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Case Report

Pilomatricoma (Pilomatrixoma) with osteoid metaplasia in an elderly male patient

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Abstract

Pilomatricoma is a benign, nodular, solitary, painless, tumour, extending from lower dermis to subcutaneous fatty tissue. Present tumour developed as a small nodule in the skin of left upper arm. It measured 1.5×1×1cm. It was bony hard tissue covered with skin. Clinically, the patient may present as nodular pricking pain. Sections were taken both from bony and soft tissues. Later, nodule was excised. Microscopically, the tumour showed sheets of large number of tumour cells with basophilic cytoplasm and anucleate 'ghost' or 'shadow' cells. Bony trabeculae with osteoblastic rimming were seen at few places. Adjacent stroma showed fibrosis. Few foci of calcification were seen in sheets of tumour cells. Small number of inflammatory cells and occasional giant cells were also seen. Lesion was finally diagnosed as pilomatricoma with osteoid metaplasia.

Keywords: Anucleate ghost cells, Calcification osteoid tissue, Benign neoplasm.

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1. Introduction

Pilomatricoma is a rare benign neoplasm in Asians arising from hair follicle.[1] It is appendageal tumour. Onset of the tumour is fast and it expands rapidly.[2] It was first reported in the year 1880.[2] Tumour is more common in females; male / female ratio being 2:3.[3] Tumour may be well-circumscribed. Cut surface of nodule may show chalk-like powdery substance which may be calcium deposit.[3] Consistency of pilomatricoma may vary according to the degree of calcification.[4] Microscopically, it shows large number of basophilic cells which may abruptly undergo keratinization without forming stratum granulosum. Later, it leads to formation of anucleate ghost cells.[4] Pigmentation may also occur due to migration of melanocytes. Rarely, extramedullary hematopoiesis may develop in the tumour. Foreign body giant cell infiltration may also occur. In addition, ossification may develop. Moreover, epithelial perforation may occur.[5] Mutation of the β-catenin gene (CTNNβ1) may occur[6] Moreover, trisomy of chromosome 18 may be associated with pilomatricoma. In most of the cases, it results in loss of regulation of β-catenin /LEF protein complex. Pilomatricoma is believed to be due to this mutation. CTNNβ1 protein is involved in cell adhesion and communication between the cells. However, it is not clear why a few pilomatricomas are malignant while most of others are benign. Several stages in development of pilomatricoma have been described. First, small cysts may develop which may be lined by basal or squamous cells. Second, cysts may enlarge further and may be lined by basophilic cells in association with ghost cells. Third, early regressive changes may occur. Fourth, advanced regressive changes may develop without epithelial element.[7] The current tumour may be etiologically related with insect-bite, trauma and surgery.[8] Familial tumour may be associated with several syndromes, e.g. Turner syndrome, basal cell nevus syndrome and myotonic dystrophy. Multiple lesions may develop in familial tumour. However, familial tumours rarely occur.[8]

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2. Case Presentation

Pilomatricoma is a benign slow-growing asymptomatic tumour. The tumour may be < 3 cm in diameter. Rarely, a giant pilomatricoma may develop size ~6x4 cms.[9] In addition, the present tumour has very low incidence. Present tumour had large number of basophilic cells with round nuclei. Tumour was encapsulated with compressed fibrous tissue. Foci of metastatic calcification were seen. Osteoid tissue formation in a patient with pilomatricoma is extremely rare histopathological feature. Microscopically, sections showed tumour tissue, comprising of proliferated basophilic cells admixed with anucleate ghost cells. Extensive keratinization was also seen. Ghost cells develop from basophilic cells. These are dead cells with definite shape and central unstained anucleate area. In the present case, bony trabeculae with osteoblastic rimming were seen. Adjacent stroma showed fibrosis. In addition, focal collections of lymphocytes and macrophages were also seen. Pilomatricomas mostly develop in children and young adults. However, present tumour developed in an elderly patient. Immunohistochemistry examination was done using monoclonal murine anti-human CD45, a leucocyte common antigen (LCA) from clone 2B11+PD7/26. Antibody was purchased from DAKO. Positive staining for Pankeratin was also done. Keratins (Pankeratins, 2-Keratins 5/6 and hard α-Keratins-HHa1, HHa2 and HH5 are known to be positive for pilomatricoma not in follicular neoplasm. In the current report, tissue sections were stained by hematoxylin Eosin and Masson trichrome methods. Basaloid cells of pilomatricoma show strong nuclear and weak cytoplasmic reactions.(**Figure 1**)

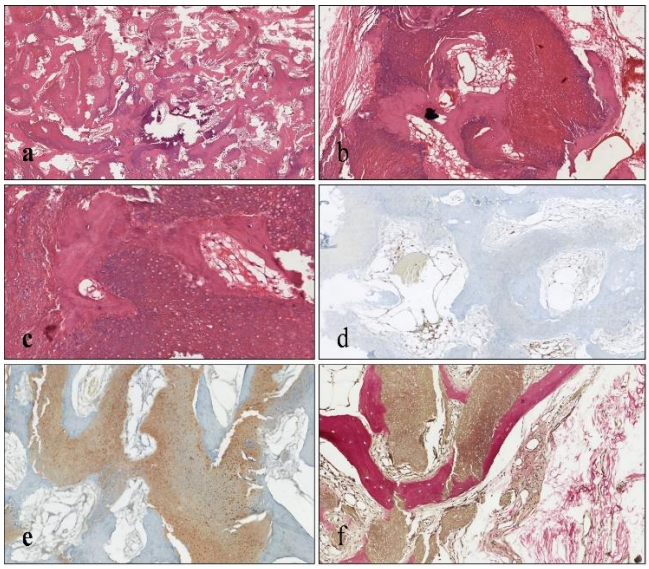

Figure 1: (**a**): Photomicrograph shows tumour cells with an area of necrosis with calcification (HE×100). (**b**): Tumour comprised of basophilic cells (HE×50). (**c**): Photomicrograph shows tumour tissue. Tumour cells were basaloid with round nuclei. Tumour cells were admixed with ghost cells (HE × 100). (**d**): Photomicrograph shows osseous metaplasia. Area between bony trabeculae shows infiltration by lymphoid cells (Masson's trichrome×100). (**e**): Shows osseous metaplasia of tumor cells (Van Gieson × 100). (**f**): Photomicrograph shows lamellar bony trabeculae with entrapped osteocytes. In addition, interstitium showed diffuse mild lymphocytic infiltration. Anti- common leucocyte antigen (CLA) antibody stained lymphoid cells. Small number of leucocytes were detected (× 100).

3. Discussion

Pilomatricoma is a benign tumour.[1] Forbis and Helwig suggested that the tumour may arise from the root of outer sheath cell of hair follicle.[10] Its incidence is very low. It represents ~1% of benign cutaneous neoplasms.[11] Tumour consists of islands of epithelial cells. Ackerman et al described pilomatricoma as a sac of epithelium that is infundibular above and metrical cells below and laterally.[12] Furthermore, other benign subcutaneous lesions, e.g. ossifying fibroma, ossifying hematoma and fibroxanthoma may be differentiated from pilomatricoma.[13] In addition, calcium deposits may occur in > 50% of the lesions. The tumour occurs mainly in the skin of head, neck and extremities.[14] Calcification, metaplastic bone formation, brisk mitoses and basophilic cells are frequently seen. However, anisonucleosis should not be seen.[14] Most of the present tumour comprised of keratin and basophilic cells.

The term pilomatricoma was used for the first time in the year 1969.[15] Furthermore, tumour may exhibit pearly-white appearance.[15] Histopathologic features of pilomatricoma were described earlier in the year 1922.[16] The cytoplasm of transitional cells is filled with large number of α-keratin filaments around nuclei. In the shadow cells, the strands of fibrils fuse around empty calcified areas. It was suggested that the tumour was derived from primitive basophilic cells of epidermis that may differentiate later into hair matrix cells.[17] Ghost cells may also be found in epidermoid cyst, chronic dermatitis and hyperkeratosis.[8] Moreover, calcification, ossification and giant cells may be seen ~ in 87%, 8% and 70% of tumour cells respectively. Foreign body giant cell formation is a common features of pilomatricoma.

Present tumour had all the features of pilomatricoma, e.g. large number of anucleate ghost cells were seen. Occasionally, basophilic epithelial cells admixed with ghost cells were seen. Another important feature of current tumour was focal inflammatory cell infiltration consisting of lymphoid cells. Rims of bony trabeculae are commonly lined by foreign body giant cells. Rarely, foreign body giant cells have been reported.[2,3,8] However, in the current case, few giant cells were seen. Furthermore, two other patients showed foci of suprabasal cleft formation.[8] Another case showed Borst-Jadassohn phenomenon (BJP). BJP is a well demarcated tumour with nests of uniform squamous cells with or without cytologic atypia in the spiny acanthotic epidermis.[18] Moreover, osteoid tissue formation was also seen in the present case; both woven and lamellar bone formations were seen. Osteoid tissue formation is extremely

rare feature of pilomatricoma. However, more commonly, osteoid formation may be found in various subtypes of pilomatricoma, e.g. pigmented, proliferating, nodulocystic, perforating bullous variants. Moreover, it may be associated with melanocytic matrixoma.[19] In addition, metaplastic bone formation has been reported in extramedullary hematopoiesis in pilomatricoma.[13] Sebaceous differentiation and acantholysis of ghost cells may be additional features of pilomatricoma. Pilomatricoma may also be detected by mammography as microcalcifications. Occurrence of multiple pilomatricomas is very rare. About 24% of cases may present with multiple lesions.[20,21] Differential diagnosis may be epidermal cysts of Gardner syndrome and metaplastic basal cell carcinoma with chondroid, osteoid and smooth muscle differentiation.[22] Recurrence following resection of tumour may occur in 2% to 6% of cases.[11] Furthermore, warning signs for malignancy may be older age, male gender, rapid growth of lesion, adherence to surrounding organs, high mitotic activity, necrosis and lymphovascular changes.[23] The skin over the tumour may be included during excision of the tumour because the tumour may adhere to dermis.[21] Moreover, 3 of 21 cases had multiple lesions and 4 cases had ossifying pilomatricoma.[24] Furthermore, acantholysis is also frequently seen in the tumour cells.[8]18 However, it was not seen in the present tumour. Pilomatricoma cases with local invasion or local recurrence have been described as aggressive pilomatricoma.[5] Malignant transformation is very rare and ~20 cases have been described in literature.[10] Recently, a case of pilomatrix carcinoma with visceral metastasis has been reported.[25] Earlier, another patient with metastasis in bone was reported. Rarely, a case of basal cell carcinoma with expression of matrix, catenin and osteopontin has been reported. Recently another case of benign pilometricoma with osseous metaplasia has been reported. Treatment of the patient comprised of surgical excision and curettement. The patient was followed for 6 months. No recurrence has occurred.

4. Conclusion

Pilomatricoma is a benign neoplasm. Rarely, a malignant change may occur. Tumour mainly develops in skin of face, neck and upper and lower limbs. Surgical excision may result in its cure. Tumour may be resected with 1 to 2 cm margins to achieve cure. Rarely, late recurrence has been reported.

5. Source of Funding

None.

6. Conflict of Interest

None.

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