CASE REPORT

Trichoblastic carcinosarcoma: an authentic cutaneous carcinosarcoma

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Accepted 3 May 2016

SUMMARY

We describe a trichoblastic carcinosarcoma on the forehead of a 65-year-old man. This is the fifth case of trichoblastic carcinosarcoma in the literature and the first to be reported from the USA. It was successfully treated with direct excision. Trichoblastic carcinosarcoma should be distinguished from epidermal carcinosarcoma. We believe trichoblastic carcinosarcoma is an authentic carcinosarcoma derived from multiple progenitor cells whereas epidermal carcinosarcomas are derived from a single progenitor cell that undergoes epithelial to mesenchymal transition, and are better considered as sarcomatoid carcinomas.

CASE PRESENTATION

A 65-year-old Caucasian man presented to a plastic surgery clinic reporting of a lesion on his forehead. The patient reported the lesion had been stable and unchanging for more than 10 years, but had begun to enlarge over the past year. Clinical examination revealed an indurated 0.6×0.8 cm erythematous nodule. The clinical impression was ruptured cyst. There was no relevant prior medical history. Physical examination was negative for regional lymphadenopathy. An excisional biopsy was submitted for histopathological evaluation.

INVESTIGATIONS

Macroscopic examination of the skin biopsy demonstrated an ill-circumscribed greyish-white mass in the dermis, measuring at least 1 cm in diameter. Microscopic examination revealed a biphasic neoplasm (figure 1). The major component of the neoplasm consisted of a proliferation of fusiform cells distributed in sheets and poorly defined fascicles with a widely infiltrative growth pattern. These cells exhibited conspicuous cellular atypia with pleomorphic, hyperchromatic nuclei and numerous mitotic division figures (5/high power field) consistent with a sarcoma. The minor component consisted of a deep dermal, circumscribed nodule of epithelial follicular germinative cells arranged in nests, strands and cribriform groups, with peripheral palisading. This component also exhibited cellular atypia including high nuclear/cytoplasmic ratios, crowded overlapping and hyperchromatic nuclei, and frequent mitotic division figures consistent with a carcinoma. The epithelial component was surrounded by trichogenic-specific stroma, which merged imperceptibly with the malignant spindle cell elements. Foci of pre-existent trichoblastoma were identified within the epithelial component of the tumour.

Immunostaining revealed strong CD10 expression by the pleomorphic fusiform cells, with less intense staining of the trichogenic-specific stroma and the peripheral epithelial cell layer (figure 2). The carcinomatous component exhibited strong keratin 5/6, p63 and high-molecular weight keratin signal. Both cell populations were non-reactive for S-100 protein, keratin 20, epithelial membrane antigen, MART-1, SOX10 and CD34. We diagnosed the tumour as a trichoblastic carcinosarcoma.

TREATMENT

The patient was treated with surgical excision of the biopsy site with 5 mm margins. Microscopic residual sarcomatous elements of trichoblastic carcinosarcoma were identified in the excision. The final surgical margins were negative for neoplasm.

OUTCOME AND FOLLOW-UP

The patient was free of disease 4 months post-surgical excision. He is currently asymptomatic and being managed with clinical observation. Imaging studies were deferred.

DISCUSSION

Carcinosarcomas are neoplasms comprised of malignant epithelial and mesenchymal cells. Most cutaneous carcinosarcomas are epidermal carcinosarcomas and consist of basal cell carcinoma or squamous cell carcinoma admixed with undifferentiated pleomorphic sarcoma.¹ Cutaneous adnexal carcinosarcomas are rare. We report the first case of trichoblastic carcinosarcoma from the USA. Trichoblastic carcinosarcoma is a biphenotypic malignant neoplasm comprised of malignant follicular germinative cells surrounded by malignant hair follicle-specific mesenchymal cells. Malignant mesenchymal cells tend to predominate. The aim of this report is to spread awareness of this entity and compare it to the more common epidermal carcinosarcomas.

There are four previously reported cases in the literature (table 1). Including our case, the mean patient age was 74 years, the male to female ratio was 4:1, and 3/5 (60%) occurred on the head.^{2–5} These findings contrast with other adnexal carcinosarcomas, which are seen in a younger age group, and have a female preponderance.⁶ Including this case, three of the five reported cases described sudden enlargement of a long-standing lesion and found elements of pre-existent trichoblastoma. The other two case reports neither described the rate of onset nor noted evidence of a pre-existent trichoblastoma.

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To cite: Colston J, Hodge K, Fraga GR. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-214977

Figure 1 (A) Scanning magnification demonstrates a biphasic skin tumour (H&E, ×20). (B) The tumour is comprised of a circumscribed nodule of basophilic follicular germinative cells surrounded by spindled cells in an eosinophilic fibroinflammatory matrix (H&E, ×100). (C) The follicular germinative cells exhibit focal crowding and aberrant mitotic activity (H&E, ×400). (D) The spindled cells exhibit pleomorphism and infiltrative growth pattern around an eccrine gland (H&E, ×400).

The rarity of trichoblastic carcinosarcoma makes it difficult to establish standards for clinical management. Complete surgical excision was curative in three of the four cases, where follow-up information is available.^{4 5} One case was associated with lymph node metastasis, tumour recurrence and eventual death from metastatic disease in a patient with comorbid B-cell chronic lymphocytic leukaemia.² Immunodeficiency from leukaemia may have contributed to the aggressive behaviour of the malignancy in this case. We believe imaging studies to exclude metastatic disease should be considered if the patient is immunocompromised. Otherwise, we suggest complete surgical excision combined with physical examination for lymphadenopathy, hepatosplenomegaly and abnormalities on review of systems as an initial management strategy. Subsequent clinical

observation at regular intervals appropriate for a high-risk non-melanoma skin cancer would be prudent.

Biphasic tumours can be produced by juxtaposition of two separate tumours (collision tumour), divergent differentiation of a single progenitor cell into malignant epithelial and mesenchymal populations (monoclonal hypothesis), and origin from two separate progenitor cells (multiclonal hypothesis). Tran *et al*⁶ suggested cutaneous carcinosarcomas could be subclassified into high-risk adnexal types and low-risk epidermal types. Paniz-Mondolfi *et al*⁷ found evidence of a common cell of origin in epidermal carcinosarcomas. Genetic studies on adnexal carcinosarcomas have not been performed. We believe trichoblastic carcinosarcomas arise from multiple progenitor cells because the precursor lesion has separate epithelial and

Figure 2 (A) The follicular germinative cells express high-molecular weight keratin (34βE-12, ×400). (B) The trichogenic specific stroma around the follicular germinative cells is weakly positive for CD10; the pleomorphic spindled cells are strongly positive for CD10 (CD10, ×400).

Table 1 Trichoblastic carcinosarcomas in the literature

Reference (year)	Sex/age	Tumour location	Pre-existent trichoblastoma	Follow-up (duration)
Regauer <i>et al</i> ² (2000)	M/55	Right deltoid region	Yes	Death from metastatic disease (21 months)
Liegl <i>et al</i> ^{3*} (2004)	F/79	Right cheek	Yes	None provided
Kazakov <i>et al</i> ⁴ (2004)	M/80	Sacrum	Not identified	No evidence of disease (7 months)
Kazakov <i>et al</i> ⁵ (2008)	M/92	Left ear	Not identified	No evidence of disease (6 months)
Our case (2016)	M/65	Forehead	Yes	No evidence of disease, (4 months)

*Liegl *et al* also present the case originally described by Regauer *et al* as part of a case series.

mesenchymal elements, and malignant trichoblastomas with solely carcinomatous or solely sarcomatous change have been described. This suggests trichoblastic carcinosarcoma is an authentic carcinosarcoma derived from two progenitor cells. Epidermal carcinosarcomas are derived from a single progenitor cell that undergoes epithelial to mesenchymal transition, and are better classified as sarcomatoid carcinomas.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Learning points

- ▶ Trichoblastic carcinosarcoma is comprised of malignant follicular germinative cells surrounded by malignant hair follicle-specific mesenchymal cells.
- ▶ Trichoblastic carcinosarcoma has a predilection for sun-damaged skin of elderly men.
- ▶ Some cases are associated with pre-existent trichoblastoma.
- ▶ Metastasis and death have been described in an immunocompromised patient.
- ▶ We believe trichoblastic carcinosarcoma is an authentic carcinosarcoma, whereas so-called epidermal carcinosarcoma is better considered a sarcomatoid carcinoma.

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