A rapidly growing tumour over arm in a middle-aged man

A 43-year-old man presented four years ago with a single asymptomatic tumour over the right lower arm of one-month duration. In the past, he had developed similar lesions over the right side of the chest, right forearm and another over the lower left arm. On examination, a single, firm, mobile, erythematous, smooth, non-tender swelling of size 4.5× 4.8 cm was present over the flexor aspect of lower right arm [Figure 1]. One and a half years later, the patient developed another similar lesion over the left upper arm.

The excisional biopsy for the lesion over the right arm revealed a relatively well-circumscribed tumour located in the dermis. The tumour consisted of a lobular proliferation of basaloid cells with areas of abrupt keratinisation. The basaloid cells were interspersed with multinucleate giant cells and foci of dystrophic calcification. Basaloid cell areas showed high cellularity, nuclear overcrowding and overlapping with minimal nuclear pleomorphism and frequent mitosis ranging from two to four per high-power field with some tiny areas of necrosis [Figures 2a-2d]. Immunohistochemistry revealed a tumour proliferating index (Ki 67) of 30–40% at the highest proliferating area [Figure 2e]. Definitive evidence of adjacent tissue infiltration was not noted even after multiple sections. Peripheral and deeper resected margins were free from the tumour.

Question

What is your diagnosis?

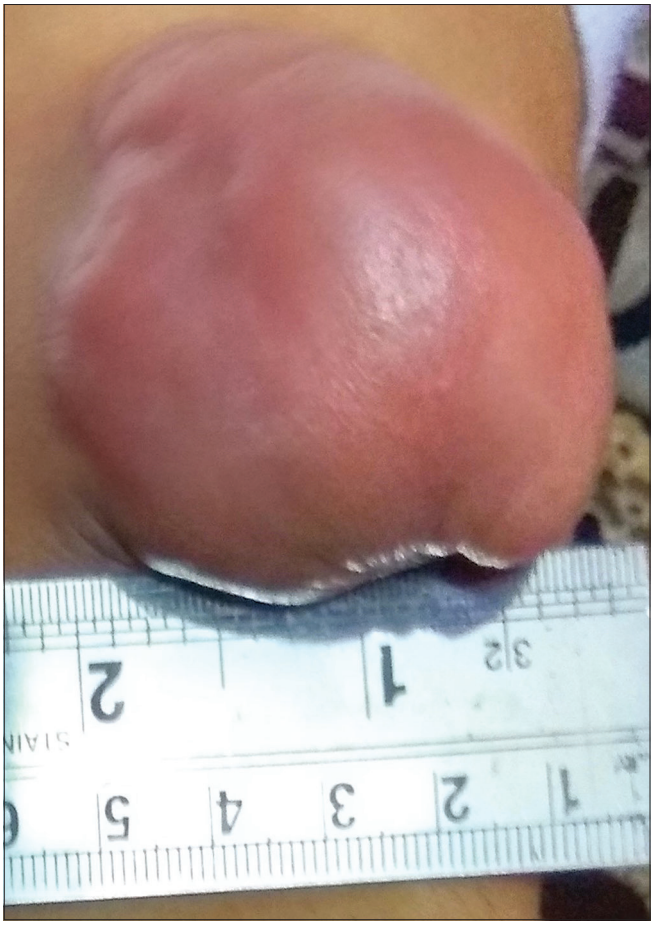

Figure 1: A firm, mobile, erythematous, non-tender, smooth tumour of size 4.5 × 4.8 cm, over the flexor aspect of lower right arm

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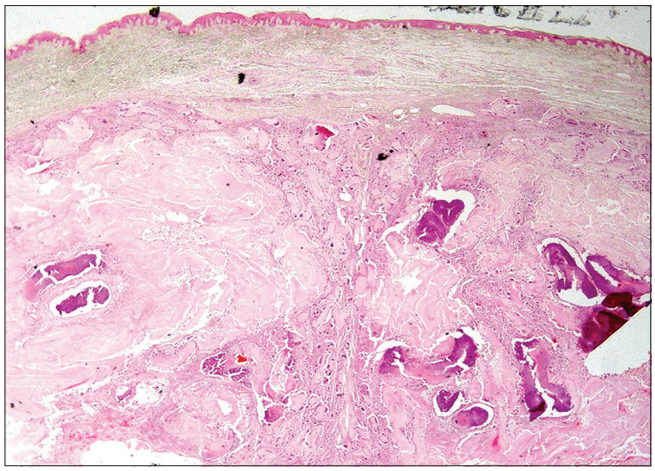

Figure 2a: Microscopic sections from the well-demarcated tumour show: Dermis showing relatively circumscribed tumour with lobular proliferation of basaloid cells (H&E, ×10)

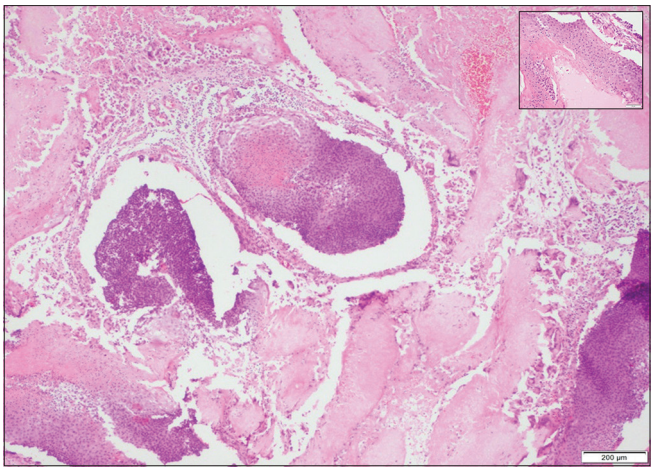

Figure 2b: Microscopic sections from the well-demarcated tumour show: Shadow cells and lobular proliferation of basaloid cells (Inset showing eosinophilic cornified material along with shadow cell [×40]) (H&E, ×200)

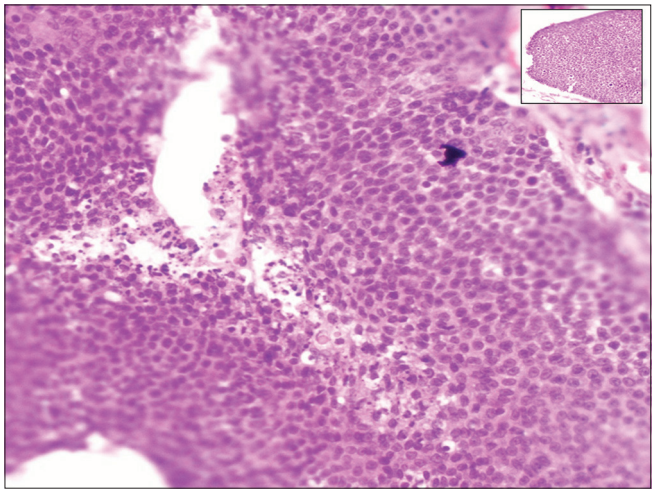

Figure 2c: Microscopic sections from the well-demarcated tumour show: Basaloid cells revealing high cellularity with nuclear crowding and overlapping (Inset showing frequent mitosis [×40]) (H&E, ×400)

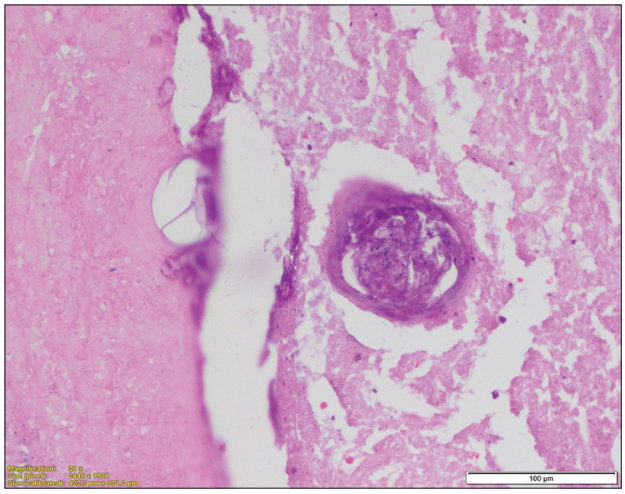

Figure 2d: Microscopic sections from the well-demarcated tumour show: Focus of calcification seen (H&E, ×200)

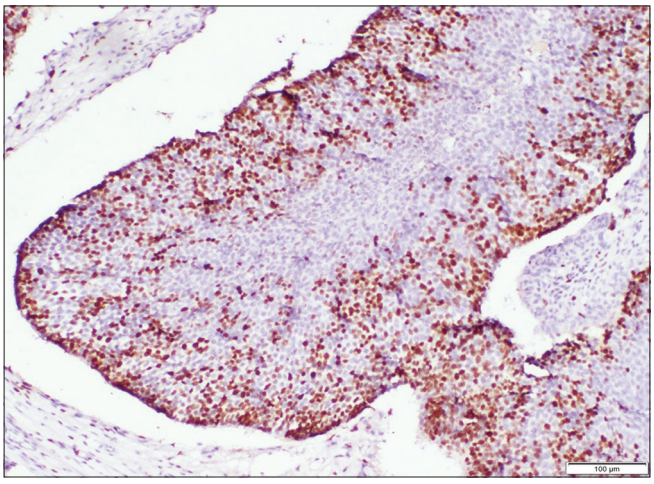

Figure 2e: Highly proliferating areas where proliferating cells are seen almost occupying the entire thickness (Ki 67, ×100)

Answer

Proliferating pilomatricoma.

Discussion

Pilomatricoma, also known as calcifying epithelioma of Malherbe, is a benign adnexal tumour characterised by hair matricial differentiation. It is frequently seen during the first two decades of life.[1] Proliferating pilomatricoma is a rare variant of pilomatricoma, first described in 1997.[1] Till now, only a few cases have been reported in the literature.[2] We report a case of proliferating pilomatricoma with a history of multiple pilomatricomas.

Proliferating pilomatricomas usually arise in the fourth to eighth decades of life;[2] although the youngest reported patient was nine years of age.[3] Kaddu *et al*. described proliferating pilomatricomas being more common in the older age group, larger in size, demonstrating matricial cells more than shadow cells, showing atypia, necrosis and a greater number of mitoses (mean value of six mitoses/high-power field was reported) on histopathology compared to ordinary pilomatricomas.[1] Our patient showed clinical as well as histopathological features consistent with proliferating pilomatricoma. Excisional biopsy of all the other lesions, however, revealed features of pilomatricoma.

The closest histopathological differential diagnosis of proliferating pilomatricoma is pilomatrix carcinoma. Pilomatrix carcinoma is an exceedingly rare variable grade malignancy with an increased chance of recurrence and a high risk of metastasis to locoregional lymph nodes, lungs or bone in 10–16% of cases. The tumour has a propensity for the head-and-neck area. It is fatal in 7–9% of patients. On histopathology, there is a lack of circumscription and presence of sheets of basophilic cells with only a few foci of shadow cells. There are numerous mitoses, areas of necrosis and atypia. Ulceration of the epidermis may be seen with infiltrative borders up to the subcutis or till muscle layer. Invasion of blood vessels or nerves can be seen.[4,5]

There are occasional reports of local recurrence with proliferating pilomatricoma.[1,4] The individual cases described by Kaddu *et al*. and Collina *et al*. had three and two recurrences, respectively.[1,4] Collina *et al.*, thus, proposed the term 'pilomatrical tumour of low malignant potential' for this entity due to its potential aggressive behaviour.[4] However, another reported case had no recurrence even at three-year follow-up.[6] Our patient showed no recurrence, on a long-term follow-up of four years.

In summary, proliferating pilomatricoma is an intermediate group between benign pilomatricoma and malignant pilomatrix carcinoma. Complete surgical excision with adequate margins along with a close and regular follow-up is recommended as treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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