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Case Report

Malignant proliferating trichilemmal tumor of thigh: a case report of an unusual location of a rare cutaneous adnexal tumor

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ABSTRACT

Proliferating trichilemmal tumour (PTT) is a well-circumscribed dermal or subcutaneous neoplasm with squamoid cytologic features and trichilemmal-type of keratinization. Usually, proliferating trichilemmal cysts are benign, although rarely, can undergo malignant transformation, at which point they are referred to as malignant proliferating trichilemmal tumours (MPTTs). Ninety percent of PTTs occur on the scalp, but they have also been found on the forehead, nose, back, chest, abdomen, buttocks, elbow, wrist, mons pubis and vulva. Here a case of an 89-year-old woman with a malignant proliferating trichilemmal tumour of right thigh is discussed, which is a very rare site for occurrence of the same.

Keywords: Malignant proliferating trichilemmal tumour, Elderly female, Thigh, Cystic swelling, Skin involved

INTRODUCTION

A proliferating trichilemmal cyst is a benign keratin-filled lesion that originates from the outer hair root sheath. They can rarely undergo malignant transformation to develop malignant proliferating trichilemmal tumours (MPTTs).[1] Clinically, MPTTs are rapidly growing masses with necrosis of underlying tissue; the scalp is the most common site of MPTTs.

Histological features of MPTTs include abrupt keratinization, anaplastic and pleomorphic cells, an increased number of mitotic figures, and infiltrative growth.[1] Surgical resection with a 1 cm margin is the treatment modality of choice.[2] Here, a case of 89-year-old female with a MPTT of right thigh is been presented.

CASE REPORT

An 89-year-old woman Eastern Cooperative Oncology Group (ECOG) 3 patient presented to the out-patient for the evaluation of a swelling on her right thigh below the inguinal region. The swelling was present for two years and was painless, there was no prior history of any trauma. The swelling was slow growing except for the past eight months, when it showed rapid increase in size, with a reddish-blue hue developing on the lateral aspect of the swelling since last three months, which alarmed the bystanders for which they consulted the hospital. According to the bystanders, the swelling was always compressible and soft. Her medical history and family history were unremarkable. On examination a single hemispherical swelling, non-tender 14×10 cm on the anteromedial aspect of right upper thigh starting 3cm below the inguinal region, with well-defined edges. The plane of the swelling was subcutaneous with no underlying muscle involvement, there was no arterial or nerve involvement as well. The surface of the swelling was smooth, tense and glossy with prominent veins over it. .A reddish blue hue was present over the lateral aspect of the swelling with fixity to the skin at that point .It was soft in consistency with positive fluctuation test but non

reducible, non-compressible, non-pulsatile. There was no inguinal lymphadenopathy (Figure 1).

The examination was followed by hematological and biochemical investigations, which were normal and imaging studies. Initially an ultrasonography was conducted which suggested a chronic collection in subcutaneous plane? abscess/hematoma (Figure 2). Suspecting neither, a contrast CT was done which described a chronic collection abutting common femoral artery and sartorius muscle with gross involvement and no lymph node enlargement (Figure 3 and 4).

She underwent a simple excision of the swelling with the involved skin, removing the cyst in total and surrounding fatty tissues under local anesthesia. Intraoperatively a biloculated cystic swelling with straw colored fluid was excised, with maintaining the fat plane between the common femoral artery and sartorius muscle, the skin was involved at the lateral aspect which was removed along with the swelling. No intraoperative pictures were taken since malignancy was not suspected.

Histopathological examination revealed cyst lined by stratified squamous epithelium showing severe dysplasia, lumen showed keratin with malignant spindle cells, squamoid nests with trichilemmal keratinization. They showed moderate to severe pleomorphism with margins in medial aspect involved focally. A provisional diagnosis of cystic squamous cell carcinoma was made with advice to do Immunohistochemistry, resulting in the final diagnosis of malignant proliferating trichilemmal tumor with poorly differentiated spindle cell areas (Figure 5-7).

Following histopathological diagnosis, patient was advised re-excision of the medial border or radiation therapy. Considering the age and the physical status of the patient, her bystanders were not willing for both and became lost follow-up.

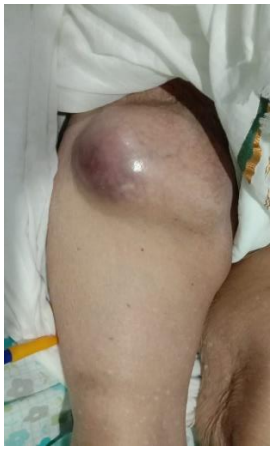


Figure 1: Right upper thigh showing a swelling 14×10 cm in the anteromedial aspect. The prominent blood vessels and the reddish blue hue over the lateral aspect at the point of skin involvement can also be appreciated.

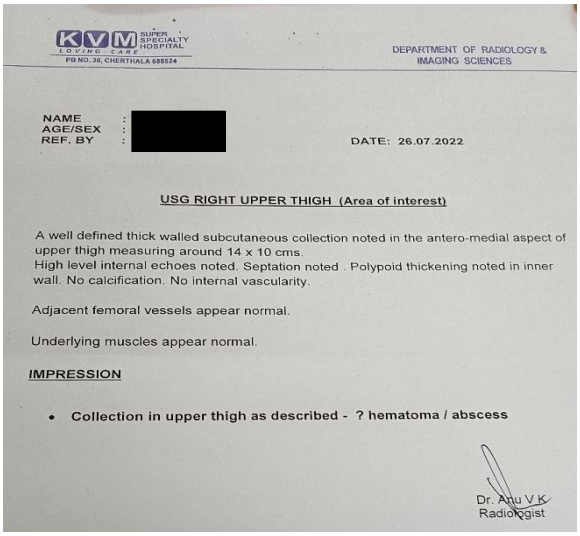


Figure 2: The ultrasonography report of the swelling.

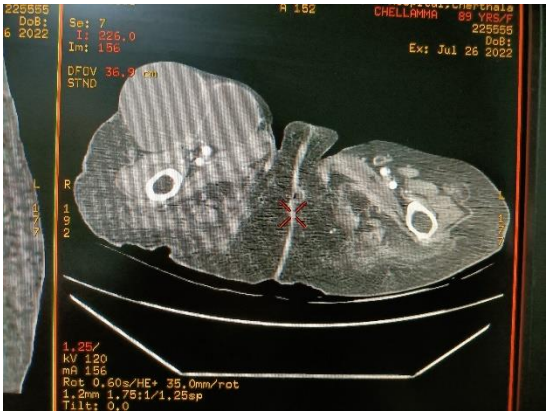


Figure 3: Axial cut of the contrast CT scan showing the biloculated nature of the swelling. A well-defined cystic swelling can be appreciated with close margins to the common femoral artery and sartorius muscle.

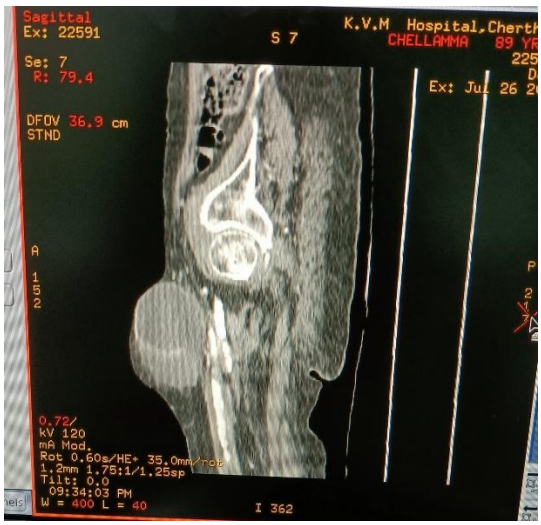
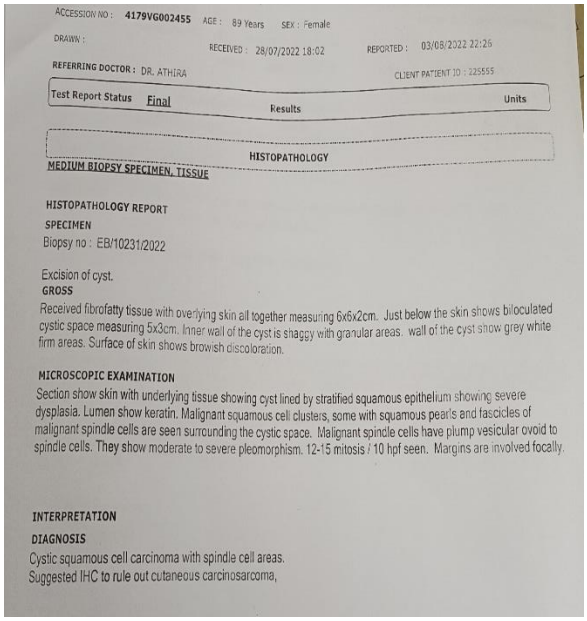


Figure 4: A sagittal cut through the contrast CT showing the cystic swelling and the relation to the common femoral artery placed posteriorly.



ACCESSION NO : 4179VG002455 AGE : 89 Years SEX : Female
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REFERRING DOCTOR : DR. ATHIRA CLIENT PATIENT ID : 225555

Test Report Status Final	Results	Units

HISTOPATHOLOGY

MEDIUM BIOPSY SPECIMEN, TISSUE

HISTOPATHOLOGY REPORT

SPECIMEN

Biopsy no : EB/10231/2022

Excision of cyst.

GROSS

Received fibrofatty tissue with overlying skin all together measuring 6x6x2cm. Just below the skin shows biloculated cystic space measuring 5x3cm. Inner wall of the cyst is shaggy with granular areas. wall of the cyst show grey white firm areas. Surface of skin shows browish discoloration.

MICROSCOPIC EXAMINATION

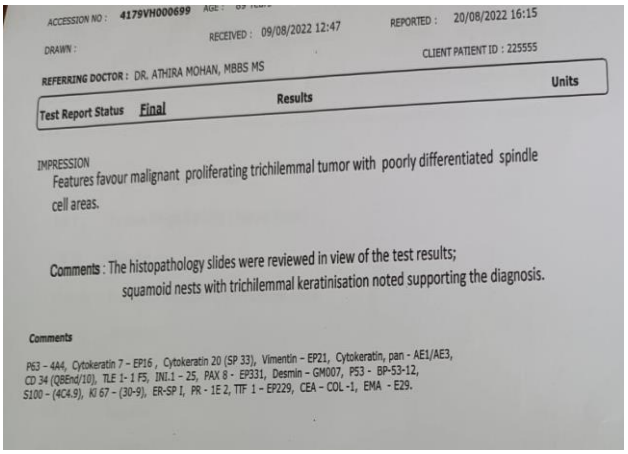
Section show skin with underlying tissue showing cyst lined by stratified squamous epithelium showing severe dysplasia. Lumen show keratin. Malignant squamous cell clusters, some with squamous pearls and fascicles of malignant spindle cells are seen surrounding the cystic space. Malignant spindle cells have plump vesicular ovoid to spindle cells. They show moderate to severe pleomorphism. 12-15 mitosis / 10 hpf seen. Margins are involved focally.

INTERPRETATION

DIAGNOSIS

Cystic squamous cell carcinoma with spindle cell areas.
Suggested IHC to rule out cutaneous carcinosarcoma,

Figure 5: The histopathology report of the excised sample.



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IMPRESSION

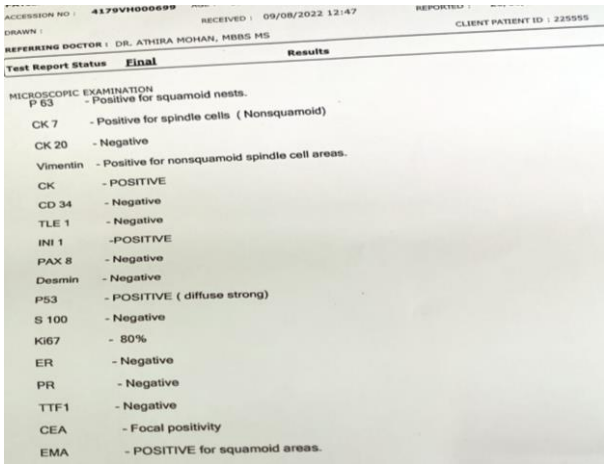
Features favour malignant proliferating trichilemmal tumor with poorly differentiated spindle cell areas.

Comments : The histopathology slides were reviewed in view of the test results; squamoid nests with trichilemmal keratinisation noted supporting the diagnosis.

Comments

P63 – 4A4, Cytokeratin 7 – EP16, Cytokeratin 20 (SP 33), Vimentin – EP21, Cytokeratin, pan - AE1/AE3, CD 34 (QBEnd/10), TLE 1- 1 F5, INI.1 – 25, PAX 8 - EP331, Desmin – GM007, P53 - BP-53-12, S100 – (4C4.9), Ki 67 – (30-9), ER-SP I, PR - 1E 2, TTF 1 – EP229, CEA – COL -1, EMA - E29.

Figure 6: The final diagnosis after immunohistochemistry.



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REFERRING DOCTOR : DR. ATHIRA MOHAN, MBBS MS

Test Report Status Final	Results

MICROSCOPIC EXAMINATION

P 63	- Positive for squamoid nests.
CK 7	- Positive for spindle cells (Nonsquamoid)
CK 20	- Negative
Vimentin	- Positive for nonsquamoid spindle cell areas.
CK	- POSITIVE
CD 34	- Negative
TLE 1	- Negative
INI 1	-POSITIVE
PAX 8	- Negative
Desmin	- Negative
P53	- POSITIVE (diffuse strong)
S 100	- Negative
Ki67	- 80%
ER	- Negative
PR	- Negative
TTF1	- Negative
CEA	- Focal positivity
EMA	- POSITIVE for squamoid areas.

Figure 7: The immunohistochemistry markers used in the study.

DISCUSSION

Proliferating trichilemmal tumour (PTT) is a well-circumscribed dermal or subcutaneous neoplasm which histologically simulate squamous cell carcinoma but with trichilemmal-type of keratinization.[3] This neoplasm was first recognized by Wilson-Jones in 1966.[4] These tumours were given many names like pilar tumour of the scalp, proliferating trichilemmal cyst, proliferating epidermoid cyst, giant hair matrix tumour, hydatidiform keratinous cyst, trichochlamydocarcinoma, and invasive hair matrix tumour.[5]

PTT can be divided into three groups based on the degree of stromal invasion and the level of cellular atypia into benign, low grade malignant and high-grade malignant. Benign group showed no infiltration to surrounding stroma and minimal nuclear atypia. Low grade tumours were composed of squamoid cells with large, hyperchromatic nuclei and irregular nuclear membranes surrounded by abundant eosinophilic cytoplasm. Necrosis, abrupt keratinization and extension into surrounding dermis are the other features. These tumours are centrally filled with homogeneous acellular eosinophilic material representing amorphous debris and pilar keratin. High grade tumours were invasive; they were cytologically anaplastic.[6]

In 1983, Saida et al reported the first case of an MPTT and distinguished MPTTs from benign trichilemmal tumours based on the degree of atypia and mitotic activity.[7] PTT is more commonly seen in elderly women. The most common site of occurrence of PTTs is the scalp, but they can also be found on the forehead, nose, back, chest, abdomen, buttocks, elbow, wrist, mons pubis and vulva.

PTT is usually benign but can rarely undergo malignant transformation in a step-wise manner either following trauma and inflammation or as de novo.[1,7,8] Malignant and benign tumours can be differentiated based on the following features - rate of growth, invasion, metastasis, anaplasia and cellular atypia.[9] Metastasis from malignant PTT is a rare occurrence.[10]

MPTT is histologically similar to squamous cell carcinoma, the presence of a trichilemmal epithelium, which is not observed in squamous cell carcinomas can be used to distinguish them.[11] IHC can also be used to distinguish both. In this case the patient had a nearly dormant swelling for 2 years, then the swelling enlarged at a rapid pace which is a feature of malignant transformation and which was proved microscopically by the, nuclear atypia and mitosis.

Surgical excision with wide (1 cm) margin is the modality of treatment.[1] Mohs surgery has been suggested as a superior alternative.[2] Radiation therapy can also be used for local tumours. For metastatic MPTTs, chemotherapy may be needed.[1] MPTTs can recur even after complete removal so follow-up is needed.[12] Follow-up at three-month intervals for two years, then six-month intervals for

three years is current norm. If no recurrence is noted at five years, then yearly follow up is done.

CONCLUSION

Malignant transformation of the proliferating trichilemmal cysts are a rare occurrence and when they occur, they are referred to as MPTTs. Histopathological examination is crucial in establishing the diagnosis of MPTTs and reveals abrupt keratinization, anaplastic and pleomorphic cells, and increased mitotic forms. Simple surgical resection with wide margins, Mohs surgery, and radiation therapy have all been reported in the management of local MPTTs that have not metastasized. A vigilant follow-up protocol may be warranted to monitor for recurrence.

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Ethical approval: Not required

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