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Auricular Helix Carcinosarcoma; an Aggressive Tumor Characterized: Case Report

Agresif Seyir Gösteren Aurikula Heliks Karsinosarkomu: Olgu Sunumu

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Abstract / Özet

Primary cutaneous carcinoma is a dermal biphasic tumor that is more commonly encountered in the upper extremities and the head and neck region, as a result of sunshine exposure. It is a rare tumor and occurs more frequently in older males. Immunohistochemical evaluation is required for the diagnosis. The epithelial components of the tumor include squamous cell carcinoma, basal cell carcinoma and adnexial tumors; mesenchymal components are pleomorphic sarcoma, atypical fibroxanthoma and osteosarcoma. We present a rare case where auricular helix carcinosarcoma metastasized to the parotid gland within the first year following total excision. The patient had a total parotidectomy as well as a modified radical neck dissection type II and postoperative adjuvant radiotherapy. In spite of the surgery and radiotherapy, the tumor had an aggressive course, with recurrence in the temporomandibular region, and the patient was lost on the 11th month of the follow-up.

Key Words: Carcinosarcoma, auricular helix, epithelial component, mesenchymal component

Primer kutanöz karsinosarkom, epiteliyal ve mezenşimal komponentleri olan sıklıkla vücudun güneş ışığına maruz kalan baş boyun ve üst ekstremite gibi bölgelerinde görülen bifazik bir tümördür. Daha çok yaşlı erkeklerde görülür ve oldukça nadirdir. Günümüzde histogenezisi hala tartışmalı olan bu tümörün tanısı immunhistokimyasal değerlendirme ile konur. Tümörün epiteliyal komponentleri daha çok squamöz hücreli karsinom, bazal hücreli karsinom ve adneksal tümörler iken, mezenşimal komponentleri pleomorfik sarkoma, atipik fibroksantoma ve osteosarkomadır. Literatürde çok az sayıda bildirilen aurikula heliks karsinosarkom vakalarına katkıda bulunmak amacıyla bir yıl önce aurikula heliksinden total eksize edilen karsinosarkom olgusunun parotis metastazını sunuyoruz. Hastaya total parotidektomi ve modifiye radikal boyun diseksiyonu tip 2 yapılmıştır. Hastanın genel durumu kemoterapi alması için uygun olmamıştır. Postoperatif 25 seans adjuvan radyoterapi yapılan hastanın takiplerinde tümör agresif bir seyir göstermiş ve altıncı ay içinde temporomandibular bölgede rekürrens saptanmıştır. Bunun üzerine hastaya 25 seans daha radyoterapi uygulanmış ve hasta 11. ayda eks olmuştur.

Anahtar Kelimeler: Koter Karsinosarkom, aurikula heliks, epiteliyal komponent, mezenkimal komponent

Introduction

Primary cutaneous carcinosarcoma is a rarely encountered tumor with epithelial and mesenchymal components. It is more common in aged males. The regions of the body exposed to sunlight, including the head&neck and upper extremity are more prone. Half of all cases are localised in the head&neck region while one third are localised in the extremities (1). A few cases of primary auricular carcinosarcoma were reported previously in the literature (2). The management is total excison of the lesion. The immunohistochemical evaluation is crucial in the diagnosis. The present case of auricular helix carcinosarcoma metastatic to the parotid gland was presented with the objective of providing an additional contribution to previously reported cases and to emphasize the aggressiveness of the tumor.

Case Report

An eighty year old male patient was referred with the compliant of a painful mass in the parotid region of two months duration. The patient had a history of a 1 cm lesion excison localised in the middle of the left auricular helix with a subsequent histopathological diagnosis of sarcomatous type squamous cell carcinoma. Examination of the left parotid region revealed a firm, fixed, tender 5x5x4 cm mass occupying the parotid lodge and expanding to the tragal cartilage anteriorly, angulus mandibula inferiorly, and 1 cm anterior to the hairy skin in occipital region posteriorly (Figure 1). Laryngeal examination revealed left vocal cord abductor paralysis. MRI confirmed a contrast positive mass in the tail of the left parotid gland (Figure 2). Radiologically, the lesion showed sternocleidomastoid muscle (SCM) and carotid sheet invasion. The patient had total parotidectomy and modified radical neck dissection type 2 (Figure 3). The peroperative frozen biopsy revealed neural invasion in the cervicofacial branch of facial nerve observed in close proximity with the tumor and was excised. The histopathology was consistent with the lesion excised from the auricular helix one year earlier and reported as a squamous cell carcinoma with a sarcomatous component. f Four of the lymph nodes in the neck dissection material had the same tumoral infiltration features and were evaluated as the metastasis of auricular helix carcinosarcoma. Parotid gland carcinoma invasion, lymphatic invasion and perineural invasion was detected in the excised parotid gland specimen.

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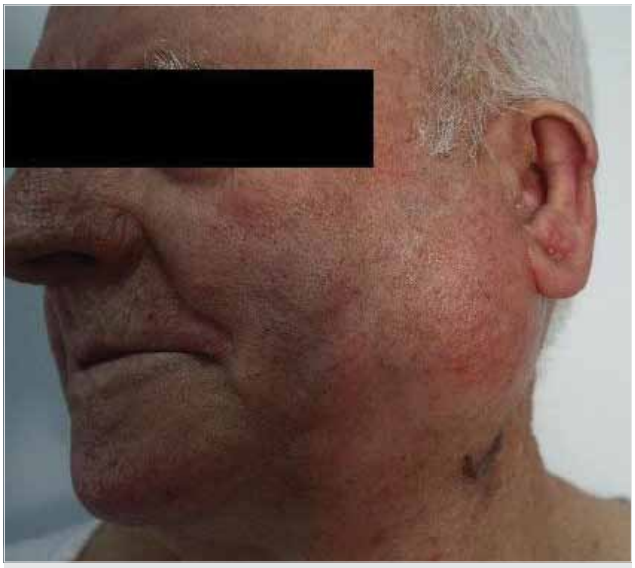

Figure 1. Clinical appearance

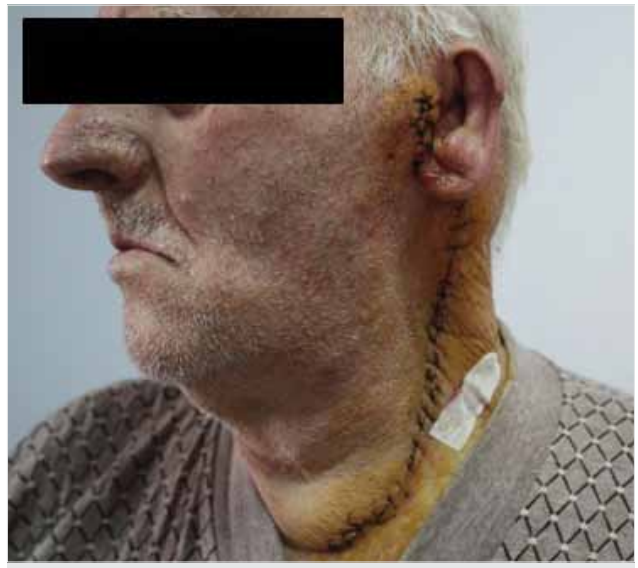

Figure 3. The patient's postoperative appearance

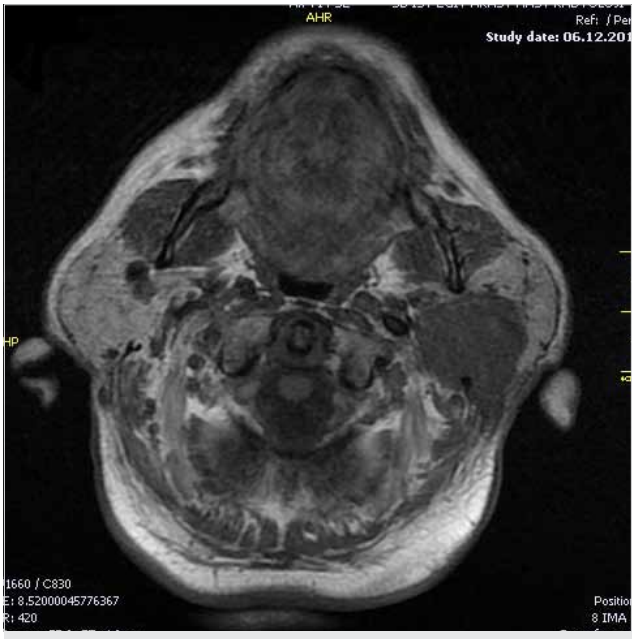

Figure 2. Preoperative MRI view

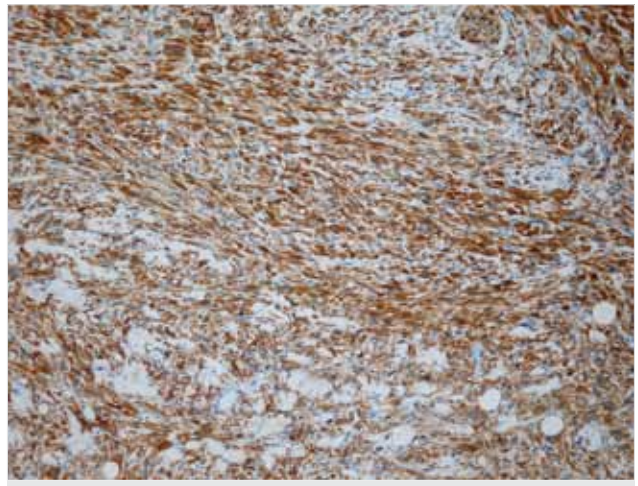

Figure 4. Vimentin positivity in tumor cells by immunohistochemistry

The cytokeratin (CK) 7 and smooth muscle actin (SMA) was positive; S100 was negative in immunohistochemical examination. Diffuse vimentin and keratine (AE 1-3) positivity was detected (Figure 4). The patient had postoperative adjuvant radiotherapy (4500cGy) in 25 treatments. Chemotherapy was avoided in order not to distrupt the general condition of the patient. The patient was lost on the 11th month of the follow-up with recurrence in the temporomandibular region despite vigorous therapy (Figure 5).

Discussion

Carcinosarcoma is a rare tumor consisting of carcinomatous and sarcomatous components (3). Carcinosarcoma has been reported to arise in many tissues, including the genitourinary tract, lung, breast, parotid gland, gastrointestinal system, thymus and skin. Half of the cases are seen in the head&neck region (1). Cutaneous carcinosarcomas are commonly encountered in old age with a male predominance. They are most commonly reported in sun exposed regions including the upper extremity and head&neck region but are also reported in the female genital tract. Adiameter of more than 2 cm, previous dermal tumoral lesion excision, different growth pattern, lymph node metastasis and visceral origin are bad prognostic factors (4). The five year survival rate was reported as 70%. The definite diagnosis was achieved through immunohistochemical evaluation. The few cases in the literature were managed with total excision and local invasion was reported. The present case had a metastatic lesion in the parotid gland following total excision of carcinosarcoma localised in the auricular helix.

Although the epithelial components are basal cell and/or squamous cell carcinoma in most cases, adnexial tumors are predominant in the remainder. The mesenchymal components include pleomorphic sarcoma (undifferentiated sarcoma), atypical fibroxanthoma or osteosarcoma. This feature is evaluated as a biphasic tumor and named as carcinosarcoma by many authors (5). The presence of epithelial and mesenchymal components in histological and immunohistochemical evaluation are diagnostic (Figure 6). Keratine AE 1 and AE 3, Vimentin, CK 7 and SMA positivity as malignant components are important in diagnosis.

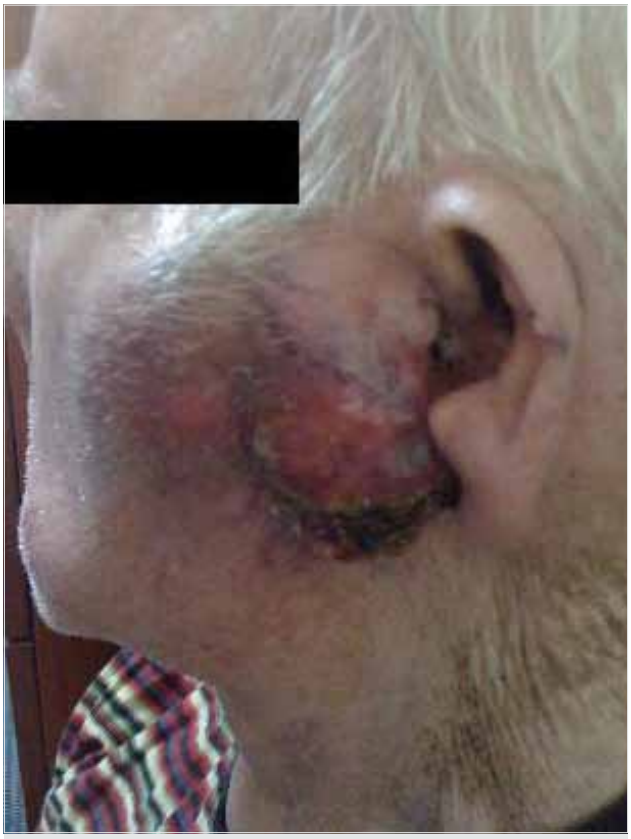

Figure 5. Recurrence of the temporomandibular region

The histogenesis of carcinosarcomas is still controversial. Various theories are reported concerning the developement of tumor. The most accepted one is the conversion theory. This theory was supported by the demonstration of p53 tumor suppressor gene mutation both in epithelial and mesenchymal components. P53, AE1 and AE3 are proven to have high sensitivity in the diagnosis of carcinosarcomas (6).

Many of the cases were treated with the combination of surgical excision,radiation therapy and chemotherapy. Chemotherapy agents are vincristine, 5-FU, cisplatin, doxorubicin and etoposide (7). In our case chemotherapy was avoided in order not to worsen the general condition of the patient.

Conclusion

Auricula helix carcinosarcoma is a rare agressive tumor. There are no evidence based and well established treatment protocols. The aggressive course should be considered and management should include surgery, radiotherapy and chemotherapy to achieve complet cure in the majority of cases.

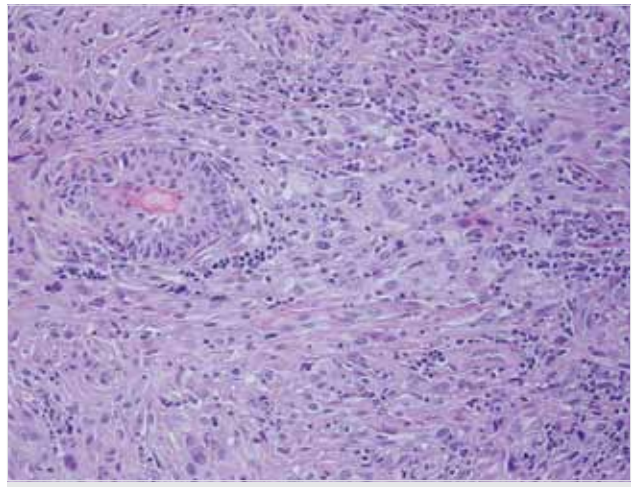

Figure 6. The presence of epithelial and mesencymal components in histological evaluation

Conflict of Interest

No conflict of interest was declared by the authors.

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