Giant Trichoblastoma of Thigh Mimicking Dermatofibrosarcoma: An Extremely Rare Entity with Unique Histology

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Abstract

Trichoblastoma is a rare benign adnexal tumor exhibiting follicular differentiation and is widely familiar for its resemblances with basal cell carcinoma. We report a case of giant trichoblastoma manifesting as a huge cutaneous nodule misdiagnosed as dermatofibrosarcoma on clinical and radiological grounds. The diverse histological features responsible for its uniqueness are discussed along with a brief review of histogenesis and histological subtypes.

Keywords: Adnexal tumor, basaloid, differentiation, nodule, trichoblastoma

Introduction

Trichoblastomas are pure benign epithelial tumors of hair germ with a differentiation directed toward the hair bulb portion of hair follicle.[1,2] Clinically, they manifest as well-circumscribed small dermal or hypodermal nodules in the region of head and neck, frequently misdiagnosed as basal cell carcinomas.[3] Being benign tumors, complete surgical excision usually suffices but local aggressiveness has been documented along with malignant transformation.[4] Occasional giant trichoblastomas have been described in the literature which some authors consider as large nodular type trichoblastomas, but their numbers are minimal.[5] The exact biologic behavior of these tumors cannot be ascertained in view of relative rarity of these tumors.[4]

Case Report

A 50-year-old male presented to the surgical outpatient department of our hospital with a chief complaint of painless nodular swelling over the postero-lateral aspect of right thigh of 1-year duration. Clinical examination revealed a well-defined nodule of size 7 × 6 cm with superficial ulceration [Figure 1]. The nodule was freely mobile, non-tender, and firm-to-hard in consistency. There was no regional lymphadenopathy. With a provisional clinical diagnosis of dermatofibrosarcoma, the patient was referred to the Department of Radiology for further evaluation. USG showed a hyperechoic lesion of size 6 × 6 × 3 cm in the subcutaneous plane with compression of the underlying muscle. A radiologic diagnosis of benign soft tissue tumor was offered. Wide local excision of the nodule under spinal anesthesia was planned. Intra-operatively, the nodule was found localized to the dermis. There was no evidence of infiltration of the nodule into the underlying subcutaneous tissue. No undue bleeding was noted. The nodule was excised *in toto* with margins of more than 1 cm and subsequently sent for histopathological examination.

Gross examination showed a well-oriented, elliptical wide excision specimen measuring 12 × 7 × 3.5 cm with a central raised nodule measuring 6 × 6 cm. Surface of

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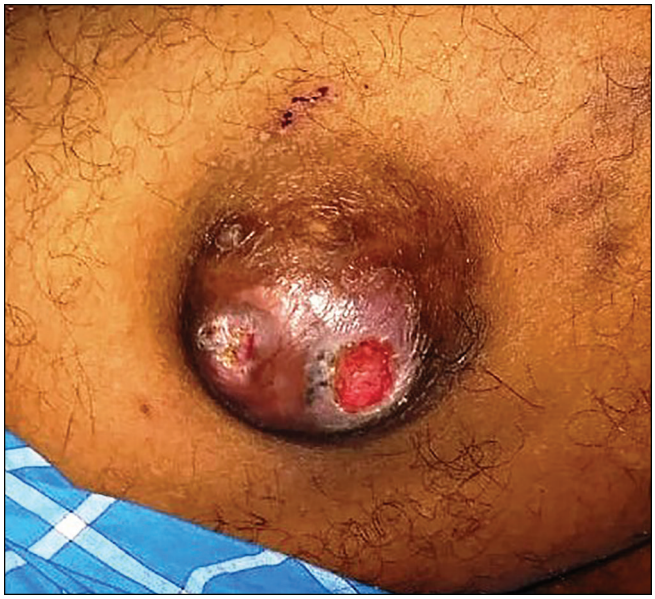

Figure 1: Nodule with superficial ulceration

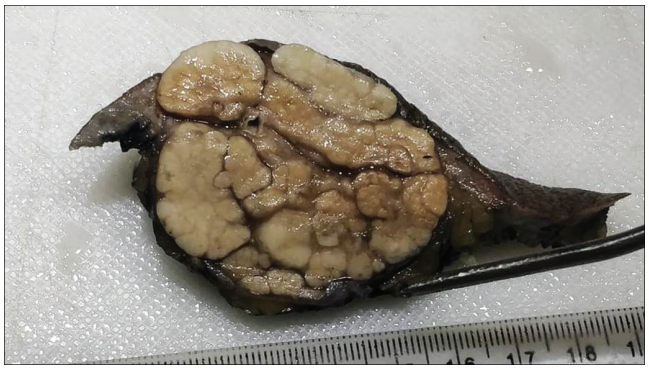

Figure 2: Lobulated gray white tumor with pushing margins

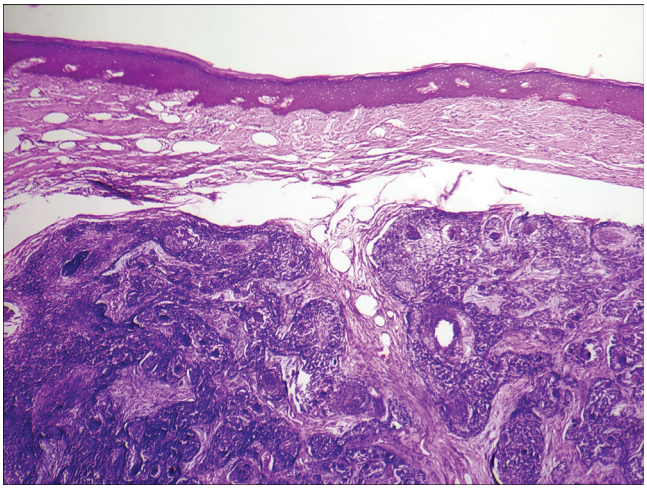

Figure 3: Skin with underlying tumor composed of nodules of basaloid cells, H & E, x40

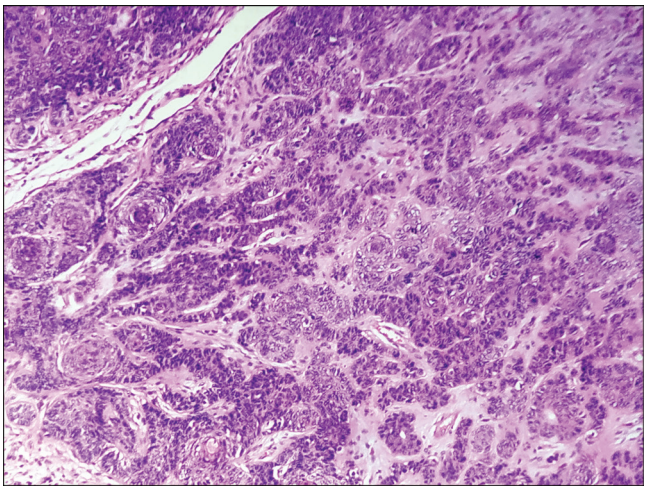

Figure 4: Tumor cells arranged in reticulate pattern, H & E, x100

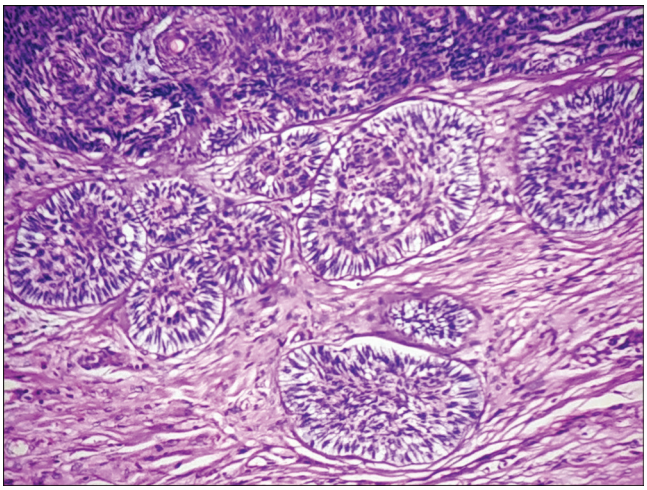

Figure 5: Tumor cells showing peripheral palisading, H & E, x100

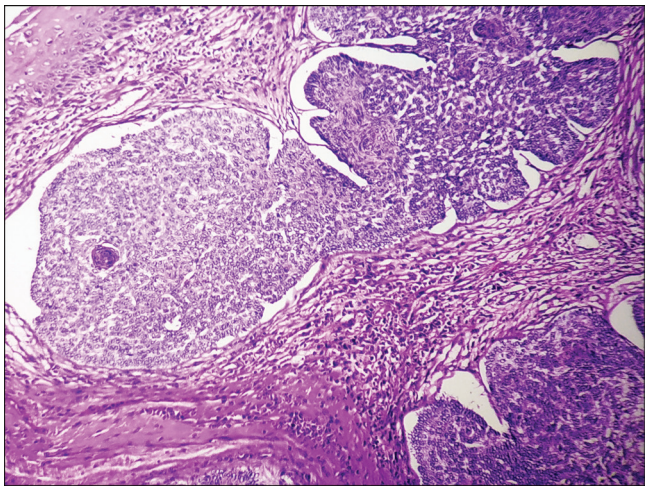

Figure 6: Tumor nodules with retraction artifact, H & E, x100

the nodule showed three ulcers. The cut surface showed a well-circumscribed, lobulated, and solid gray white tumor with pushing margins [Figure 2]. The deep resected surgical margin was free from tumor involvement. Microscopy showed a tumor seated over the deep dermis exhibiting a nodular configuration with intervening fibro-myxoid

stroma. The tumor component revealed predominantly small basaloid cells with evidence of peripheral palisading and focal cleft artifacting, cords of larger tumor cells demonstrating reticulated pattern, papillary mesenchymal bodies, squamous eddies, and extensive areas of calcification. Focal areas showed marked cytologic atypia, abnormal mitoses, and melanin pigment [Figures 3–9]. However, there was no involvement of epidermis by the tumor. With extensive tumor sampling and thorough literature review, a diagnosis of trichoblastoma was considered.

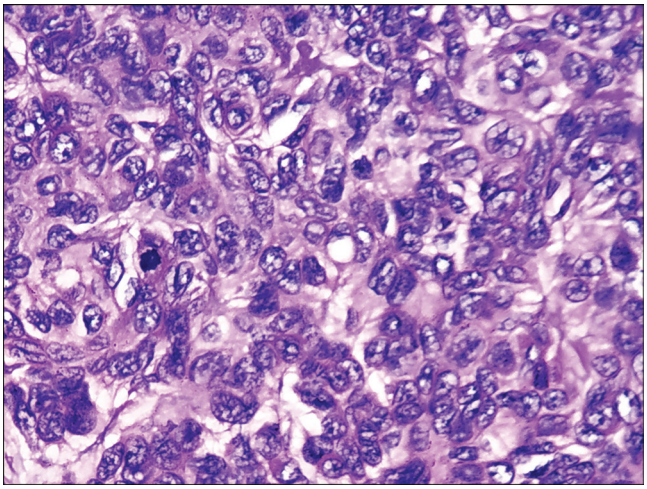

Figure 7: Marked nuclear atypia with mitosis, H & E, x400

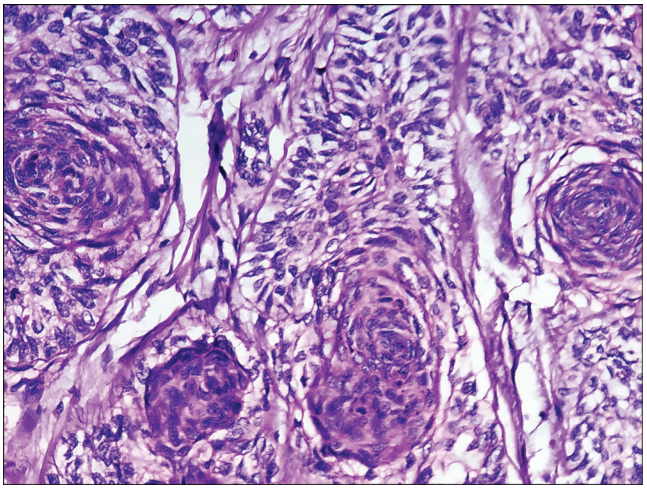

Figure 8: Squamous eddies, H & E, x400

Discussion

The oldest known description related to hair follicle tumors dates back to 1962 where Headington and French initially described a tumor in a young 25-year-old Caucasian female. "Primary neoplasm of hair matrix" was the preferred umbrella term used to describe these tumors which showed evidence of distinct morphodifferentiation toward hair follicles and they coined the term "trichogenic adnexal tumor" to describe this entity. Later in 1970, they used the term trichoblastoma to describe these tumors. Based on the proportion of epithelial and mesenchymal elements in the tumor, Headington divided trichoblastomas into several subtypes. However, Ackerman preferred a single designation.[4]

Trichoblastomas are rare, slow-growing benign tumors with differentiation toward primitive hair follicles. Most often they manifest between fifth and seventh decades of life. However, any age group is afflicted except young children.[6] Clinically, they present as asymptomatic, solitary, well-circumscribed skin-colored to brown or blue-black nodules localized to head and neck with a prediliction for scalp. Involvement of trunk, proximal

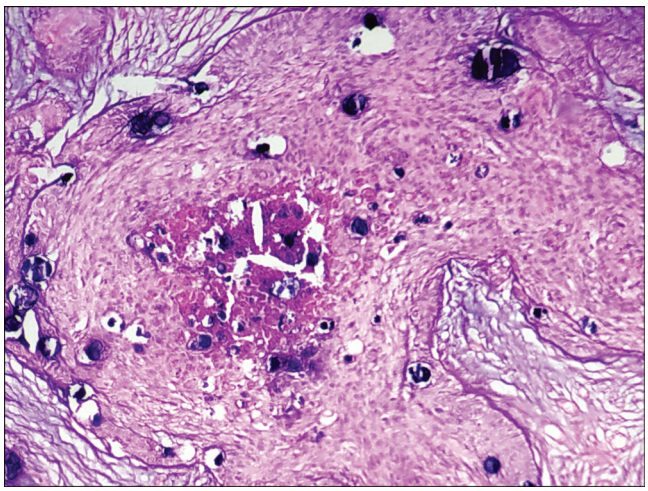

Figure 9: Extensive dystrophic calcification, H & E, x100

Table 1: Clinico-pathological profile of various giant trichoblastomas in literature

Sl. no.	Author	Year	Tumor size (cm) (max dimension)	Location
1	Requena and Barat[5]	1993	8	Scalp
2	Ogata *et al.*[8]	1999	9.5	Upper arm
3	Morillo *et al.*[9]	2006	6	Right buttock
4	Pragati *et al.*[6]	2009	6.6	Suprapatellar region
5	Landolsi *et al.*[10]	2011	5	Scalp
6	Nguyen *et al.*[11]	2016	5.5	Right thigh
7	Frings *et al.*[12]	2017	5	Right upper leg
8	Our case	2021	6	Right thigh

extremities, perianal and genital areas has been reported by some authors, but involvement of distal extremities is a rare event with less than 10 cases reported in the literature. They usually take the form of dermal nodules or papules measuring less than 2 cm, but enormous sizes up to 8 cm have been recorded.[7]

Majority of the tumors are sporadic and isolated. Some sporadic cases have been associated with certain genetic mutations particularly 9q22.3. An association with familial syndromes such as Brooke–Spiegler syndrome and Brooke–Fordyce syndrome is well known.[7]

Histologically pure trichoblastomas show lobules of uniform basaloid cells with prominent peripheral nuclear palisading and occassional cleft artifacting as described originally by Headington. Tumors with evidence of induction are characterized by the mesenchymal component, follicles in various stages of differentiation, and a secondary population of larger cells with pink cytoplasm resembling outer root sheath cells. Infundibulocystic, squamous, and sebaceous differentiation may be found. Cytologic atypia and mitoses are extremely rare.[1,3] Numerous histological subtypes such as nodular (small and large), retiform, cribriform, racemiform, columnar, and adamantinoid have been described.[7]

To the best of our knowledge, our case is one of those unique entities. A wide range of abnormal clinical and pathologic findings was recorded. To start with, the location of the tumor was at an unusual site with abnormal size misleading the clinician to an erroneous diagnosis of dermatofibrosarcoma. On microscopy, a constellation of rare findings such as cytologic atypia, abnormal mitoses (2/10 hpf), squamous eddies, extensive dystrophic calcification, retraction artifacts, focal melanin pigmentation, and mixed histological patterns were made out. Basal cell carcinoma which is the closest differential diagnosis was easily excluded on morphological and immunohistochemical grounds. Morphologically, the tumor had no connection anywhere to the overlying epidermis. Immunohistochemically, the tumor cells were negative for BerEP4, an epithelial cell adhesion molecule (EpCAM). Strong CK positivity was noted in the tumor cells indicating differentiation toward follicular outer root sheath. Ki 67 shows a proliferative index of 64%. Moreover, the presence of other histological features such as relatively larger tumor cells in reticulate pattern, infundibulocystic structures, and papillary mesenchymal bodies aided in making a diagnosis of trichoblastoma.

Tumors usually greater than 5 cm are considered as giant trichoblastomas and their reported numbers in literature are very few. A brief compilation of some of the reported cases is shown in Table 1.

In general, the behavior of these tumors is quite indolent, often persisting for years before a histological diagnosis is made. Local aggressive behavior is well-recognized contemplating the need for wide local excision as these tumors show evidence of invasion into the underlying skeletal muscle as reported by Cowen *et al*.[13] Such tumors have been designated by various terminologies such as low-grade trichoblastic carcinomas or unusually aggressive trichoblastomas.[14] Malignant transformation with demonstration of distant metastases in long-standing trichoblastomas has been reported by Regauer *et al*.[15] and Schulz *et al*.[16]

To conclude, trichoblastoma is a rare benign cutaneous tumor derived from follicular germinative cells showing varying histological features. Precise clinical diagnosis is essential as complete surgical excision is usually curative despite local aggressiveness.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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