ABSTRACT: We have studied a total of 59 skin adnexal tumors that has been reported for the past 10 years (2004 – 2014) in our hospital and we have documented that 7 cases of them are pilomatricoma. Features of myeloid metaplasia with extensive areas of calcification was observed with an old hematoma in only one case. Herewith a case of ancient/old pilomatricoma with extensive areas of calcification and ossification along with the formation of marrow elements is being presented. The lesion occurred in the shoulder region of a 16 year old female. Pilomatricoma (Calcifying epithelioma of Malherbe) is a tumor of hair follicle, composed of basaloid cells with squamous differentiation and formation of shadow cells.

KEYWORDS: Pilomatricoma, Extensive calcification, Ossification with myeloid metaplasia, Extramedullary hematopoiesis.

INTRODUCTION: Pilomatricoma is a benign follicular skin appendage tumour first described by Malherbe and Chenantais. In 1880, they described calcifying epitheliomas, initially thought to be tumors of the sebaceous glands. Malherbe corrected this view in 1905. Since then, this uncommon entity has been called “calcifying epithelioma of Malherbe”. The term pilomatrixoma proposed by Forbis and Helwigin 1961 was subsequently changed to the more etymologically correct pilomatricoma in 1977. Anatomically, the head and neck is the favoured site, representing more than 50% of its distribution.

MACROSCOPY: The specimen was 2cms in diameter, bony hard in consistency and was gritty to cut. Cut sections showed friable white material along with firm to hard areas. The entire lesion was well circumscribed.

MICROSCOPY: A well circumscribed lesion with extensive areas of calcification and ossification was noticed. There were squamous cells with adjacent shadow cells. Fibro fatty tissue among the seams of bone containing mononuclear, round, poorly differentiated cells approximately twice the size of lymphocytes were observed. Also seen were cells approximately 4 times the size of lymphocytes. These large cells had nuclei which were smooth as well as lobulated in configuration, probably representing megakaryocytes. The morphology of all these cells appears similar to that of marrow elements. There are collections of cholesterol clefts along with adjacent osteoclastic type of giant cells.

FINAL DIAGNOSIS: Pilomatricoma with extensive areas of calcification and ossification and evidences of myeloid metaplasia, secondarily complicated by an organized hematoma.
DISCUSSION: Pilomatricoma is a tumor of hair follicle origin. The tumors of hair follicles can be classified as: benign tumors and malignant tumors.

The benign tumors are: Trichofolliculoma, Pilar sheath acanthoma, Fibrofolliculoma, Trichodiscoma, Trichoeplithelioma, Trichoblastoma, Tricho adenoma, Pilomatrixicoma, Trichilemmoma, Trichilemmal horn and Proliferating trichilemmal cyst.

The malignant tumors are: Pilomatrix carcinoma, Malignant proliferating trichilemmal tumor, Trichilemmal carcinoma and Trichoblastic carcinoma.

Some of them are cystically dilated in the case of pilar cyst.

Pilomatricoma is a tumor with differentiation towards the hair matrix such as basophilic cells, shadow cells\(^1\), outer root sheath\(^2\) and any structure of the follicle with the presence of follicular germinative cells.\(^3\)

Four clinical variants have been described: an eruptive type,\(^4\) a perforating type,\(^5\) a familial type associated with myotonic dystrophy\(^6\) and a recurrent invasive, non-metastatic pilomatrix carcinoma.\(^7\)

Noguchi et al suggested that the distribution of pilomatricoma corresponds to the density of hair follicles at a particular site.\(^8\) Pilomatricoma are commonly seen on face and scalp. The next common location is upper extremities.

It is a known fact that no new hair follicles are formed after birth, but partially formed follicles might be dormant and become activated at puberty or in later life.\(^9\) The position of these follicles might be a factor that determines their fate or subsequent reactivation.\(^10\) If they lie too deep for inducing agents to exert their normal effects, then these dormant structures might on reactivation only partially differentiate and form a pilomatricoma.\(^11\)

Pilomatricoma often present as gradually enlarging, asymptomatic, mobile, hard subcutaneous mass. In the more superficial lesions in a study, the overlying skin was often stretched, and calcifications could be seen as yellow or white flecks throughout the surface.\(^12-16\)

As a rule, two types of cells, basophilic cells and shadow cells comprise the islands. Calcification occurs in more than two-thirds of the tumors and is usually in the shadow cells. Ossification of the stroma occurs in about 13%.

Extramedullary hematopoiesis may occur adjacent to the spicules of bone. Bone morphogenetic protein (BMP)-2, which plays an important role in ectopic bone formation, has been found in the shadow cells, suggesting that it may play a role in generating bone formation in pilomatrixomas.

Osteoblasts are derived from primitive stem cells. Marrow is often found in the metaplastic bone. It could either be the result of the local mesenchymal cells or else follows the deposition of primitive haematopoietic cells from the blood stream.

Cutaneous extramedullary hematopoiesis has been encountered exceptionally in association with haematological malignancies. The localized form of extramedullary hematopoiesis is not associated with haematological disorders or bone marrow disturbances and is usually seen in the context of degenerative tissue changes, especially necrosis and calcification.\(^17\)

Ackerman et al have lately depicted old lesions of pilomatrixoma with histopathologic features of bone marrow tissue, including metaplastic calcification, osteoclasts and a highly vascular stroma.\(^18\) Histopathological findings of extramedullary hematopoiesis were noted in seven of 120
cases of pilomatricoma. In six of the lesions, variable areas of calcification were noted. Osseous metaplasia, however, was present in only two cases.

In our institution, we studied 59 cases of skin adnexal tumors during the period of 2004-2014, of which 7 were pilomatrixicoma. Among the 7 cases only one showed features of myeloid metaplasia with extensive areas of calcification with an old hematoma.

CONCLUSION: Pilomatrixicoma is an uncommon tumor but still rarer is the one with extensive areas of calcification, ossification, and formation of myeloid metaplasia. Secondary hematoma with formation of cholesterol clefs and giant cell reaction can occur as was observed in our study report.

EQUIPMENT USED:
Nikon Coolpix-8400.
x denotes the power of objective.
Stain used – (H & E).

MICROSCOPIC IMAGES

Clusters of squamoid cells and shadow cells. Field also shows mature lamellar bone and areas of calcified necrotic bone and calcification.

Clusters of shadow cells and osteoclastic giant cells.

Fig. 1: H & E stained 4 x
Fig. 2: H & E stained 10 x
Fig. 3: H & E stained 40 x
Fig. 4: H & E stained 40 x
Necrotic bone surrounded by osteoclastic giant cell.

Cholesterol clefts.

Fat spaces, fibro collagenous tissue and dark marrow elements.
Marrow elements.

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AUTHORS:
1. M. Valluvan
2. Divvya B
3. P. Viswanathan
4. Rehana Tippoo
5. R. Ramesh

PARTICULARS OF CONTRIBUTORS:
1. 2nd Year Post Graduate, Department of Pathology, Rajah Muthiah Medical College, Annamalai University.
2. 2nd Year Post Graduate, Department of Pathology, Rajah Muthiah Medical College, Annamalai University.
3. Professor, Department of Pathology, Rajah Muthiah Medical College, Annamalai University.
4. Professor, Department of Pathology, Rajah Muthiah Medical College, Annamalai University.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:
Dr. P. Viswanathan,
Professor,
Department of Pathology,
Faculty of Medicine,
Rajah Muthiah Medical College,
Annamalai University,
Chidambaram-608002,
Tamilnadu, India.
Email: drpviswanathan2013@gmail.com

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