

Carcinosarcoma of the Skin : A New Combination of Squamous Cell Carcinoma and Chondrosarcoma

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Carcinosarcoma is a rare malignant tumor composed of a mixture of neoplastic epithelial and mesenchymal elements, which has been reported in many organ systems, but its occurrence is very rare in the skin.

An 81-year-old female patient presented with a 1-year-history of a painful exophytic mass on the tip of the middle finger of the right hand involving the distal part of the nail bed. The mass was 1×1×0.6 cm in size, hard, flesh colored, ulcerated and easily bleeding. Microscopically, the epidermis at the margin of the tumor showed the characteristic findings of squamous cell carcinoma and the carcinomatous component was mixed with malignant mesenchymal components focally differentiating into chondrosarcoma. This combination of squamous cell carcinoma and chondrosarcoma has not been reported yet in the skin. She had a distal interphalanx amputation and no recurrence appeared for 1 year on follow-up.

(Ann Dermatol 10:(2) 81~85, 1998).

Key Words : Carcinosarcoma, Chondrosarcoma, Skin, Squamous cell carcinoma

Carcinosarcoma is a rare tumor composed of a mixture of malignant epithelial and mesenchymal elements. Development of this entity has been reported in many organ systems, including the uterus, ovary, breast, thyroid, esophagus, stomach, larynx, lung and urinary tract in order of frequency.¹ However this neoplasm has only very rarely been documented in the skin.² We describe a case of carcinosarcoma having features of both squamous cell carcinoma in the epithelial component and of chondrosarcoma in the mesenchymal component involving the nail bed. We also review the literature reported up to date regarding the carcinosarcoma of the skin.

CASE REPORT

A 81-year-old woman presented with a 1-year history of a painful exophytic mass on the tip of the middle finger of the right hand on March 8, 1996.

She had undergone a left hemicolectomy 1-year previously with ischemic colitis and visited this hospital periodically with hypertension and diabetes mellitus.

A physical examination revealed a 1×1×0.6 cm sized, hard, flesh colored, ulcerated mass with tenderness on palpation on the tip of the middle finger of the right hand (Fig.1). The mass separated the nail from the nail bed. Neither cervical nor axillary lymph nodes were palpable.

Routine laboratory examinations showed no abnormal findings. An X-ray revealed soft tissue swelling and a pathological fracture line on the distal phalanx (Fig.2).

The biopsied specimen revealed an admixture of basaloid cells forming small nests or cord structures and a malignant stromal component with

Received October 20, 1997.

Accepted for publication February 3, 1998.

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Table 1. Summary of reported cases of carcinosarcoma of the skin in the literature

Case	Author	Year reported	Sex/ Age	Site	Size (cm)	Histologic combination	
						Epithelial element	Mesenchymal element
1	Dawson ²	1972	M/75	Chest wall	10	BCC & SCC	Fibrosarcoma
2	Quay, et al ⁵	1981	M/74	Lt. axilla	8	BCC	Fibro-, Chondro-, Osteo-, & Synovial-sarcoma
3	Iakovides, et al ⁶	1988	M/73	Temple	N/A	SCC	MFH
4	Iakovides, et al ⁶	1988	M/54	Lt. leg	9 × 7 × 4	SCC	MFH
5	Tschen, et al ⁷	1988	F/91	Forehead	7	BCC	Osteosarcoma
6	Takasaki, et al ⁸	1989	F/89	Cheek	3 × 3.5 × 3	SCC	Fibrosarcoma
7	McKee, et al ⁹	1990	F/52	Lt. iliac crest	6	MES	Rhabdomyosarcoma
8	McKee, et al ⁹	1990	F/45	Lt. upper arm	1.0 × 1.0 × 0.5	MES	Osteosarcoma
9	Parham, et al ¹⁰	1991	F/54	Vulvar area	1.5	SCC	Osteosarcoma
10	Izaki, et al ¹¹	1993	M/44	Upper back	15 × 13 × 6.5	BCC	Spindle cell sarcoma like structure
11	Leen, et al ¹²	1995	M/86	Ant. to Rt. tragus	1.5 × 1.2 × 0.3	BCC	Osteosarcoma
12	Present case	1996	F/83	Finger tip	1.0 × 1.0 × 0.6	SCC	Chondrosarcoma

N/A, accurate information not available

MES, Malignant eccrine spiradenoma; MFH, Malignant fibrous histiocyoma

Fig. 1. Bean head sized, hard, flesh colored, ulcerated and bleeding mass on the tip of the middle finger of the right hand.

plump spindle cells in a myxoid and chondroid background. The pathological differential diagnosis was malignant chondroid syringoma, malignant

Fig. 2. X-ray revealed soft tissue swelling and pathological fracture line (←) on the distal phalanx.

peripheral nerve sheath tumor, synovial sarcoma and carcinosarcoma. She had the distal interphalangeal joint amputated.

Fig. 3. The tumor consists of a carcinomatous and sarcomatous component infiltrating to the bone (◄) and marrow space (Hematoxylin and eosin stain; original magnification $\times 40$).

The tumor, 1.2×1.0 cm in size, showed an ulcerated external surface and a pale gray to bluish myxoid cut surface macroscopically. Microscopically, the tumor consisted of carcinomatous and sarcomatous components infiltrating into the bone and marrow space (Fig. 3). The epithelial component was composed of nests or cords of basaloid cells and squamoid cells (Fig. 4). The squamoid cells formed small and large nests. They had abundant eosinophilic and clear cytoplasm, and hyperchromatic nuclei with moderate pleomorphism. Several atypical squamoid cells showed enlarged bizarre nuclei. The epidermis at the margin of the tumor showed a continuous change to dysplastic squamous cells and finally to squamous cell carcinoma. The carcinomatous component was closely associated with histologically malignant mesenchymal components which focally differentiated into chondrosarcoma (Fig. 4). The cartilaginous foci had multiple lacuna-appearing vacuolations, and were blended or surrounded with the undifferentiated round, oval or spindle-shaped cells. The individual cells in the cartilaginous foci possessed hyperchromatic nuclei of similar size and shape separated by large amounts of mucoid material. There were 11 to 12 mitoses per high power field in most cellular areas. An immunohistochemical study was carried out using a standard avidin-biotin complex immunoperoxidase technique. The epithelial cells in the tumor nests weakly demonstrated EMA- and cytokeratin- positive reactions and negative immunoreactions for vimentin, S-100 protein and

Fig. 4. The epithelial component, squamous cell carcinoma (*) is associated with a chondrosarcomatous area (★) (Hematoxylin and eosin stain; original magnification $\times 200$).

desmin. The mesenchymal component showed positive immunoreactivities for S-100 protein, desmin and vimentin. The final pathological diagnosis was carcinosarcoma.

DISCUSSION

The term carcinosarcoma was introduced by Virchow³ in 1864 to describe a tumor in which both malignant epithelial and malignant mesenchymal elements were identified. In 1938 Saphir and Vass¹ recorded 153 examples from all sites including 5 tumors of the skin, but none of which was acceptable as carcinosarcoma. The description of carcinosarcoma is applied to a wide range of neoplastic morphotypes including squamous carcinoma, adenocarcinoma, or transitional carcinoma in an admixture with clearly identifiable rhabdomyosarcoma, osteosarcoma, chondrosarcoma, or liposarcoma.⁴ The criteria for the diagnosis of carcinosarcoma in any location are twofold: 1) the lesion must contain malignant epithelial and mesenchymal components; 2) the arrangement of the epithelial and stromal components must be mainly in the form of an intimate ad-

mixture.⁵ Many locations have been reported to be the sites of this tumor including the female genital tract, breast, thyroid, larynx, GI tract and lung. Of these sites, the uterine corpus is the most frequently involved,⁴ but in the uterus, it is a relatively rare neoplasm. Most patients, not necessarily everyone, with carcinosarcomas are elderly and they typically have symptoms and signs suggesting relatively rapid growth of their tumors. The long clinical history, the large size of the tumor and the polypoid pattern of growth are considered features which strengthen the diagnosis.⁵

This tumor of the skin is rare, as only 11 cases of carcinosarcoma in the skin have been reported to our knowledge (table I). This tumor showed no sex predilection and affected relatively elderly patients between the ages of 44 and 91 years (mean 68 years) as those in other organs. The typical clinical history was a cutaneous huge mass that had been present for many years, although there were some patients with small tumors of short duration including this case. The duration ranged from 6 weeks to 13 years,^{5,9} and the phase of rapid growth was preceded by a quiescent or indolent phase in the cases with long history.^{2,7,9,11} It shows diversity in the sites involved, suggesting that this malignancy may occur at any site. Our patient was also an aged person, but the present tumor was much smaller than most of those previously described.

Histologically, the characteristic feature of carcinosarcoma in any location is the presence of heterogeneous malignant epithelial and stromal components. The eleven reported cases of carcinosarcoma of the skin showed various combinations of epithelial and mesenchymal elements. The epithelial component exhibited mainly basal cell carcinoma,^{2,5,7,11,12} squamous cell carcinoma,^{2,6,8,10} or malignant eccrine spiradenoma in two cases.⁹ The commonly encountered mesenchymal component was osteosarcoma.^{5,7,9,10,12} Chondrosarcoma,⁵ rarely, malignant fibrohistiocytoma,⁶ rhabdomyoblastoma,⁹ or fibrosarcoma^{2,5,8} were reported. The malignant epithelial component in the present case represented the characteristics of squamous cell carcinoma. The carcinomatous nests were haphazardly arranged in a fibrotic, focally myxoid stroma. The stroma which consisted of malignant spindle cells showed areas of homogenous chondroid material in which the individual cells possessed large hyperchromatic nuclei and a narrow rim of deeply

eosinophilic cytoplasm, characteristic features of chondroblasts. This area showed positive reactions for S-100 protein and desmin as expected. To our knowledge, this is the first account of an unusual combination of squamous cell carcinoma and chondrosarcoma, which was different from those reported as carcinosarcoma in the skin. The pathological differential diagnosis was that the tumors showed biphasic patterns including malignant chondroid syringoma, malignant peripheral nerve sheath tumor, synovial sarcoma and carcinosarcoma. The epithelial element formed small or large cysts in malignant chondroid syringoma. In biphasic synovial sarcoma, epithelial cells and fibroblast-like spindle cells coexist. Not infrequently a diagnosis of squamous cell carcinoma may also be suggested by focal squamous metaplasia. Calcification with or without chondroid changes is another characteristic feature, but it is morphologically benign. Malignant peripheral nerve sheath tumors resemble fibrosarcoma in its overall organization, and have islands of cartilage which are mature unlike this case. Squamous metaplasia was also reported in malignant peripheral nerve sheath tumors, but it was only one case.

Wide excision and/or radiotherapy was performed in these 11 cases. Carcinosarcoma at any site is often associated with a poor prognosis.^{9,13} But three of them showed no sign of local or distant recurrence at the time of the report.^{7,9,10} The fact that the eighth patient has survived without evidence of disease for 10 years is especially interesting. A couple of patients had local recurrence,^{6,10} one 6 months later and the other 3 years later. One patient¹¹ died 1 year later as a result of hemorrhage due to invasion by the neoplasm and metastasis developed in three cases.^{5,8,9} There was no clinical evidence of metastatic spread in our patient at presentation and she was well after one year of surgery without any signs of recurrence or metastasis. In view of the reported rate of recurrence and metastasis, carcinosarcoma of the skin may behave in a biologically malignant fashion.

In summary, we herein report a case of carcinosarcoma of the skin. The skin is an exceedingly rare site for such a neoplasm. Our case was found to be a carcinosarcoma characterized by a new combination of squamous cell carcinoma and chondrosarcoma involving the nail bed. The careful and close examination both clinically and histo-

logically facilitate the diagnosis of carcinosarcoma which is known as a rare malignancy in the skin.

REFERENCES

1. Saphir O, Vass A: Carcinosarcoma. *Am J Cancer* 33: 331-361, 1938
2. Dawson EK: Carcinosarcoma of the skin. *J R Coll Surg(Edinburgh)* 17: 242-246, 1972
3. Virchow R: *Die Krankhaften Geschwulste*. Hirschwas, Berlin, 1864, pp 180-182 cited from reference 5
4. Wick MR, Swanson PE: Carcinosarcomas: Current perspectives and an historical review of nosological concepts. *Sem Diagn Pathol* 10: 118-27, 1993
5. Quay Sc, Harrist TJ, Mihm MC Jr: Carcinosarcoma of the skin: case report and review. *J Cutan Pathol* 8: 241-246, 1981
6. Iakovides J, Delides GS: Carcinosarcoma of the skin-Report of two cases. *Arch Geschwulstforsch* 58: 461-464, 1988
7. Tschen JA, Goldberg LH, McGavran MH: Carcinosarcoma of the skin. *J Cutan Pathol* 15: 31-35, 1988
8. Takasaki S, Itami S, Fujiwara S, et al: Carcinosarcoma: report of a case (in Japanese). *J West Jpn Dermatol Soc* 51: 261-266, 1989
9. McKee PH, Fletcher CDM, Stavrinou P, et al: Carcinosarcoma arising in eccrine spiradenoma. A clinicopathologic and immunohistochemical study of two cases. *Am J Dermatopathol* 12: 335-43, 1990
10. Parham DM, Morton K, Robertson AJ, et al: The changing phenotypic appearance of a malignant vulval neoplasm containing both carcinomatous and sarcomatous elements. *Histopathology* 19: 263-268, 1991
11. Izaki S, Hirai A, Yoshizawa Y, et al: Carcinosarcoma of the skin: Immunohistochemical and electron microscopic observations. *J Cutan Pathol* 20: 272-278, 1993
12. Leen EJ, Saunders MP, Vollum DI, et al: Carcinosarcoma of skin. *Histopathology* 26: 367-371, 1995
13. Carson HJ, Tojo DP, Chow JM, et al: Carcinosarcoma of salivary glands with unusual stromal components: Report of two cases and review of the literature. *Oral Surg Oral Med Oral Pathol* 79: 738-746, 1995