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Case Report

Ossifying pilomatricoma: A rare entity

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ABSTRACT

Pilomatricomas are rare benign skin adnexal tumors usually occurring in the head, neck or upper extremities of children and young adults. Ossifying pilomatricoma is a still rarer entity. Here, a case of a 22-year-old lady is reported, who presented with a firm to hard swelling in the right side of the neck. Histopathological examination of the resected specimen showed features of ossifying pilomatricoma with the formation of bone marrow elements. The patient was free of recurrence on 6-month follow-up. The case highlights a rare entity with characteristic histopathological features.

1. Introduction

Pilomatricoma, formerly known as pilomatrixoma, is a rare benign tumor of the skin appendage which usually presents as a well-defined, slow-growing, asymptomatic subcutaneous swelling of firm to hard consistency, more commonly in the head and neck region in the first two decades of life. Malherbe and Chenantais first described the lesion in 1880 and coined the name calcifying epithelioma of Malherbe as they thought that the calcified tumor arose from the sebaceous gland.[1]

In 1922, Dubreuilh and Cazenave described the characteristic histology comprising of shadow (ghost) cells and basaloid cells[2] but it was in 1942 that Turhan and Krainer postulated that the neoplasm arises from hair matrix cells and due to its histological similarity, it has been considered as a dermal analog of the calcifying odontogenic cyst.[3] However, the term 'Pilomatricoma' was coined by Forbis and Helwig much later in 1961.[4]

Though these usually present as solitary lesions, multiple pilomatricomas have also been described, particularly when associated with syndromes like Gardner Syndrome, Turner Syndrome, Rubinstein-Taybi syndrome etc.[2] Pilomatricoma with florid osseous metaplasia, also known as ossifying pilomatricoma, is a rare and distinct variant of pilomatricoma, the pathogenesis of which is still not well understood.[5]

2. Case Report

A 22-year-old lady presented with a painless swelling on the right side of the neck. The swelling was slowly increasing in size for the last 1 year and now she complained that the swelling was gradually becoming harder. On examination, a 3 cm diameter, firm to hard, well-circumscribed, non-tender, mobile swelling was palpated, adherent to the overlying skin but without any skin changes. Fine needle aspiration done from the lesion yielded paucicellular smears with an occasional cluster of basaloid cells. A possibility of pilomatricoma was made as to the cytological diagnosis. The swelling was resected under local anaesthesia and sent to the histopathology laboratory for examination. Grossly, it was a soft tissue swelling with adherent skin having a gritty sensation on cutting. The microscopic examination showed a well-circumscribed

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tumor composed predominantly of basaloid cells and some shadow cell clusters with prominent laminated bony trabeculae. (Figure 1) Between the bony trabeculae, the mononuclear cells, multinucleated megakaryocytes and fat spaces could be well appreciated consistent with bone marrow material.(Figure 2) A histopathological diagnosis of ossifying pilomatricoma was rendered. The patient showed no recurrence at 3-month and 6-month follow-ups.

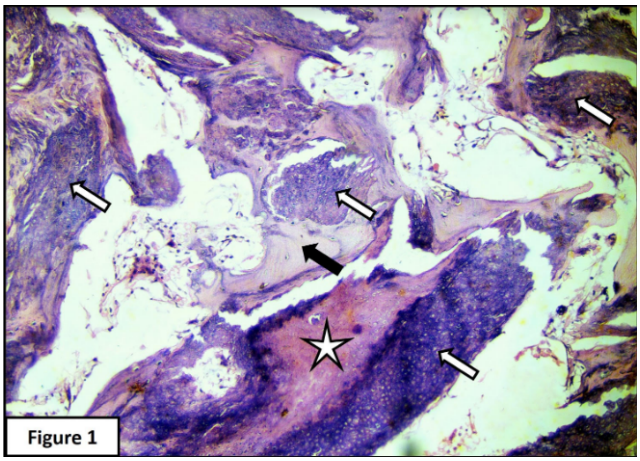

Fig. 1: Photomicrograph showing basaloid cells (white arrow), ghost cells (star) and bony trabeculae (black arrow) (H&E stain; 100X magnification).

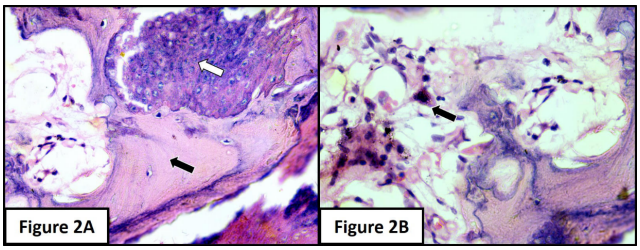

Fig. 2: A: Photomicrograph showing basaloid cells (white arrow) with adjacent bony trabeculae (black arrow); **B:** Hematopoietic cells in the intertrabecular spaces with megakaryocyte marked by black arrow (H&E stain; 200X magnification).

3. Discussion

It is generally believed that the formation of pilomatricoma represents a disturbance in the hair follicle cycle characterized by limited cytologic differentiation of pilar keratinocytes but a lack of further development into mature hair.[2]

Histopathologically, pilomatricoma shows two major cell types viz the basaloid cells which represent the proliferating portion of the tumor, and the mature, larger, pinkish shadow cells. This may be accompanied by foreign body giant cells, calcification, and rarely ossification. Shadow cells are not pathognomonic of pilomatricomas and may be seen in a myriad of conditions like craniopharyngiomas, odontogenic tumors etc.[5]

The pilomatricomas pass through 4 chronological stages viz early, fully developed, early regressive and late regressive stages depending on the duration of the lesion.[6]

Early lesions are small, cystic structures containing squamoid and basaloid cells with the presence of faulty, aberrant hair matrix material in the form of shadow cells. In the fully developed stage, the lesions are larger and are composed of lobules of basaloid cells with the shadow cells at the centre. The early regressive stage shows inflammation with the presence of multinucleated foreign body type giant cells around the shadow cells caused by the release of the hair matrix material into the surrounding tissue. The late regressive phase lacks the epithelial content and the inflammatory reaction. It shows confluent masses of the faulty matrix material with the presence of secondary changes like desmoplasia, dystrophic calcification, metaplastic ossification etc.

Pilomatricomas characterized by florid osseous metaplasia have been termed as ossifying pilomatricomas. They usually have a longer clinical history and firmer consistency than conventional pilomatricomas.[5] Usually in most reported cases of ossifying pilomatricomas, there is minimal or absence of basaloid components with the only presence of shadow cells and bone formation around the shadow cells with some cases showing extra medullary hematopoiesis.[2,5–7]

Osseous metaplasia is a well-described type of aberrant ossification encountered in various neoplastic and non-neoplastic conditions ranging from epidermal cyst, trichilemmal cyst to chondroid syringoma and craniopharyngioma.[8–11]

Ossification in pilomatricoma is generally seen next to the areas of shadow cells and is thought to be a sequelae of the longstanding fibroblastic response. This fibroblastic reaction follows the foreign body reaction due to release of the faulty hair matrix material into the surrounding tissue. Thus, ossification is seen only in the old pilomatricomas. However, in the present case, there was the presence of both basaloid cells and shadow cells with trabecular bone and hematopoietic elements.

The metaplasia of fibroblasts into osteoblasts may occur by the action of bone morphogenic protein 2 (BMP-2).[12]The other probable candidate for osteoblastic differentiation of precursor cells is oncostatin M produced by activated macrophages.[13]

Macrophage-derived osteopontin is thought by some to be responsible for deposition of calcium phosphate in the shadow cell nests.[14]

Hence, in spite of several theories, the exact mechanism of ossification in pilomatricoma is still under speculation.

The standard treatment of ossifying pilomatricoma is surgical resection and the rate of recurrence is low.[15]

4. Conclusion

Ossifying pilomatricoma with bone marrow elements is a rare variant of pilomatricoma which is seen in longstanding cases particularly in the late regressive stage of the tumor. However, ossification may be present in fully developed or early regressive stage of the disease as seen in the present case. The origin of the ossification is either metaplastic or heterotopic. Surgical resection is usually curative.

5. Conflict of Interest

The authors declare no relevant conflicts of interest.

6. Source of Funding

None.

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