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Case Report

Pathology Section

Extramedullary Haematopoiesis in a Case of Pilomatricoma

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ABSTRACT

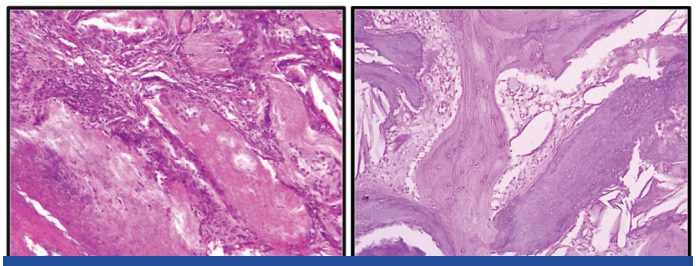
Pilomatricoma is a benign follicular skin appendage tumour. It usually occurs as solitary lesion and is most commonly seen in the face and upper extremeties. Here we report a rare case of extramedullary haematopoiesis (EMH) in pilomatricoma in a 38-year-old lady.

Keywords: Adnexal tumours, Calcification, Ossification

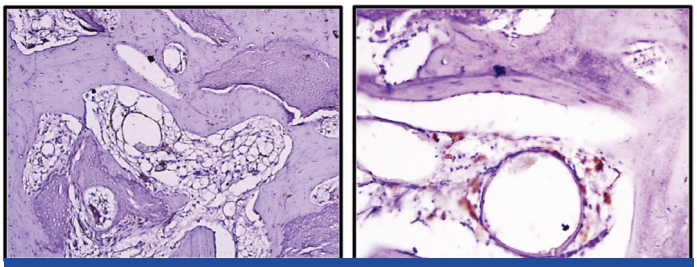
CASE REPORT

A 38-year-old lady presented with a left shoulder swelling. The swelling had been present for the last 5 years and showed gradual enlargement since the last 1 year. On examination a firm swelling was noted in the left shoulder measuring 2x1.5x1cm. The swelling was not fixed to overlying skin. She had no other significant history. No significant haematological findings or systemic disease. With a clinical suspision of calcified parasitic cyst the patient was operated upon.

We received a single globular grey white mass that measured 1x0.5x0.5cm. It was firm to hard in consistency. On cut section it was gritty. On microscopic examination a well circumscribed lesion was present with extensive areas of calcification and ossification. It was composed of islands of epithelial cells with varying amounts of basaloid cells along with adjacent anucleated ghost or shadow cells [Table/Fig-1]. Fibrofatty tissue was seen among the seams of bony trabeculae showing erythroid and myeloid precursors along with large cells with multilobulated nuclei, representing megakaryocytes [Table/Fig-2]. On immunohistochemistry these multilobated cells were positive for CD61 and Anti-myeloperoxidase (anti-MPO) was positive in the myeloid precursors [Table/Fig-3,4]. Based on histological immunohistochemical features diagnosis of pilomatricoma with extensive areas of calcification and ossification with evidence of extramedullary haematopoiesis was rendered.



[Table/Fig-1]: Islands of epithelial cells with varying amounts of basaloid cells along with adjacent anucleated ghost or shadow cells. **[Table/Fig-2]:** Fibrofatty tissue was seen among the seams of bony trabeculae showingery throid andmyeloid precursors along with large cells with multilobulated nuclei, representing megakaryocytes.



[Table/Fig-3]: Immunohistochemistry showing multilobated cells positive for CD61 and anti-MPO was positive in the myeloid precursors.
[Table/Fig-4]: Immunohistochemistry showing anti-MPO was positive in the myeloid precursors.

DISCUSSION

Pilomatricoma, also known as "calcifying epithelioma of Malherbe" is a tumour which shows differentiation towards hair cells, particularly hair cortex cells [1]. Four clinical variants have been described. They are: 1) eruptive type; 2) perforating type; 3) familial type (associated with myotonic dystrophy); and 4) recurrent invasive, non-metastatic pilomatrix carcinoma [2].

In a study by M. Valluvan et al., on skin adnexal tumours over a span of 10 years only a single case of pilomatricoma with myeloid metaplasia was reported [2]. Also, in another study by Steven Kaddu et al., seven cases of pilomatricomas with EMH were found in a span of 16 years. Two out of these seven showed myeloid erythroid and megakaryocytes while rest were with myeloid and or erythroid precursor cells [3]. They found that this phenomenon is not uncommon in pilomatricoma with an overall incidence of 5.8%. Ackerman et al. in 1993 studied 120 cases of pilomatricomas and had noted EMH in seven cases [4]. Only two lesions showed osseous metaplasia. The variable incidence of this entity can thus be ascertained from the incidences quoted by different authors.

Cutaneous EMH has rarely been documented. It usually occurs in patients with generalized EMH cases in association with a haematological disorder, especially myelofibrosis [5]. Several authors have proposed that the cause could be due to the compensatory mechanism of the disturbed haematopoietic organ [3]. Studies have shown EMH may occur adjacent to the spicules of bone. It has been shown that the Bone Morphogenetic Protein (BMP)-2 plays an important role in ectopic bone formation [2]. Degenerative changes could also be providing environment for the colonization by the circulating stem cells as have been encountered with pilomatiocomas showing degenerative changes [3]. Degenarating leiyomomas have also been found to show EMH [6].

EMH when seen in a localized form may not be associated with haematological disorders or bone marrow disturbances. They may be seen in areas of degenerative tissue changes, especially ossification and calcification [3].

Extensive search of literature have yielded scant knowledge of this entity with few of the authors quoting variable incidence of EMH in pilomatricoma. The exact cause of EMH is not known in such cases. Our case had no significant haematological or systemic disease and is a case of localized EMH occurring in a benign skin appendageal tumour. The patient however lost to follow up.

CONCLUSION

Pilomatricoma with extensive areas of ossification and presence of extramedullary haematopoiesis is extremely rare. It should be further stressed that extramedullary haematopoiesis in regressive lesions of pilomatricomas is a localised phenomenon and that these findings may not be linked with a systemic haematological disorder.

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