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Dermatology

A CASE REPORT OF MULTIPLE BENIGN CALCIFYING PILOMATRICOMAS OF SCALP

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ABSTRACT Pilomatricoma (PMC) is a relatively uncommon benign skin neoplasm arising from the skin adnexa. Due to varied presentation, difficulties persist in making the clinical and cytologic diagnosis. Multiple pilomatricomas on the other hand, are rare. They have been associated with various syndromes of which myotonic dystrophy has been the most described(1). We report the clinical and histopathological findings of a case of pilomatricoma with unusual presentation. In this case, a 36-year-old female presented with multiple asymptomatic swellings in the scalp. However, subsequent histopathologic examination confirmed the case as pilomatricoma. This report reveals that pilomatricoma is a frequently misdiagnosed entity in clinical practice.

KEYWORDS : Pilomatrixoma, calcifying epithelioma of Malherbe, hair matrix, basaloid cells.

INTRODUCTION

Pilomatricoma is also known as Trichomatricoma or Benign calcifying epithelioma of Malherbe. It is a benign tumor of hair matrix differentiation (hamartoma), which is considered to differentiate toward hair follicular cells, particularly the cortex but can show cellular evolution toward the other parts of the hair follicle, such as the outer and inner root sheaths, sebaceous and infundibular components. Therefore, it can be considered a panfollicular neoplasm.

It is the commonest hair follicle tumor, accounts for 20% of all hair follicle related tumors. It is frequently seen in children, may occur at any age with slight female preponderance.

The lesion is usually a solitary, deep, dermal, or subcutaneous tumor 3–30 mm in diameter situated on the head, neck, or upper extremities. The skin over the tumor is normal and the lesion has a firm to hard consistency indicating calcification and a lobular shape on palpation.

Some familial cases are reported. Genetic studies reveal that 75% of pilomatrixoma possess activating mutations of the β‐catenin gene, CTNNB1. High levels of β‐catenin increase cell proliferation, inhibit cell death leading to neoplastic growth. Associations with myotonic dystrophy, Turner syndrome, and Rubinstein–Taybi syndrome have been described. MYH associated polyposis, a variant of familial adenomatous polyposis with susceptibility to colorectal carcinoma is associated with multiple pilomatricomas.

Histopathology typically consists of basaloid, eosinophilic, and shadow cells admixed with multinucleated giant cells and areas of calcification(2). The tumor is situated in the dermis and is composed of well circumscribed rounded islands giving a lobulated contour. The outer cells are small, and their rounded nuclei crowded together to make this region deeply basophilic. The cytoplasm is scanty and the cell margins indistinct, but intercellular connections can be seen. Towards the center of the mass, the cytoplasm becomes more abundant and eosinophilic. The nuclear outline persists, but the chromatin is sparse and clumped in dark granules, when all basophilic material disappears, a mummified 'ghost cell' remains. The stroma that encapsulates the masses usually contains inflammatory and foreign body giant cells, and occasionally ossifies.

The course is usually benign. Malignant change may arise, chiefly in large tumors.

Local excision is the main modality of treatment with reported recurrences.

CASE DESCRIPTION

A 36-year-old female who is a homemaker, presented with multiple asymptomatic swellings in the scalp for 1 year. History of similar lesion on scalp four years back for which excision was done at a local hospital.

On examination, a single skin-colored non-tender nodule of 1.5x1.5 cm size with smooth surface and firm consistency was noted on the vertex of the scalp in the midline, 10 cm above the hairline. The skin over the swelling was normal.

Four nodules of sizes ranging from 0.5cms to 1cm, firm to hard in consistency, and non-tender were noted on both parietal regions of the scalp and the tenting sign was positive for them.

No growths or lesions were noted elsewhere on the body.

No systemic signs of illness were present. The patient gave no history of recent trauma, surgery, infection, or radiation.

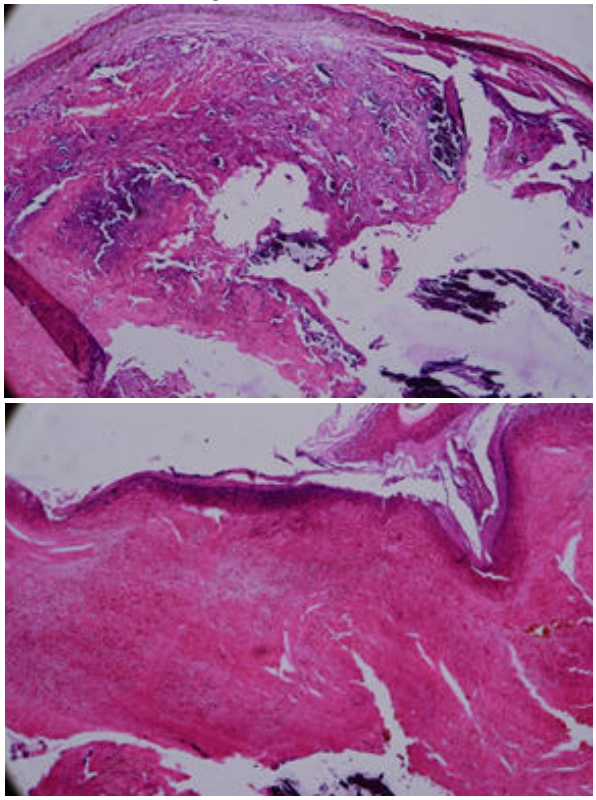
Serum calcium levels were normal.

An excisional biopsy was done for the large tumor on the midline of the vertex and the specimen was sent for histopathological examination.

The clinical differential diagnoses were pilomatricomas, pilar cysts, calcinosis cutis

Fig: pilomatricoma- small, firm, skin-colored nodule.

Histopathological examination showed a relatively well-circumscribed cyst composed of a basaloid proliferation resembling the hair matrix cells, maturing into structureless eosinophilic cells lacking nuclei “shadow cells”. Focal areas of calcification are seen within the shadow cell regions.



DISCUSSION

Pilomatricomas are commonly misdiagnosed, benign neoplasms of the skin, thought to arise from hair follicles(3). They appear most frequently as solitary, firm, deeply seated, nontender subcutaneous nodules adherent to the skin but not fixed to the underlying tissue. Stretching of the skin over the tumor shows the "tent sign" with multiple facets and angles, a pathognomonic sign for pilomatricoma. In addition, pressing on one edge of the lesion causes the opposite edge to protrude from the skin like a "teeter-totter". Both these "tent sign" and "teeter-totter sign" are the most helpful clinical clues to the diagnosis of pilomatricoma(4).

Pilomatricomas are asymptomatic and usually found in the head and neck area and, upper extremities, but rarely identified on the chest, trunk, or lower extremities. Four distinct morphological stages of pilomatricoma are defined as: (a) early: small and cystic lesions, (b) fully developed: large and cystic, (c) early regressive: foci of basaloid cells, shadow cells, and lymphocytic infiltrate with multinucleated giant cells, (d) late regressive: numerous shadow cells, absence of basaloid and inflammatory cells. The only truly reliable means of diagnosis remains pathological evaluation. The classic histology is defined by the presence of ghost or shadow cells and basaloid cells. At low power, it is usually seen as a well-circumscribed nodulocystic tumor. Calcium deposits are present in well over half the lesions identified. Thus, the skin lesion is also described as a calcifying epithelioma(4).

CONCLUSION

This case is reported because of its unusual presentation with multiple tumors in the 4th decade and with no associated conditions.

Multiple pilomatricomas are usually associated with other conditions. Hence long term follow-up is recommended for these patients.

No conflicts of interest.

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