CALCIFYING EPITHELIOMA OF MALHERBE IN THE PINNA: A CASE REPORT WITH REVIEW OF LITERATURE

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HOW TO CITE THIS ARTICLE:
Vinay Kumar R, Siva Ranjan D, Sunil Kumar K, Rama Mohan Rao B, Epari Sanjeeva Rao. "Calcifying epithelioma of malherbe in the pinna: a case report with review of literature". Journal of Evolution of Medical and Dental Sciences 2013; Vol. 2, Issue 45, November 11; Page: 8845-8848.

ABSTRACT: Benign calcifying epithelioma of Malherbe or pilomatricoma is an uncommon lesion in the pinna, arising from the matrix cells at the base of the hair. Histologically it was characterized by the presence of ghost cells, basophilic cells and foreign body giant cells. It is a rare benign tumor, starting in the hair matrix and clinically resembling a hard subcutaneous swelling. We report a case of 50 year-old female with a pilomatricoma in the pinna. We discuss the clinical, histopathological characteristics of pilomatricoma with review of literature.
KEY WORDS: Calcifying Epithelioma, Pilomatricoma, Pinna

INTRODUCTION: Pilomatricomas are of ectodermal origin and arise from the outer root sheath cell of the hair follicle. Pilomatricomas usually are asymptomatic, deeply seated, firm, non tender subcutaneous masses adherent to the skin but not fixed to the underlying tissue.[1] Pilomatricoma, or calcifying epithelioma of Malherbe, was first described in 1880 by Malherbe and Chenantais.[2] They described it as a benign subcutaneous tumor arising from sebaceous glands. In 1922, Dubreuilh and Cazenave described the unique histopathological characteristics of this neoplasm, including islands of epithelial cells and shadow cells.[3] In 1961, Forbis and Helwig proposed the term pilomatricoma, to describe the condition to avoid a connotation of malignancy and denote its origin from hair matrix cells.[4] In 1977, the term was changed as pilomatricoma to be more correct etymologically.[5]

CASE REPORT: A 50-year-old woman with an unremarkable medical history, presented with a nodule in the pinna. The patient had noted the mass in her left pinna, of one year duration. The lesion had begun growing over the last few months. There was no history of pain, infection or trauma. Clinical examination revealed a solitary, firm, well-circumscribed mass located over left pinna.[Figure 1] Surgical excision was done and the specimen was sent to pathology lab. Since it was not commonly suspected preoperatively over pinna, certain distinctive clinical features of tumor should suggest clinical diagnosis followed by histopathological confirmation.

Pathological findings: Gross examination revealed nodular mass measuring one centimeter in diameter, well-circumscribed, firm and gritty to cut. Cut surface showed grayish white areas. Microscopic examination showed numerous lobules with basophilic cells in the periphery and of ghost-like squamous cells towards the center with a few anucleated cells. [Figure 2] There were numerous foci of calcification, a few foreign body giant cells [Figure 3] and a few lymphoplasmacytic infiltrates. The histopathological features were consistent with a diagnosis of pilomatricoma (benign calcifying epithelioma of Malherbe).

DISCUSSION: Pilomatricoma is an uncommon lesion that arises from the matrix cells at the base of the hair. Since the first description of Pilomatricoma in 1880, there has been a gradual increase in understanding of the morphologic features and clinical presentation of this tumor. It was first described by Malherbe as benign calcifying epithelioma.[6] Subsequently numerous ultrastructural and electron microscopic studies[7,8] provided strong evidence of its origin from the matrix cells and the term "Pilomatricoma" was then coined by Forbis and Helwig keeping the histogenesis into consideration.[9] Pilomatricoma has been reported not only as a benign lesion, or as a low-grade malignant lesion with a tendency to recur locally, but also as a highly malignant tumor.

Pilomatricoma are found particularly on the head and neck region (over 50% of cases) with a female predominance.[10] Other locations include the upper extremity, trunk and lower extremity in decreasing order of frequency. A very few cases were reported in the pinna. No cases have been reported on the palms, soles or genital region.[11] The present case was reported in 50 year old female who presented with a swelling in the left pinna. Pilomatricoma can occur at any age but demonstrates bimodal peak presentation during the first and sixth decades of life. However; 40% of cases occur in patients younger than 10 years of age and 60% of cases occur within the first two decades of life.[12]

Although pilomatricoma is an uncommon benign tumor and frequently misdiagnosed as epidermoid or dermoid cyst, it has some distinctive clinical features that suggest the correct diagnosis. Pilomatricomas usually are asymptomatic, firm, non tender subcutaneous masses 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.[13]

Clinically differential diagnosis includes epidermoid cysts, dermoid cyst, sebaceous adenoma or carcinoma, juvenile xanthogranuloma, capillary hemangioma, and rhabdomyosarcoma. Although they grow slowly, they occasionally demonstrate rapid growth and may resemble keratoacanthoma. They can rarely undergo malignant transformation into pilomatrix carcinoma.[14] Histopathological examination is must for confirmation of diagnosis.

Macroscopically the specimen was firm to hard, calcified in its central portion, measuring, 1 x 1 cm and was grayish-white in color. Histopathologically, the hematoxylin and eosin stained sections showed a tumor composed of an epithelial component exhibiting the typical population of basaloid and ghost cells. The basaloid cells were round to oval, hyperchromatic nuclei and scanty cytoplasm. The ghost cells were eosinophilic with a central unstained shadow in the site of the lost nucleus. In addition, a few multinucleated giant cells and areas of calcification were present. Based on these histopathological findings it was diagnosed as pilomatricoma.

To conclude Pilomatricoma is a rare benign neoplasm derived from hair follicle matrix cells. Review of world's literature has showed that this was a rare case of a calcifying epithelioma of Malherbe in the pinna. The clinical course is generally benign although, malignant transformations have been reported. Management includes a complete surgical excision. Recurrence is uncommon after adequate excision.

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Fig. 1: Nodule over the left pinna

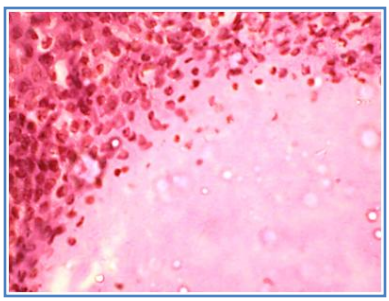

Fig. 2: Basophilic cells in the periphery and of ghost-like squamous cells towards the center. [H&E, x400].

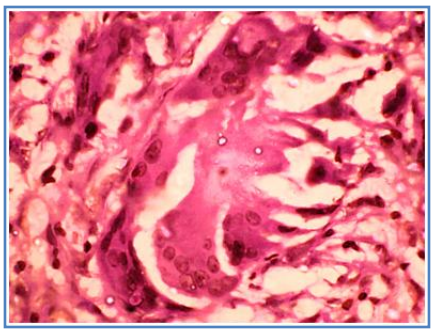

Fig. 3: Numerous multinucleated foreign body giant cells. [H&E, x 400].

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Date of Submission: 27/10/2013.
Date of Peer Review: 28/10/2013.
Date of Acceptance: 05/11/2013.
Date of Publishing: 07/11/2013