest in children younger than 10 years[1]. Calcium deposits are seen in about 75% of the tumors. In fact, it was due to this feature that the tumor was initially named as calcifying epithelioma of Malherbe when it was first described in the year 1880[2]. Though it was initially thought to arise from hair matrix cells, it is now believed to be a complex panfollicular neoplasm since it may exhibit differentiation of the entire follicle. Ackerman et al have described differentiation into other parts of hair follicle, like the inner sheath, the infundibulum and sometimes the sebaceous and the isthmic epithelium[3]. This makes this panfollicular neoplasm with divergent differentiation an exceptional entity, which can be easily mistaken for any neoplasm arising from any of these cell lines. We present a case of pilomatricoma with some divergent features like, elderly age at presentation and absent calcification. We also present a brief review of literature with an emphasis on the importance of differentiating this benign entity from basal cell carcinoma showing hair matrical differentiation.

CASE SUMMARY

A 70 year old male patient presented to the surgical department with a swelling over the right side of the face below the inner canthus of the right eye of 3 month's duration. He gave history of sudden increase in size. Local examination showed a solitary non- tender nodule measuring 3 cms x 2 cms over the right side of the face near the inner canthus of right eye. Skin over the swelling was pinchable. There was a focal area of surface ulceration. General examination revealed no significant findings. A provisional clinical diagnosis of "sebaceous cyst" along with the possibility of basal cell carcinoma to be ruled out was formulated. A complete wide excision was performed and the specimen was submitted for histopathological examination.

Histopathological findings:

Gross examination: The Specimen consisted of single, skin covered soft tissue mass which measured 2.3 x 1.6 cms.

Surface was nodular and showed focal ulceration. Cut section showed an encapsulated, circumscribed, nodular, grey-white to yellow tumor below the normal overlying skin.(Figure.1)

Microscopy: The epidermis was normal. The dermis showed a well encapsulated tumor with large, irregular islands of epithelial cells of two types (Figure 2). Central areas of keratinization and keratinized shadow cells(Figure.3)with peripheral areas of basaloid cells with indistinct cell borders.(Figure.4) The transition between the two cell types was abrupt in some areas and gradual at places. Foreign body reaction was seen adjacent to keratinized areas (Figure 5).

Histochemistry: Per-iodic- Schiff positivity was observed in the keratinized areas and the ghost cells.

Final diagnosis: Pilomatricoma

DISCUSSION

Pilomatricoma, was originally termed "Calcifying tumor of Malherbe" due to the calcification which is present in majority of these tumors[1].An occasional tumor which does not exhibit calcification is also not a hard nut to crack despite the absence of calcification when the other histopathologic features are pathognomonic as in our case. Considering that calcification is not an invariable or diagnostic feature, the term 'pilomatricoma' meaning that it is a tumor involving the hair cortex, seems more appropriate.

Ackerman et al in their extensive research on pilomatricoma, described this tumor as a sac of epithelium which is infundibular at the top, matrical along the sides and supramatrical below. They also observed that the matrical cells of pilomatricoma can cornify and turn into shadow cells or also differentiate and evolve along the other components of the hair follicle like the inner and outer root sheaths, sebaceous and infundibular structures[3]. This justifies consideration of pilomatricoma as a "panfollicular neoplasm". The histopathological features of pilomatricoma depend upon the predominant line of differentiation that the cells fall under.

The classic appearance with eosinophilic cells and basaloid cells indicative of matrical differentiation as seen in our case is a universal feature in all pilomatricomas. C. M. Simi et al in their study of 21 cases of pilomatricoma observed that, in addition to these features, all their cases also showed supramatrical differentiation in the form of cellular areas with pale stained, moderate cytoplasm but with an arrangement not as crowded as in matrical differentiation[4]. Focal areas of supramatrical differentiation were seen in our case. The differentiation observed in our case was predominantly matrical which was seen in the form of closely packed basaloid cells with round, pale stained cytoplasm, finely stippled chromatin and prominent nucleoli. Besides matrical and supramatrical differentiation, one may also find clear large cells with vacuolated cytoplasm. When present, they are indicative of outer root sheath differentiation. Root sheath differentiation was absent in our case. In occasional instances, focal sebaceous differentiation in the form of cells with vacuolated cytoplasm and scalloped nuclei may be present[4]. Sebaceous differentiation was not seen in our case.

Signs of infundibular differentiation which include keratohyaline granules and basket weave type of orthokeratosis may be occasionally present[4].They were absent in our case. Trichohyaline granules and keratinocytes which were observed by C. M. Simi et al in some cases [4] were not seen in our case. Ossification and apocrine cells, which were observed in a small percentage of cases by Simi et al were not seen in our case. Foreign body giant cell reaction observed in most of the other reported cases was also seen in our case.

The important differential diagnosis include trichelemmal cyst and basal cell carcinoma with basaloid differentiation[1].The basaloid cells in both these entities show peripheral palisading which is absent in pilomatricoma[1].Basal cell carcinoma very closely mimics pilomatricoma even in clinical presentation because of which it is often clinically misdiagnosed as basal cell carcinoma as in our case.

CONCLUSION

Pilomatricoma, though considered a tumor with hair matrix differentiation, fits in more appropriately under the category of panfollicular neoplasm owing to its capability to differentiate along divergent lines because of which it may pose diagnostic dilemmas. But its classic histopathological features and invariable presence of matrical and supramatrical differentiation help in resolving the confusion. Differentiating it from its closest malignant differential entity like basal cell carcinoma is imperative considering the prognostic implications.

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Figure 1: Encapsulated, circumscribed, nodular, grey white to yellow tumor

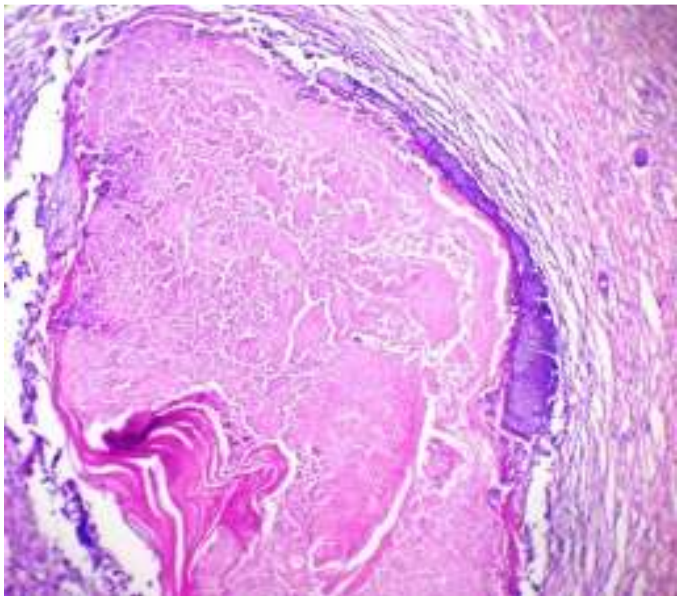

Figure 2: Large irregular island with central area of keratinization,ghost cells and peripheral basaloid cells, (H & E, ×100)

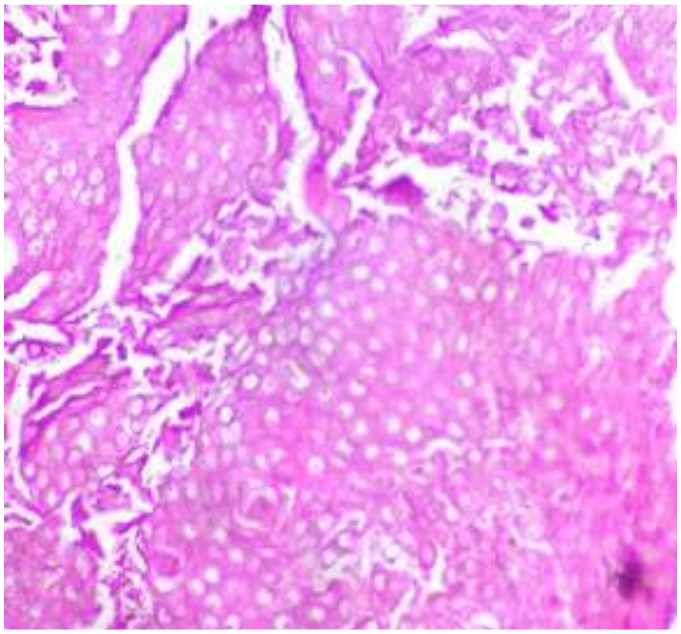

Figure 3: Keratinized shadow cells in the tumor, (H & E, ×400)

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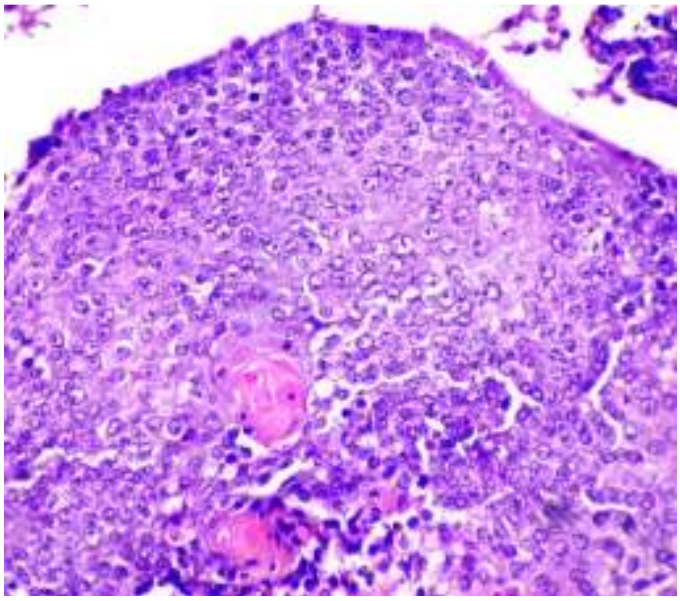

Figure 4: Basaloid cells with indistinct cell borders, (H & E, ×400)

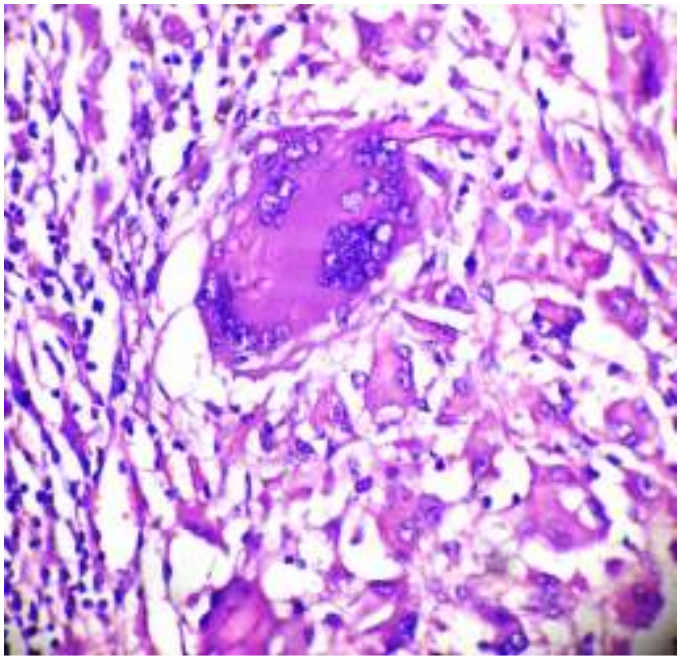

Figure 5: Foreign body giant cell reaction, (H & E, ×400)

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