

Pilomatrix Carcinoma At The Popliteal Fossa: A Case Report With A Review Of The Literature

D Kotasthane, V Kotasthane, G Koteeswaran

Citation

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Abstract

Pilomatrix carcinoma is a rare, low-grade malignant tumour of hair matrix. In this report, we present a case of Pilomatrix carcinoma in a 52 year male patient, which occurred on a popliteal fossa, which is a very rare site. Histological findings have been discussed.

INTRODUCTION

Pilomatrix carcinoma is a malignant counterpart of pilomatrixoma. It is so rare a disease that only 80 cases have been reported (1). Gromiko observed pilomatrix carcinoma invading adjacent tissues and recurring for the first time in 1927. (2) Lopansri and Mihm reported a case of pilomatrixoma with malignant features and proposed the term "pilomatrix carcinoma" or "calcifying epitheliocarcinoma of Malherbe." (3) This neoplasm can exhibit local aggressive behaviour and occasionally distant metastasis. Most pilomatrix carcinomas occur on the head and neck. (4) Few cases have been reported at other sites like axilla and spine. (5,6) To the best of our knowledge, this rare tumour has not been reported in popliteal fossa.

CASE REPORT

A 52 year-old male patient came to General Surgery Out Patient Department with a slowly growing fungating growth over the popliteal fossa. The patient had this growth for a period of 2 years. Clinical diagnosis was Squamous cell carcinoma. Wide local excision was done and sent for histopathological examination. The specimen received showed a fungating, proliferative ulcerated mass with surrounding skin. The mass measured 12x11x10 cm. External surface of the mass was necrotic, friable and showed sand grain appearance. (fig 1) Cut section was grayish white with cystic change and areas of necrosis (fig 2) Haematoxylin and Eosin sections studied showed a tumor covered by ulcerated epidermis. The tumor showed nests of basophilic cells with deeply basophilic nuclei and scanty cytoplasm and indistinct cell margins. The nests of tumor cells showed eosinophilic areas of abrupt keratinisation (fig

3) These cells were seen surrounding eosinophilic shadow or ghost cells which show loss of nuclei. (fig 4) The basaloid cells showed pleomorphism, darkly stained nuclei, anisonucleosis and mitotic figures. (fig 5 and 6) Focal areas of necrosis were also seen. (fig 7) Margins of tumour mass were seen infiltrating surrounding stroma. (fig 8) The tumor cells showed areas of perineural invasion. (fig 9) The stroma of the tumor showed foreign body giant cells and lymphocytic reaction. Based on these findings, histopathological diagnosis of pilomatrix carcinoma was given.

Figure 1

Figure 1: External surface of the growth showing fungating ulcerated exophytic growth with sand grain appearance (arrow showing surface ulceration)



Figure 2

Figure 2: Cut surface of the growth showing areas of cystic change (shown by arrow)



Figure 3

Fig 3: showing nest of basophilic cells with abrupt keratinisation

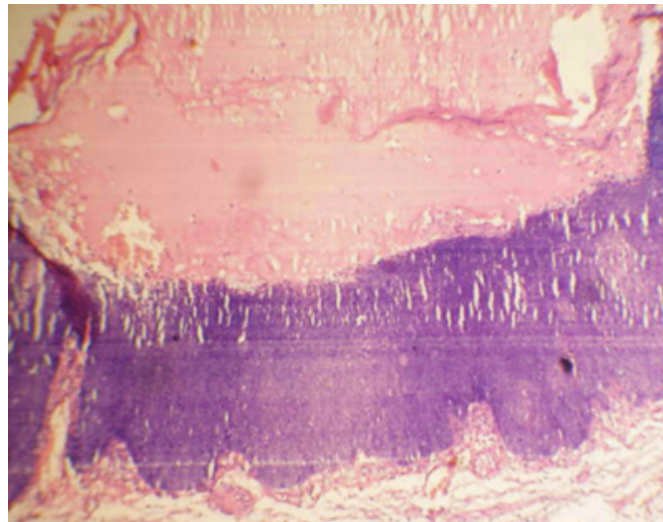


Figure 4

Fig 4: showing ghost cells shown by arrows

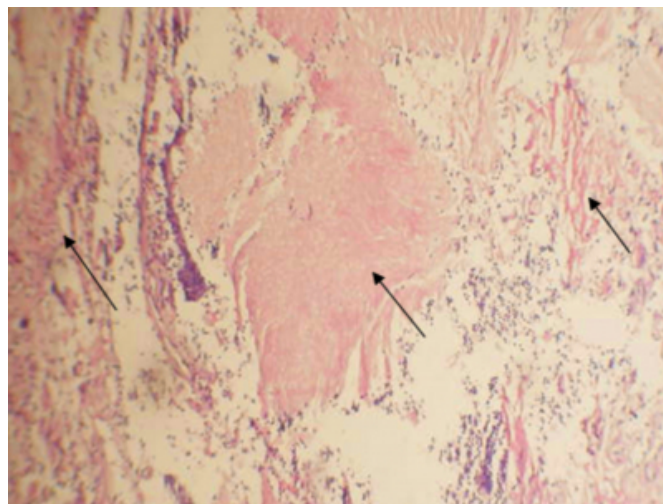


Figure 5

Fig 5: Showing basaloid cells with darkly staining nuclei. Arrow showing mitotic figures

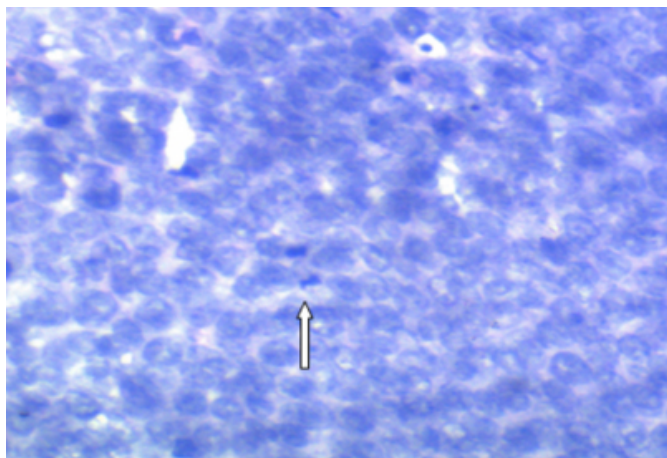


Figure 7

Fig 7: Arrow showing necrosis.

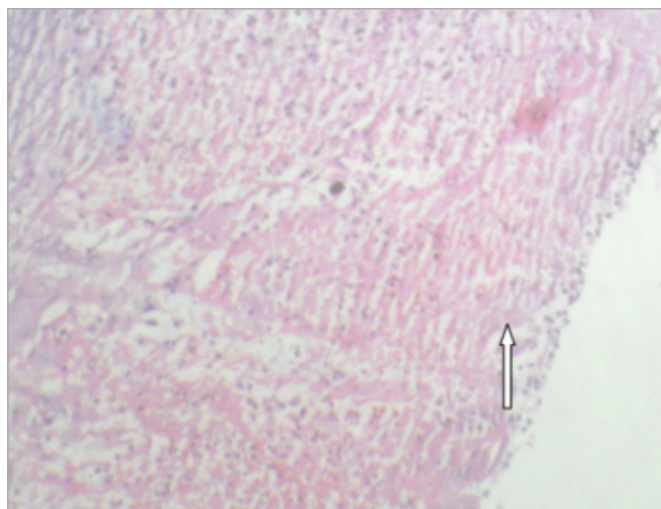


Figure 6

Fig 6: Basaloid cells show pleomorphism and anisonucleosis.

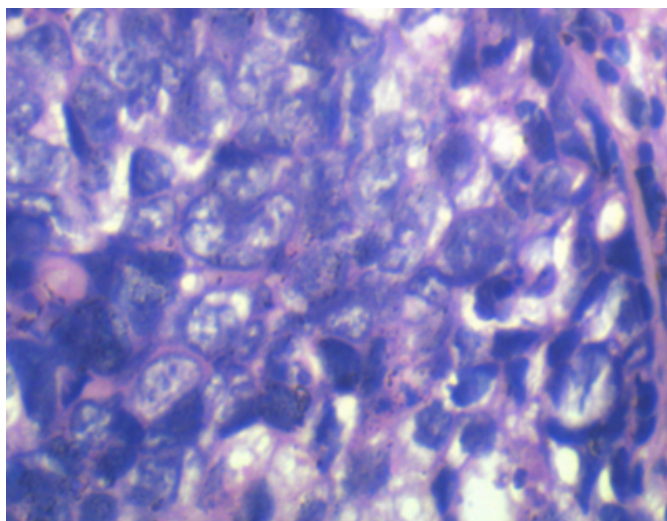


Figure 8

Fig8: Nests of basophilic cells with margins infiltrating the stroma, as shown by white arrows.

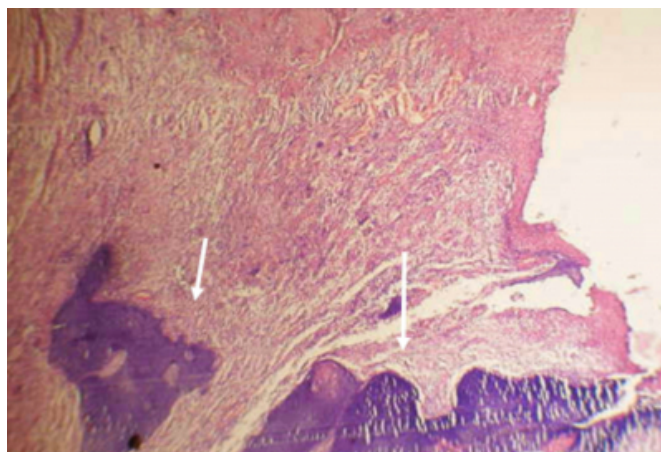
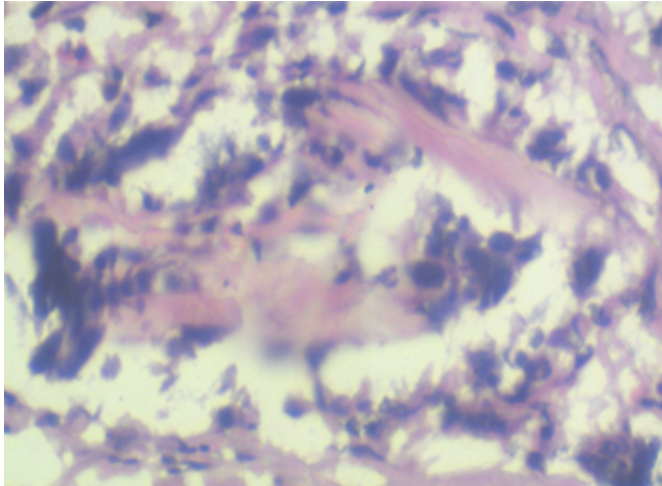


Figure 9

Fig 9: Cells showing perineural invasion.



DISCUSSION

In case of the benign counterpart, i.e, pilomatrixoma, the male to female ratio is 2:3, with about 60% cases occurring under age of 20 years. (7) In contrast to this, pilomatrix carcinoma, the ratio of male: female occurrence is 3: 1. Review of literature reveals a broad age range with a mean age of 46 years in case of pilomatrix carcinoma. (7, 8) Our patient was a male patient of 52 years.

The most common site of pilomatrix carcinoma is the head and neck, occurring in 60% of patients, and followed by upper extremities, trunk, lower extremities. (8) The tumor was present on popliteal fossa which is an uncommon site. On extensive review of literature, it was found that pilomatrix carcinoma has not been reported in popliteal fossa.

The clinical appearance of pilomatrix carcinoma is generally not distinctive. Patients show solitary, occasionally ulcerated or fungating nodules ranging in size from 1-10 cm in diameter. (9) The mean size of pilomatrix carcinoma as found in literature is variable. The mean size was 4.8 cm in the case study of Sau P (10), 1.78 cm in the case study of Hardison D (11) and 3.5 cm in the case study of Li X. (12) In Our patient, the size of the tumor was 12x10x11 cm, which is larger as compared to that described in literature. Skin nodules are often of long duration ranging from several months to years before diagnosis, although occasional cases of recent onset and a history of rapid growth have been reported (11). In our patient, the tumor was slowly growing for a period of 2 years to attain the present size. The clinical features are similar in benign pilomatrixoma and malignant

pilomatrixoma, except for the course and development of the disease

Features which are helpful in making the diagnosis include asymmetry and poor circumscription, presence of several markedly sized and variably shaped basaloid aggregations of tumor cells, (12,14,15) Basaloid cells exhibit hyperchromatic nuclei, with one or more prominent nucleoli and ill-defined cytoplasmic margins as well as variable numbers of occasionally atypical mitotic figures (up to 10 mitoses per high-power field). Foci of geographical necrosis, calcification and ossification are observed. Mitotic activity is not a reliable indicator of malignancy, because mitoses are common in pilomatrixoma. Other parameters, such as an infiltrative growth pattern, as well as angiolymphatic, perineural and bone invasion are more reliable features. (15,16) In our case, the basaloid cells had deeply basophilic nuclei and scanty cytoplasm. Few Mitotic figures were seen. Foci of necrosis were also seen. The tumor margins were seen infiltrating the surrounding stroma, at places. Some neoplasms show a variable desmoplastic stroma surrounding the basaloid cell aggregations. Focal connections of basaloid cell aggregations to the overlying epidermis and/or ulceration are often noted. In our case, the epidermis showed ulceration. The histological examination can be challenging, especially because there are no clear histologic criteria distinguishing this neoplasm from other matriceal tumours. Immunohistochemical investigations have failed to delineate a specific marker for this tumour. (17) Differential diagnosis includes Basal cell carcinoma with matriceal differentiation, in which retraction spaces between neoplastic cells and stroma is seen. (18) Trichoblastic carcinoma is another malignant tumor of hair follicle. It is made up of large cells with abundant cytoplasm and lack the typical shadow cells. There is no peripheral palisading of neoplastic cells and no cleft between the tumor lobule and the surrounding stroma. (19)

Some pilomatrixomas show apparent transformation into carcinomas. (11,12) Other cases are malignant from the onset. (20,21) Pulmonary, lymphnode and bone metastases may occur. The lesions have a tendency to recur if not widely excised. (22,23,24,25) Gould et al. reported the first case of distant metastasis to both lungs, which occurred 4 years after operative excision. Metastases were identified from 4 months to 4 years after the first diagnosis. (26) Mean life expectancy was reported to be from 3 months to 2 years. If wide local invasion or metastasis is identified, surgical excision and postoperative radiation therapy or

chemotherapy can be done, but there are no standard treatments known to produce good results. (27) Our case did not show metastasis, when it was diagnosed. Wide local excision was done in our case and close clinical follow up was advised.

CONCLUSION

Pilomatrix carcinoma is a very rare low grade malignant tumor. To the best of our knowledge, no case has been reported in Popliteal fossa. Gross appearance mimics Squamous cell carcinoma. Pilomatrixoma, Basal cell carcinoma with Pilar differentiation and Trichoblastic carcinoma should be considered as a differential diagnosis. Wide local excision is the treatment of choice.

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Author Information

Dhananjay Shrikant Kotasthane

Professor and HOD, Department of Pathology, Mahatma Gandhi Medical College and Research Institute

Vaishali Dhananjay Kotasthane

PG student, Department of Pathology, Mahatma Gandhi Medical College and Research Institute

G Koteeswaran

Associate Professor, Department of Pathology, Mahatma Gandhi Medical College and Research Institute