



LETTER TO THE  
EDITOR  
EDİTÖRE  
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# Cutaneous Carcinosarcoma: A Case Report and Review of The Theories on Histogenesis

## *Kutanöz Karsinosarkoma: Olgu Sunumu ve Histogenez Teorilerinin Gözden Geçirilmesi*

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Dear Editor,

Carcinosarcoma has been reported in various sites, such as the lung, uterus, breast, bladder, and salivary glands (1-6). Primary carcinosarcoma of the skin is very rare. To the best of our knowledge, almost 50 cases have been reported to date in English in the literature. The disease is defined by two different malignant components (7). In the reported cases, the epithelial component was basal cell carcinoma, squamous cell carcinoma, or adnexal carcinoma of the skin, such as porocarcinoma, trichilemmal cystic carcinoma, and spiradenocarcinoma (8-10). The mesenchymal component was osteosarcoma, chondrosarcoma, fibrosarcoma, or malignant fibrous histiocytoma (7-14). Carcinosarcomas at extracutaneous sites are characteristically aggressive tumours. Cutaneous basal cell carcinoma is an indolent tumour, but sarcomatoid transformation would increase the aggressive potential of the tumour.

Because of its rarity and variable terminology, cutaneous carcinosarcoma is still not completely characterised with regards to the histopathologic features and clinical behaviour. We present a particular case of cutaneous carcinosarcoma on the scalp with a long duration and rapid growth in a short time. Our case may contribute to the theories on the mechanism of the development of these tumours.

A 70-year-old male patient was admitted to hospital with a mass on the scalp. By clinical examination, an ulcero-vegetant mass was detected in the right parietal region of the scalp. The mass was 8 cm in diameter. According to the patient's history, the mass had been there for 50 years, but it had grown rapidly in the previous 2 months. Clinical or radiological findings of metastatic lesions were not found. The mass was excised with 1 cm of the surrounding tissue. Macroscopically, an 8 cm solid tumour with a hard consistency and bleeding areas was detected. Under microscopic examination, extensive surface ulceration and mixed inflammatory cell infiltration were observed. There were typical basal cell carcinoma islands consisting of basophilic cells with peripheral palisading and cleft formation. Between the islands of the basal cell carcinoma, the stroma was not hypercellular and did not have any atypical changes (Figure 1a). In the deeper portions of the lesion highly cellular another tumour, with different morphological features, was observed. There were spindle-shaped tumour cells with storiform pattern, histiocyte-like cells, and bizarre and multinucleated giant cells (Figure 1b and c). Each high-power field showed 1-2 mitotic figures. Extensive necrosis and an atypical mytotic figure was noted (Figure 1d). Between the epithelial islands, areas of osteoid formation were detected.

For differential diagnosis, immunohistochemical studies were performed. Cytokeratin (PAN-CK Cocktail, RTU, Neomarkers), EMA (E29, RTU, Neomarkers), vimentin (V9, RTU, Neomarkers), CD68 (KP-1, RTU, Neomarkers), S100 (4C4.9, RTU, RTU, Neomarkers), HMB45 (HMB50, HMB45+, RTU, Neomarkers), myoglobin (Ab-2, RTU, Neomarkers), desmin (d33, RTU, Neomarkers), and actin (1A4, RTU, Neomarkers) were used. The epithelial component was positive for cytokeratin (Figure 1e) and EMA. The mesenchymal component showed strong positivity with vimentin and CD68 (Figure 1f). Actin was weakly positive. Also, the mesenchymal component exhibited a lack of reactivity for the epithelial markers, myoglobin, desmin, S100, and HMB45. With these immunohistochemical findings, the mesenchymal component was evaluated as malignant fibrous histiocytoma. Also, the tumour was reported as carcinosarcoma of the skin because of the basal cell carcinoma and malignant fibrous histiocytoma association.

Carcinosarcoma of the skin occurs predominantly in elderly males (14). The most common sites involved are the distal extremities, head, and neck (7-9, 11, 12). Cutaneous carcinosarcoma has a better prognosis than the other sites (11). Local recurrence rate is 19%, visceral metastasis rate is 26%, and the 5-year disease-free survival rate is 50%. Tumours with an adnexal carcinoma have worse prognosis and, as expected, those with basal cell carcinoma have better prognosis than the others (8, 13). Osteosarcomatous and chondrosarcomatous components can be easily identifiable but different mesenchymal components can cause diagnostic problems. In order to avoid misdiagnosis,

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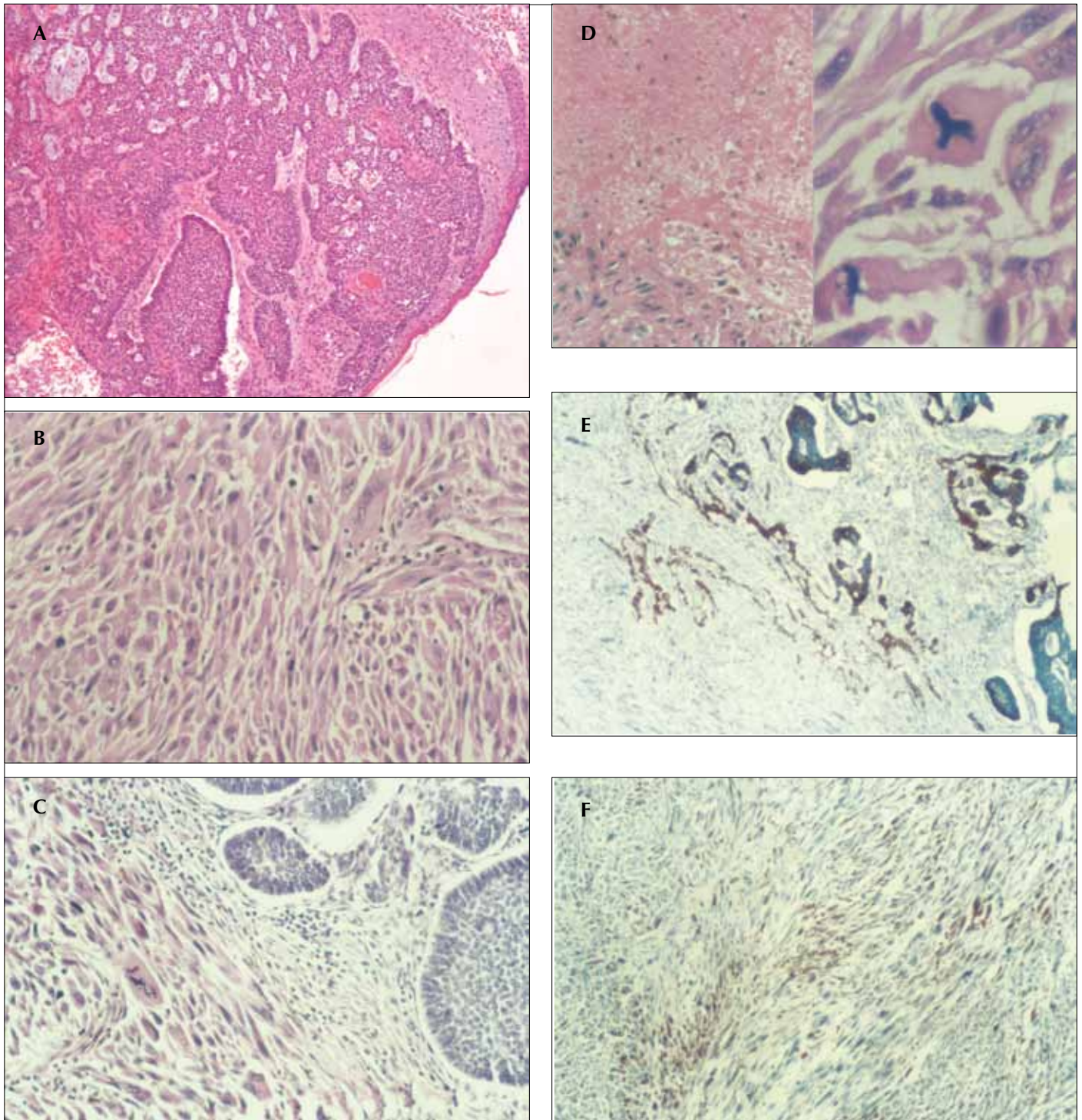
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**Figure 1.** (a) Typical basal cell carcinoma islands with palisading (HE x40), (b) spindle-shaped tumour cells with storiform pattern (HE x100), (c) histiocyte-like cells, and bizarre and multinucleated giant cells (HE x100), (d) (i) extensive necrosis (HE x100), (ii) atypical mitotic figure (b) (HE x400), (e) the epithelial component with cytokeratin (x40), (f) the mesenchymal component with CD68 (x100)

immunohistochemical studies should be performed, and ultrastructural studies may be needed (15).

Various mechanisms have been put forward for the pathogenesis of these tumours, such as *divergence*, *clonal evolution*, *combination*, *conversion*, and *collision theories*. Some of them are about monoclonality, but others postulate polyclonality. According to the *divergence*

(*monoclonal*) *theory*, the epithelial component undergoes metaplastic transformation in order to form the mesenchymal component of the tumour. This is the most widely accepted hypothesis and is supported by clinical, histopathological, immunohistochemical, ultrastructural, and tissue culture evidence (10). Patel has published four carcinosarcoma cases with osteosarcoma, leiomyosarcoma, rhabdomyosarcoma, and chondrosarcoma components, and came up with the idea that

the mesenchymal component arises from metaplastic transformation of malignant epithelial cells (11). Bigby et al. (10) reported p53 expression of equal intensity in both the epithelial and mesenchymal components. Carlson grafted human basal cell carcinoma cells into the subcutis of immunosuppressed mice and showed the development of the sarcomatous differentiation of the epithelial tumour cells (16). However, the *clonal evolution theory* postulates that a second genetic event occurring in the neoplastic epithelial cells gives rise to the development of another epithelial component or heterologous metaplastic elements, such as bone, cartilage, skeletal muscle, or smooth muscle (17). On the other hand, the *combination theory*, supported by the monoclonality of the tumour, holds that the carcinomatous and sarcomatous components arise from pluripotent stem cells (10). According to the *conversion theory that is founded on the carcinosarcomas of the female genital tract*, the mesenchymal component is pseudosarcomatous reactive reaction to the epithelial component (18). However, this theory has little support because the mesenchymal component is clearly malignant, and has the capacity to metastasise. The *collision theory* is about polyclonality and suggests that two independent tumours have collided (7). İzaki reported a case and proposed that these two components did not have common immunohistochemical or electron microscopic features. A pre-existing carcinomatous tumour stimulated the underlying stroma, giving rise to stromal proliferation and sarcomatous transformation (7).

In our case, the epithelial and mesenchymal components had different immunohistochemical features and there were transition focuses between the stroma of the basal cell carcinoma and malignant fibrous histiocytoma. We think our findings suggest that the malignant mesenchymal and epithelial components were different tumours and they did not share histogenetic mechanisms. Basal cell and squamous cell carcinomas are not rare tumours. In addition, they may be seen on sun-damaged skin with concomitant superficial malignant fibrous histiocytoma. This supports the *collision theory* (7). The patient's history about the rapid growth of the mass in the previous 2 months after a long duration also supports this idea.

In summary, primary cutaneous carcinosarcomas are rare tumours of unknown aetiology. These neoplasms have a high recurrence rate and metastatic potential. It is important to recognise the two malignant components of the tumour by thorough examination. In addition, immunohistochemical studies should be performed in order to confirm the diagnosis. The theories about the histogenesis may be right for some cases but not all the cases reported in the literature can be explained by these theories.

#### Conflict of Interest

No conflict of interest was declared by the authors.

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Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

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