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Case Report

Multiple malherbe's calcifying epithelioma: A case report

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ABSTRACT

Pilomatricoma, is an appendageal tumour of the skin usually seen in head and neck area. They are usually solitary, derived from the ectoderm and arise from the outer root sheath cells of the hair follicle. Multiple pilomatricomas are rare. We present a case of multiple pilomatricomas over the arm of a 13 year old girl.

1. Introduction

Pilomatricoma, also known as calcifying epithelioma of Malherbe, is an appendageal tumour of the skin seen in head and neck regions.[1] Embryologically, Pilomatricomas are derived from the ectoderm and arise from the outer root sheath cells of the hair follicle.It is more commonly seen in children. It presents clinically as a well-demarcated, hard, mobile superficial nodule that slowly progresses in size. Although all hairy areas of the skin can be involved, pilomatricoma frequently occurs in the head and neck region and is usually solitary. Involvement of the upper extremities is relatively uncommon.[2,3] We report a case of an unusual presentation of multiple pilomatricomas in the arm of a 13-year-old girl.

2. Case Report

A 13 year old female patient presented with well defined nodules. One was over the right arm just above the elbow and one was over the left shoulder. Both lesions were present since one year. Lesions were initially small and gradually progressed in size over a period of one year. There was no history of insect bite prior to onset of the lesions and no history of pain or discharge from the lesions. Patient gave a history of similar lesion over the right arm 6 years ago which was excised 4 years ago. There was no history of neurological or musculoskeletal abnormalities.

Family history was negative.

On examination, 1- solitary nodule measuring 3x4 cm in size, mobile, firm, non-tender and not fixed to the skin present over right arm (Figure 1). Tenting sign was positive.

2- solitary nodule measuring 4x4 cm in size, mobile, firm, non-tender and not fixed to the skin present over left shoulder, overlying skin was thinned out and atrophic. Tenting sign was positive.

Pallor, icterus, cyanosis, clubbing, lymphadenopathy and edema were absent.

There were no systemic features present.

FNAC report revealed keratinous cyst.

Excision biopsy was performed on both lesions.

Gross description, 1- The excised specimen revealed an irregularly shaped nodular tumor mass in the reticular dermis, firm in consistency, calcified in its central portion, measuring 3x4 cm, and was yellowish-white in color.

2- The excised specimen revealed a well circumscribed nodular tumor mass in the reticular dermis, extending into the subcutaneous fat, firm in consistency, calcified in its central portion, measuring 4x4 cm, and was chalky-white

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in color.

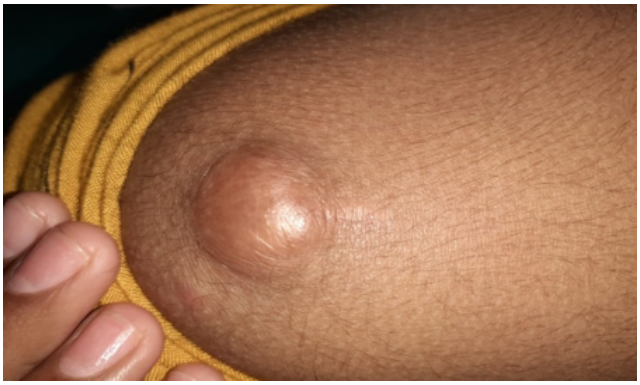

Fig. 1: Solitary nodule on the right arm

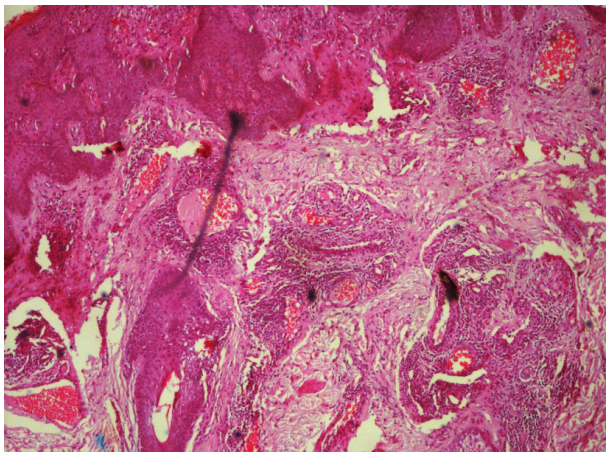

Fig. 2: Irregularly shaped well-circumscribed masses of epithelial cells in mid and lower dermis

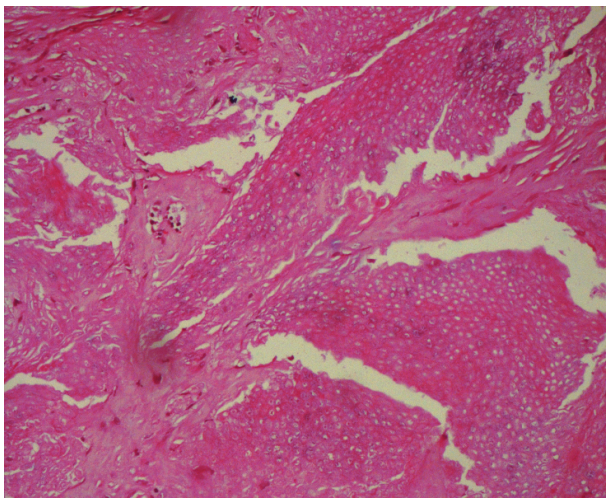

Fig. 3: Islands of basaloid cells with central area showing ghost cells

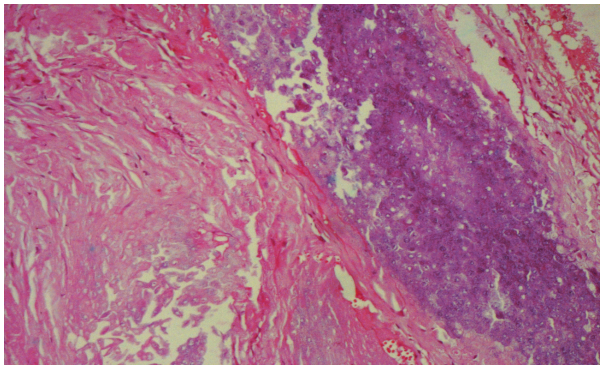

Fig. 4: High power view showing calcified areas and ghost cells

Histopathology revealed- Histopathologically, hematoxylin and eosin stained sections from both the specimens revealed few well demarcated lesions in the dermis and extending into the subcutaneous fat in a few areas. The lesions consist of islands of epithelial cells comprised of both basophilic cells with scanty cytoplasm and eosinophilic 'ghost cells' that have a central unstained area with loss of nucleus. High powered view showed some areas of calcification.

3. Discussion

Pilomatricoma also known as calcifying epithelioma of Malherbe, is a benign skin appendageal tumor with differentiation towards the matrix cells of the hair follicle. This lesion occurs over a wide age range with two peaks in the age less than 20 years and over 50 years.[4]

Pilomatricomas constitute 0.12% of all skin tumors.It has a female to male ratio of 1.5-2.5:1.[5]

Pilomatricomas are usually found in head and neck region, though it has been occasionally also reported in upper extremities and other sites.[4]

Pathogenesis is related to abnormal position of follicles which are located very deep in the dermis due to which regular differentiation induction agents cannot act on them properly. As a result of this, such partially differentiated follicles form pilomatricomas. These tumors can be familial, and maybe associated with Gardner's syndrome, Steinerd's disease, and Sarcoidosis.[5]

Pilomatricomas are generally asymptomatic and pain only occurs with associated inflammation and ulceration. They are solitary, deeply seated, firm, nontender subcutaneous masses adherent to the skin but not fixed to the underlying tissue.

A pathognomonic sign for pilomatricoma involves stretching of the skin over the tumor which shows the "tent sign" with multiple facets and angles.In addition, pressing on one edge of the lesion causes the opposite edge to protrude from the skin like a "teeter- totter". Both these signs are the most helpful clinical clues to the diagnosis of

pilomatricoma.[6]

The blue red discoloration of the overlying skinis another typical feature of pilomatricoma which definitely excludes the possibility of epidermal inclusion or dermoid cyst.

The accuracy rate of the preoperative diagnosis of pilomatricoma ranges from 0% to 30%.[6]

Fine needle aspiration can be used to diagnose pilomatricoma. The presence of basaloid cells, ghost cells, and foreign body giant cells are essential for a confident diagnosis of pilomatricoma. Additional supporting features included calcification, naked nuclei, and nucleated squamous cells. Differentials on FNAC were squamous cell carcinoma, giant cell lesions, and epidermal inclusion cysts. However, skin biopsy remains the definitive means of diagnosis.[7]

Histopathology of pilomatricoma shows lobules and nests of epithelial cells composed of two cell types i.e-basophilic cells and eosinophilic shadow cells. Early lesions show a predominance of basophilic cells grouped in islands at the tumor periphery. With tumor progression, the basophilic cells acquire more cytoplasm and gradually lose their nuclei to become eosinophilic shadow cells. These shadow cells constitute the central portion of the tumor and frequently calcify. Gradually this calcified foci increase in size, thus imparting the firm consistency to the lesion.[6]

Multiple pilomatricomas in a single patient is rare. The occurrence of multiple pilomatricomas has been associated with the development of myotonic dystrophy (MD). Myotonic dystrophy usually presents during the teenage years or early adulthood, but the onset of pilomatricomas ranges from many years before to many years after the onset of symptomatic MD. There have also been various reports of other syndromes being associated with multiple pilomatricomas, including Turner syndrome, Gardner syndrome, and sarcoidosis.[7]

The standard treatment of pilomatricoma is complete surgical excision. Spontaneous regression is never observed and malignant transformation is rare. There is an incidence of 0% to 3% chance of recurrence of the lesion after surgery. Malignant transformation to a pilomatrix carcinoma should be suspected in cases with repeated recurrences at the same site.[6]

4. Conclusion

The present case highlights the importance of considering pilomatrixoma in the clinical and pathologic differential diagnosis of dermal or subcutaneous nodule even in locations other than head and neck region. To conclude, pilomatricoma, a tumor deemed to be of hair matrix differentiation, commonly shows cellular evolution toward other parts of the hair follicle such as the outer and inner root sheaths and sebaceous and infundibular components. These features are often not stressed in the literature. If one is not aware of this fascinating array of appearances, it is quite likely that the diagnosis may be missed.

5. Acknowledgements

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6. Conflict of Interest

None.

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