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Calcifying Epithelioma of Malherbeat Unusual Sites: Fine Needle Aspiration Cytology Diagnosis and Review of Literature

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Abstract

A pilomatricoma is a benign appendage tumor related to hair cells matrix. Pilomatricoma usually occurs as a solitary firm lesion with predilection for head and neck, and upper extremities. The cytologic findings had features which allowed a correct diagnosis. Findings included are presence of basaloid cells, delicate pink fibres, shadow cells, giant cells, naked nuclei and calcium deposits. *Material and Method:* The present study includedcytologicallysuspected 8 cases ofpilomatricoma at unusual sites, confirmed on histology. *Conclusion:* Pilomatricoma generally present as firm subcutaneous nodules in children and young adults and are often misdiagnosed clinically. The physicians and surgeons should be familiar with this entity and consider it in the differential diagnosis of a superficial mass. FNA cytology is characteristic and will allow a conclusive diagnosis even in cases with an aberrant clinical presentation.

Keywords: Pilomatricoma; Unusual Sites; Calcifying Epithelioma; Aspiration Cytology.

Introduction

Pilomatricoma is a benign appendage tumor related to hair cells matrix [1]. This tumor was first described by Malherbe and Chenantais in 1880 as a benign, subcutaneous tumor arising from hair cortex cells. Since then, this uncommon entity has been called calcifying epithelioma of Malherbe [2]. They are slow growing, hard masses found beneath the skin. Most commonly they are seen on the face and neck but are sometimes found on the scalp, eyelids and arms. They are usually solitary, but occasionally, multiple masses are seen. Most cases occur in the first two decades of life. The condition is twice as common in females as males. Treatment is usually surgical excision. In cases with a typical presentation it should be possible to make a tentative clinical diagnosis of pilomatricoma. However, the variation in age, size and location makes it difficult in some cases even to suspect this tumor.The lesions are raised in relation to the skin surface and are characteristically hard and knobblies or multifaceted on palpation. They may be of normal skin color or white and may have a bluish tinge. Some can become inflamed, presenting as a granulomatous swelling, with occasional extrusion of calcium as a thick, white, gritty substance. Histopathology demonstrates a well-demarcated tumor, often with a connective tissue capsule. Irregularly shaped islands of epithelial cells are embedded in a rather cellular stroma. Usually two types of cells are seen within the islands. These cells can be seen transforming into "ghost" or shadow cells, which lack nuclei. They have a distinct border and a central unstained area which is a shadow of the lost nucleus. Calcification is commonly encountered as fine basophilic granules within the cytoplasm of the shadow cells or as large sheets of amorphous basophilic material. Ossification is a rare feature. The stroma of the tumor often shows a foreign body giant cell reaction adjacent to the shadow cells.

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Material and Methods

The present study includes analysis of cytological diagnosis of pilomatricoma, confirmed by histopathology. The aspirations were done by a 23 G needle attached

to a 10-ml disposable syringe. The smears made were air-dried and stained with May-Grünwald Giemsa (MGG), smears were also fixed in 95% ethanol and stained with Papanicolaou stain.

Clinical Details of Selected Cases were Shown in Table 1

A cytological analysis was done and following features were noted mainly: presence of basaloid cells, squamous nucleated cells, shadow cells, inflammatory cells, multinucleated giant cells, calcium deposits, naked nuclei, keratin, and most consistent finding pink fibrillarmaterial (Figure 1 a, b, c).

Unusual clinical presentation in case no. 2 as papillomatous growth in thigh with restricted mobility is unlikely for the diagnosis of pilomatricoma, even on cytology with the presence of characteristic feature like presence of basaloid cells and shadow cells. X-ray and ultrasound findings is compulsory before committing pilomatricoma in this particular case. All investigative modalities were unremarkable. Lesion was excised and confirm the diagnosis of pilomatricoma.

Another case no. 3 presented with dual mass - first at the midline neck swelling moves with the deglutination, suspected as thyroglossal cyst on sonogram. Slides prepared from aspirated material showed presence of basaloid cells, shadow cells and calcified material. Similar cytological features noted from another lesional mass in the thigh. Repeat guided FNAC was done to confirm our doubt of pilomatricoma, again showed similar cytological findings, confirmed on histopathology.

Because of the short duration of history in the case no. 5 clinician suspected this case as metastatic deposit, unlikely for the pathologist, was diagnosed as pilomatricoma.

Case no. 8 was suspected as fibroma clinically at the tip of the nose, cytology slide showed only presence of dystrophic calcified material, suspected as tumoral calcinosis with a differential of skin adnexal tumor. Histopathology also reveal predominantly presence of dystrophic calcified material with presence of ghost cells also, lead to the diagnosis of pilomatricoma.

Table 1: Clinical details of suspected pilomatricoma cases

S.N.	Age/Sex	Site of FNAC	Clinical details with duration	Clinical Diagnosis
1	18/F	Left eyebrow	Soft red-blue nodule, 2x2 cm - 8 months	Epidermal inclusion cyst
2	52/F	Inner aspect of left thigh	Papillomatous firm growth, 3.5x3 cm – 1year	Papilloma
3	58/F	Upper cervical region & right upper thigh	Soft lump 3x 3 cm moves with deglutination over the neck since 1 year& firm reddish-white nodule at thigh region 3x3.5 cm – since 18 months	Thyroglossal cyst – neck region and fibroma - thigh
4	35/M	Left eyebrow	Solitary cystic nodule, 1.5 cm – 5 months	Epidermal inclusion cyst
5	18/F	Upper inguinal region	Solitary nodule, 4x 4.5 cm- 1 months	Metastatic lesion
6	22/M	Forearm	Solitary nodule, 2x2.5 cm – 2 years	Fibroma
7	28/M	Forehead	Solitary nodule, 3x 3 cm – 1.5 years	Dermoid cyst /Sebaceous cyst
8	24/F	Tip of the nose	Solitary nodule,1.5x1.5cm – 2 years	Fibroma

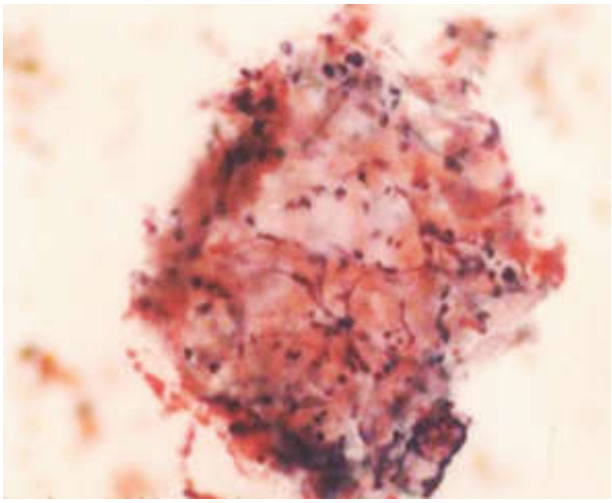

Fig. 1a. Photomicrograph show fragments of squamous epithelium with attached keratotic material and ghost cell. (Papanicolaou stain; 400X)

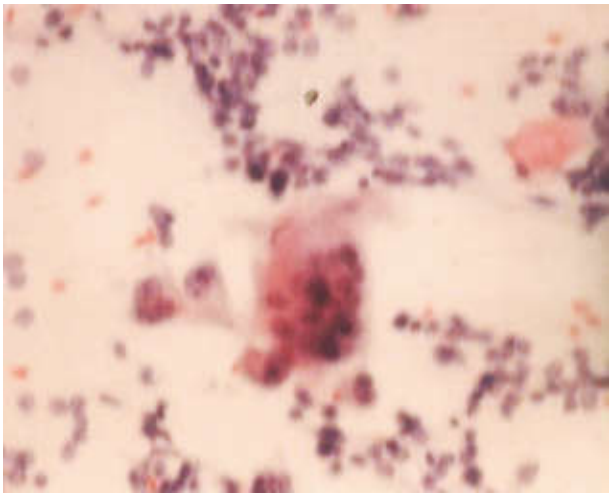

Fig. 1b. Photomicrograph show multinucleated giant cells of foreign body type and loosely dispersed germinative epithelial cells. (Papanicolaoustain; 200X)

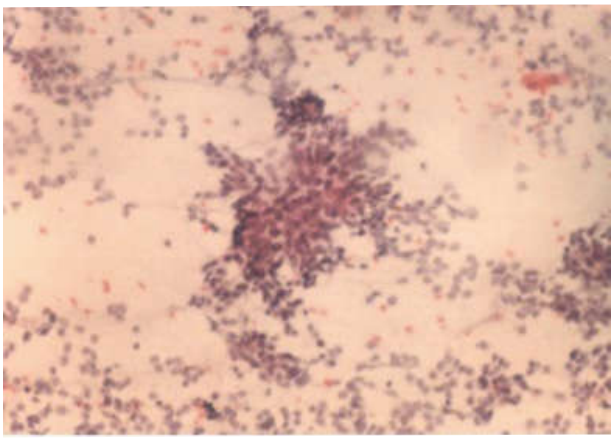

Fig. 1c. Photomicrograph show clusters of cohesive and isolated bare nuclei germinative epithelium- basaloid cells. (Papanicolaou stain; 200X)

Discussion

Pilomatricoma, referred to as calcifying epithelioma of Malherbe is uncommon skin tumor that occur predominantly in young people [1]. It usually present as a solitary firm, deep seated subcutaneous nodule and the overlying skin frequently show bluish-red discoloration [2]. It is more common in females than males. Pilomatricoma was first described by Malherbe and Chenantais in 1880 as calcified epithelioma of sebaceous gland [3]. Turhan and Krainer in 1942 advanced the concept that the tumor is derived from hair matrix or cells that are hair matrix like [4]. Histochemical staining reaction confirmed the origin of the tumor from hair matrix [5]. Forbis and Helwig on this basis designated it as pilomatricoma [6].

Though it may occur at any age,60% cases have been reported to occur in first 2 decade of life [1]. Behnke et al had reported occurrence of pilomatricoma in 4 elderly patients [7]. In Kaddu et al study 58 patients out of 118 patients were more than 45 years of age [8]. There study showed that clinical and histopathological features in more than 45 years of age were similar to in patients < 45 years of age. Differential diagnosis in both group were also comparable except in some cases where differential diagnosis included basal cell carcinoma, keratoacanthoma and metastasis. Recurrence of the lesion after simple excision as observed in one patient over 45 years of age.

Literature report suggests that in about 56-72% cases, Pilomatricoma occur in the head and neck region [9]. They are uncommon tumors on the eyelid and eyebrow region of young adults and are frequently misdiagnosed when evaluation is based on clinical evidence alone [10]. In Yap et al study of patients with tumors involving the eyelids and eyebrows, all the cases were 20 years old or younger. Tumor diameter was 1 cm or less in all the cases and most of them were misdiagnosed preoperatively [11]. One of our cases presented with a cystic swelling in the neck moving with deglutition and was misdiagnosed as a case of thyroglossal cyst. Pilomatricoma is commonly misdiagnosed preoperatively as an epidermoid or dermoid cyst. Fine needle aspiration cytology findings seen in pilomatricoma patients are cellular smears comprising primarily of clusters of small primitive appearing basaloid cells with overlapping nuclei. The basaloid cells had relatively large, round, regular and basophilic nuclei with evenly dispersed chromatin and large nucleoli. Sheets of ghost epithelial cells, multinucleated giant cells with squames and keratin fragments are also seen [12].

The results of the cases discussed here, is possible to arrive at a conclusive diagnosis of pilomatricoma on FNA smears after a careful analysis of all cytological features, even in cases with an uncommon clinical presentation. Thus the finding of a smear with clusters of tightly arranged basaloid cells surrounded by delicate fribrillarmaterial, keratin flakes, shadow cell, giant cells, calcium deposits and numerous naked nuclei with inflammatory cells in the background should lead to a diagnosis of pilomatricoma [13,14]. Naked nuclei with distinct nucleoli mixed with shadow and basaloid cells were reported as the main cause of a false-positive diagnosis of malignancy. [13-15]

The differential diagnosis should include giant cell tumour, epidermal inclusion cyst, basal and squamous cell carcinoma, Merkel cell tumour, adnexal tumour and basal cell adenoma from salivary gland [13,15].

A predominance of giant or shadow cells with a few basaloid cells should suggest a cytologic diagnosis of giant cell tumour or epidermal inclusion cyst, respectively. Dissociated small fragile cells with moulding and individual cell necrosis would favour Merkel cell tumour. In basal cell carcinoma, well defined clusters with peripheral palisading and sharp borders are prominent cytological features. None of these tumour show the typical clusters of basaloid cells surrounded by a delicate red (MGG) fibrillar material, calcium deposits and naked nuclei found inpilomatricoma.

Our cases contains several interesting aspects in its clinical profile. First, despite the fact that these tumors can be seen at unusual sites. Second the lesions showed atypical clinical presentation which is not a regular finding of pilomatricoma. Having all these peculiarities makes these cases quite unique and increases the value of keeping pilomatricoma as a differential diagnosis in patients presenting with such

clinical features.

Following recommendation have been made to avoid errors.

1. The FNAs should be carried out and the smears should be interpreted by the same person.
2. Clinical data, particularly age and location are of paramount importance.
3. Shadow cells are pathognomonic of pilomatricoma's.
4. Basaloid nuclei with prominent nucleoli should not be overdiagnosed.
5. Both Papanicolaou and Giemsa stained slides should be used.
6. Think of pilomatricoma when performing and interpreting aspirations from subcutaneous growths located in any region and in any age group.

Conclusion

Pilomatricoma is an uncommon tumor that occurs on the eyelids and of brow region of children and adolescents. It is commonly misdiagnosed preoperatively. Distinctive clinical features, however, suggest the correct diagnosis. Pilomatricoma should be kept in the differential diagnosis even at unusual sites. The cytologic presentation had features which allowed a correct diagnosis in all our cases and included basaloid cells, delicate pink fibres, shadow cells, giant cells, naked nuclei and calcium deposits. It is concluded that in pilomatricoma, FNA cytology is characteristic and will allow a conclusive diagnosis even in cases with an aberrant clinical presentation.

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