INTRODUCTION

Trichilemmal carcinoma (TLC) is a rare unusual malignant adnexal neoplasm arising from the outer root sheath of the hair follicular epithelium. TLC generally has an indolent clinical course and is found usually as a solitary lesion occurring in sun exposed areas especially the face, head and neck, extremities and trunk. Differentiation from other high grade malignant tumors such as squamous cell carcinoma (SCC), basal cell carcinoma (BCC) and other proliferating trichilemmal tumors is essential as TLC is a low grade carcinoma with low metastatic potential. Hence, complete wide excision of tumor and follow-up of the patient is sufficient as treatment protocol unlike other high grade skin tumors.

CASE REPORT

The present case report is about a 60-year-old female patient presented with nodule over the right side of neck since 3 months. The patient was apparently in good health and did not have any history of systemic pathology. Local examination revealed a skin nodule with overlying cutaneous pigmentation over the right side of the neck. Nodule measured 2 cm × 1 cm and was associated with tenderness. The regional lymph nodes were not palpable. A clinical diagnosis of cutaneous horn was made. Excision was performed under local anesthesia and specimen was sent for histopathological examination.

Gross examination of the specimen revealed a single grey brown nodular soft tissue mass measuring 1.5 cm × 1 cm. External surface showed a friable grey black outgrowth measuring 0.5 cm × 0.5 cm. Cut section revealed grey white area beneath the pigmented outgrowth.

Histopathological examination revealed marked hyperkeratosis of stratum corneum. Dermis showed tumor cells arranged in a lobular pattern extending deep in the dermis up to the subcutaneous fat. Tumor cells were polygonal with abundant cytoplasm and showed moderate nuclear atypia and mitotic activity <5/10 hpf along with peripheral palisading and trichilemmal keratinization [Figure 1]. Some of the lobules showed clear cell change, which showed periodic acid Schiff (PAS) positivity [Figure 2]. The immunohistochemistry (IHC) showed tumor cells positive for pan-cytokeratin AE1/AE3, but epithelial membrane antigen (EMA) was negative. Hence a diagnosis of TLC was made. Patient did not return for follow-up later.

DISCUSSION

TLC is a rare locally invasive malignant tumor arising from the external root sheath hair follicular epithelium. The term
TLC was first given by Headington and French in 1962, who originally described this neoplasm as a tumor of clear cells in continuity with a coexisting trichilemmoma. TLC occurs usually in elderly age group with no sex predilection over the sun exposed areas of head and neck, face, extremities and trunk. Few case reports have described this tumor occurring over the eyelid and upper lip.

This tumor usually presents as an epidermal papule, plaque or nodule which may be erythematous, tan or flesh colored. These can be associated with superficial ulceration or keratosis.

A study by Swanson et al. analyzed the clinical findings in 10 cases of TLC and found that the tumor occurred in the hair bearing and sun exposed skin which included the scalp, face, trunk or upper extremities. Size of tumor usually ranged from 0.4 to 2 cm with less than 1 year history. However, few reports show TLC occurring in unexposed parts of the body such as thigh, presence of metastasis and recurrence.

The pathogenesis of this neoplasm remains unclear, though significant numbers of cases of TLC develop de novo. However, factors such as actinic damage, long dose irradiation and transformation from benign trichilemmoma have been postulated. Schell and Haneke reported 11 cases of TLC and found that the tumor often occurred in the slightly damaged skin of elderly people.

Microscopically TLC consists of multiple intradermal relatively well circumscribed lobules or trabeculae with a peripheral palisade of basaloid cells in continuity with the epidermis. The predominant tumor cells are polygonal with abundant clear, glycogen rich PAS positive cytoplasm. Atypia and a high mitotic index are usually present. Trichilemmal keratinization, characterized by abrupt keratinization without a granular layer is a characteristic finding.

Wong and Suster, reviewed thirteen cases of TLC and observed similar histologic features in all cases.

Headington suggested criteria for the histological diagnosis of TLC, i.e.,
- Continuity with a co-existing benign epithelial tumor (trichilemmoma) and/or
- Continuity with the outer sheath epithelium of a co-existing hair follicle and/or
- Light microscopic evidence suggestive of outer sheath architecture: Glycogen rich epithelium, peripheral cell palisading, prominent basement membrane zone and/or
- Focal epithelial differentiation (keratinization) in a trichilemmal model: (Absent or minimal granular layer, abrupt single cell keratinization and formation of dense non-lamellar keratin) and/or
- Electron microscopic cellular detail similar to that found in either normal sheath epithelium or trichilemmoma and/or
- Immunocytochemical similarities of normal outer sheath epithelium or to trichilemmomas, e.g. positive for monoclonal antibodies to hair associated keratins or other antigens of the outer sheath.

The histopathologic features of our case fulfilled the criteria 3, 4 and 6.

On IHC, TLC is positive for cytokeratin 7, 8, 18 and 19 and also found to be positive for hair follicle differentiation, but negative for carcinoembryonic antigen and EMA. CD34 expression is also a feature said to support outer root sheath differentiation.

The differential diagnosis to be considered is SCC, BCC and proliferating trichilemmal tumors. Clear cell SCC lacks trichilemmal keratinization, lobular growth and adnexal extension. BCC mimics TLC, but areas of clear cell change
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often involves a minor portion of the tumor and peripheral palisading cells rarely show subnuclear vacuolization which is distinctive of hair follicular differentiation. Proliferating trichilemmal tumor shows well-circumscribed nodulocystic lesion containing homogenous keratotic material unlike TLC, which shows extensive infiltration into the adjacent structures.[13]

The present case showed lobular pattern with tumor cells showing clear cytoplasm, peripheral palisading and trichilemmal keratinization. IHC was positive for AE1 and AE3 but negative for EMA. Hence a diagnosis of TLC was made. The presence of extensive hyperkeratosis in stratum corneum leads to a clinical diagnosis of cutaneous horn.

As TLC is a tumor of low malignant potential and has indolent clinical course, the treatment is exclusively surgical. Simple excision with adequate margins is safe and inexpensive. TLC generally has a good prognosis and reports of deep invasion and local recurrence are uncommon. Recurrence of TLC though rare, has been reported in a few cases.

REFERENCES


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