

AGGRESSIVE TRICHOLEMAL CARCINOMA OF THE AXILLA

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Tricholemmal carcinoma is a rare cutaneous adnexal tumor of external hair sheath origin generally occurring on sun-exposed, hair-bearing skin of elderly individuals. The scalp and face are the most common localizations of the tumor and a tricholemmal carcinoma in the axilla has not been reported previously. Presented here is a case of axillary tricholemmal carcinoma with an aggressive nature leading to invasion of the thoracic wall and the parietal pleura. The treatment protocol included amputation of the upper limb, partial resection of the thoracic wall and reconstruction with a free flap.

Key Words: Tricholemmal Carcinoma, Axilla, Aggressive Local Invasion, Recurrence, Amputation.

Aksillada Agresif Trikolemmal Karsinoma

Trikolemmal ca (TLC) saç kılıfı kökenli, nadir bir adneksal tümör olup skalp ve yüz en sık görüldüğü lokalizasyonlardır. Burada sunulan, torasik duvarı ve parietal plevrayı invaze eden agresif yapıda, şimdiye kadar hiç rapor edilmemiş bir aksiller TLC vakasıdır. Tedavi protokolü üst ekstremitenin tamamının ve göğüs ön duvarının kısmi amputasyonunu ve defektin serbest doku aktarımıyla rekonstrüksiyonunu içermiştir.

Anahtar Kelimeler: Trikolemmal Karsinom, Aksilla, Agresif Lokal İnvazyon, Rekürrens, Amputasyon.

Epithelial cysts and tricholemmomas are common benign lesions of the skin with extremely rare malignant degeneration, and are usually excised for cosmetic purposes. Merkel cell carcinoma, basal cell carcinoma, Bowen's disease, Paget's disease, sebaceous carcinoma and squamous cell carcinoma were thought to originate from cystic lesions, and malignant transformation of proliferating tricholemmal cysts have also been reported (1,2). Tricholemmal carcinoma is a rare cutaneous adnexal tumor of external hair sheath origin that must be differentiated from the malignant and premalignant lesions above and some benign conditions like verrucous cysts and pseudocarcinomatous hyperplasia in ruptured cysts. Typical localization of a tricholemmal carcinoma is the hair-bearing, sun-exposed skin of the elderly (2-5). We hereby report a patient with a recurrent, aggressive axillary tricholemmal carcinoma showing pleural invasion and requiring amputation of the upper limb.

CASE REPORT

A previously healthy 66-year-old man had noticed an indolent, colorless, olive-sized nodular mass in his left axilla 2.5 years before. A fine needle aspiration biopsy of the cystic lesion revealed malignant epithelial cells, but the patient refused any treatment until necrotic drainage had begun, 15 months later. The axillary mass, reaching 10 cm in diameter, was resected with a simultaneous lymph node dissection in another medical center and the patient received 60 Gy external radiotherapy to his left axilla in the following 4 months after the histological diagnosis of apocrine carcinoma. Two months later, a 2 x 2 cm mass protruding from the left axilla was detected on physical examination and he was referred to our hospital for further treatment. On admission, the patient had a large, fixed axillary mass, severe upper extremity edema, and no range of motion in the shoulder joint (Fig. 1).

An incisional biopsy followed by further evaluation of the previous specimen with specific dyes indicated a diagnosis of tricholemmal carcinoma (Fig. 2). Clear cytoplasmic tumor cells showing prominent atypia and a high mitotic rate were observed with hematoxylin-eosin staining. These cells were of tricholemmal type with abrupt keratinization without a granular cell layer. AB PAS positive, diastase digestible deposits of glycogen were found in the cytoplasm of most of the clear cells, constituting a cytological hallmark. Features of outer-root sheath differentiation were shown by LMWK and HMWK stains. Basal layer columnar clear cells showing a tendency toward palisading, prominent basement membrane and focal involvement of interfollicular epidermis were detected. Infiltrating margins, abnormal mitotic features, marked cellular and nuclear pleomorphism were all suggestive of tricholemmal carcinoma, eliminating a possible diagnosis of squamous cell carcinoma.

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Figure 1: Severe edema in the left upper extremity of the patient.

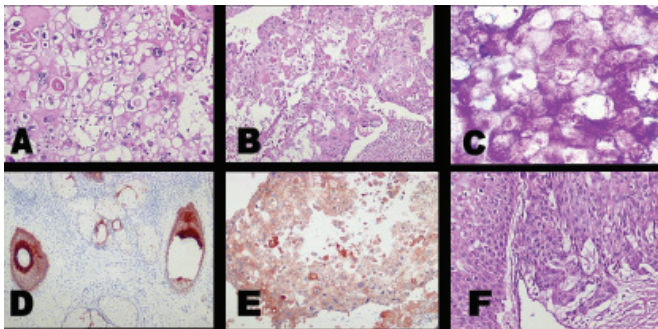
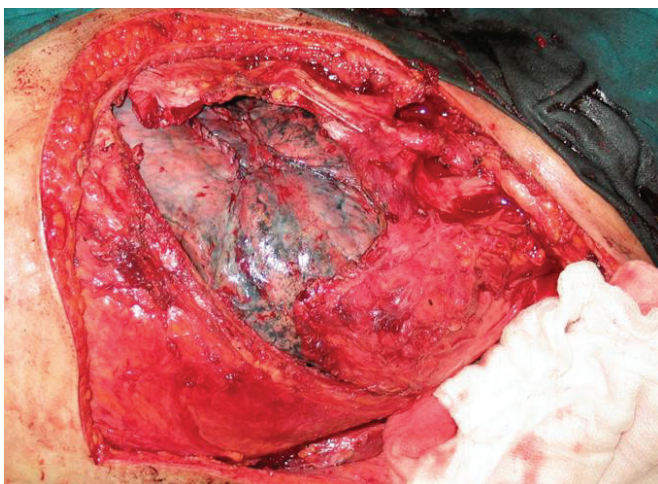


Figure 2: A; Clear cytoplasmic tumor cells showing prominent cellular atypia (x100). B; tumor cells showing prominent keratinization (x40). C; glycogen positivity with Alcian blue PAS stain in clear cells (x200). D; dominant staining with LMWK in follicular infundibulum (x40). E; positive staining with HMWK (x40). F; tumor cells with peripheral palisading (x100).



The preoperative evaluation with MRI showed that Figure 3: The limb amputated, thoracic wall resected and the lung exposed.

the axillary neurovascular structures were invaded by the tumoral tissue. During surgery, it was impossible to dissect the neurovascular elements out of the tumor and an upper extremity amputation including the scapula was performed. It was not feasible to salvage the extremity with a vascular graft, because the whole soft tissue of the arm including the muscle and the periosteum of the humerus were invaded. The third and fourth ribs were also partially resected because of local invasion (Fig. 3). Reconstruction was achieved with a fillet flap from the deltoid region and a transverse epigastric fasciocutaneous flap covering the dual mesh that was used to repair the thoracic wall. A final reconstruction with a free latissimus dorsi myocutaneous flap was required for complete healing after partial necrosis in the tips of the flaps (Fig. 4). The final histological examination confirmed the diagnosis of tricholemmal carcinoma with invasion of the parietal pleura. The patient underwent systemic chemotherapy, and was free of any symptoms for six months.

DISCUSSION

Tricholemmal cyst walls resemble hair follicles in the catagen phase. When the anagen phase ends, a high percentage of the outer hair sheath undergoes apoptotic reduction and the follicle again goes into the catagen phase. When this is genetically mediated in an abnormal way, tricholemmal cysts occur (6). These cysts are mostly detected on the scalp, but are also found on the trunk, face and extremities, less often (2,7). The biological behavior of a cutaneous cyst undergoing malign transformation is important as it destroys tissues locally or by metastasis (1). Tricholemmal and other epithelial cysts may be the most common benign lesions, but their malign degeneration is extremely rare. Infiltrating margins, marked cellular and nuclear pleomorphism, abnormal mitotic figures and aneuploidy are the affirmative histological criteria for malignancy (1,7-9). The malignant transformation ratio was ascertained as 2-3%, including other clear cell tumors or



Figure 4: The patient three months after the final reconstruction with the latissimus dorsi free flap.

their variants (1). Although extremely rare, their differential histologic characteristics are determined as a prominent clear cell component and outer hair sheath differentiation type keratinization without granular layer interposition (2,7). They are most often confused with basal cell carcinoma, keratoacantoma, squamous cell carcinoma, Bowen's disease, Paget's disease, sebaceous carcinoma, mucin-producing adenocarcinoma, metastatic renal cell carcinoma, balloon cell malignant carcinoma and most sweat gland carcinomas (1,2,5,8,9).

Tricholemmal carcinoma must be differentiated from other malignant, premalignant and benign conditions to ensure appropriate management. Common agreement on the treatment protocol as complete excision without any other treatment was based on the metastasis-free nature of this lesion (1-5,8). However, Noto et al. reported a patient having many local relapses that required several surgical excisions, radiotherapy and chemotherapy (7). Our case also showed that tricholemmal carcinoma leads to amputation of a limb and by reaching the pleura with local invasion requires systemic therapy, if treatment is delayed. Although seen extremely rarely, a surgeon or the pathologist must be suspicious of a possible tricholemmal tumor in an axillary localization; it requires an aggressive treatment protocol with close follow-up.

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KAYNAKLAR

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