Primary Carcinosarcoma of the Skin

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Abstract Primary cutaneous carcinosarcoma is a biphasic tumour containing both malignant epithelial and malignant mesenchymal elements. The disease is a rare malignant skin tumour of unknown aetiology with significant potential to recur locally and to metastasise. Surgery is the primary therapeutic modality. We describe an unusual case of primary cutaneous Carcinosarcoma localised in the hand.

Keywords Skin neoplasms · Cutaneous carcinosarcoma

Introduction

Carcinosarcoma is a biphasic tumor composed of intimately admixed epithelial and mesenchymal elements, both of which are malignant. It occurs in a variety of organs, including female and male genital tracts, lung, kidney, upper and lower urinary systems, breast, oronasopharynx, larynx, gastrointestinal tract, thyroid, thymus, and head and neck [1–4]. It is extremely rare as a primary tumor in the skin. The prognosis of these tumors remains unclear because the number of reported cases is small, often with limited follow-up. The histogenesis remains controversial, although various hypotheses have been suggested. We describe an unusual case of

primary cutaneous carcinosarcoma that consisted of squamous cell carcinoma and liposarcoma.

Case Report

A 58-year-old woman presented with a 8 cm ulcerated exophytic nodule on her right hand (Fig. 1). The lesion was nontender with overlying erythema. The patient reported that the lesion began as a small red papule 6 months earlier and had rapidly grown in size. No history of trauma in the area could be elicited. No regional lymph nodes were found to be enlarged on clinical examination. Histopathologic diagnose of biopsy of the lesion was squamous cell carcinoma of the skin. The magnetic resonance imaging showed an interdigital extension of the lesion (Fig. 2). Chest radiograph and computed tomography scan of the abdomen were normal. An amputation of the hand was carried out.

Microscopic examination of the lesion revealed a tumor with two closely intermingled components. The epithelial tumoral component, arranged in cords and cellular tablecloths nests. The sarcomatous portion showed fascicular arrangement of atypical spindle cells with atypical lipoblasts. The tumoral stroma displayed mixed inflammatory infiltrate and variable vascular structures. Immunohistochemical analysis showed the cellular areas with atypical cells and frequent mitoses to be positive for keratin (AE1/AE3) and mdm2.

The patient has had no evidence of disease 7 months after the treatment.

Discussion

By definition, carcinosarcoma is regarded as a biphasic tumor composed of intimately admixed epithelial and mesenchymal

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Fig. 1 Macroscopic aspect of the tumour showed an ulcerated exophytic nodule on the right hand

components, both of which must be malignant. It should be distinguished from metaplastic carcinoma, pseudosarcoma, spindle cell carcinoma, and sarcomatoid carcinoma [4].

Several theories of carcinosarcoma pathogenesis have been proposed [5]:

- _ The collision of two synchronous but otherwise unrelated tumours.
- _ The combination of two divergent cell lines from a single stem cell.
- _ The conversion from carcinoma to carcinosarcoma by metaplastic transformation of part of the carcinoma into a sarcoma.

The disease is most likely to occur in the eighth and ninth decades but the average age at presentation (70, 4 years) was skewed by a wide range of ages (minimum 36 to maximum 93). Insufficient data have been reported to draw conclusions on predisposing factors but the history of previous skin tumours and the anatomical distribution of lesions suggest that sun exposure may be a factor. It affects men and women with equal frequency.

The majority of these neoplasms present as a polypoid or exophytic mass of variable color, often with overlying ulceration and bleeding secondary to trauma. the polypoid morphology results from the sarcomatous component. Although subjective, the duration of these tumors before clinical presentation ranges from several weeks to 30 years,

with many patients reporting a recent period of accelerated growth [4].

The differential diagnosis includes the more common spindle cell squamous carcinoma but immunohistochemistry may help as the spindle cells are usually positive for keratin markers. In addition, there tends to be a gradual transition between the epithelial and spindle cell elements. It is possible to see pseudoepitheliomatous hyperplasia overlying some tumors such as atypical fibroxanthoma and malignant melanoma but the keratinocytes are less atypical and, again, immunohistochemistry may clarify the diagnosis. Malignant mixed tumor of the skin may have a stroma with osteoid formation; however, this is usually benign in appearance. They demonstrate eccrine or apocrine gland differentiation, which was not identified in our cases, and very rarely focal follicular differentiation.

Surgery is the primary therapeutic modality for carcinosarcoma. All the reported patients underwent excisional surgery. Surgical margins are not reported in the literature. At our patient, an amputation of the hand was carried out because she has the interdigital extension of the lesion. The nodes clearance is indicated in the event of nodal disease [6].

Adjuvant radiotherapy can be administered to patients with recurrent disease or after partial excision but it has not demonstrated significant benefit [6].

The prognosis of carcinosarcoma of the skin is better than that of visceral areas, perhaps because of the easily detectable nature of cutaneous growth, which may prompt early diagnosis and treatment. However, long-term follow-up is needed [4]. The long-term prognosis of these tumors remains unclear. The number of reported cases is small, often with limited follow-up, and many studies group all tumors with different epithelial components together. This makes it difficult to establish the behaviour of individual tumor types [7].

Fig. 2 The magnetic resonance imaging showed an interdigital extension of the lesion

References

1. Wick MR, Swanson PE (1993) Carcinosarcomas: current perspectives and an historical review of nosological concepts. *Semin Diagn Pathol* 10:118–127
2. Biernat W et al (1996) Carcinosarcoma of the skin: case report and literature review. *Am J Dermatopathol* 18:614–619
3. Patel NK et al (1997) Primary metaplastic carcinoma (carcinosarcoma) of the skin: a clinicopathologic study of four cases and review of the literature. *Am J Dermatopathol* 19:363–372
4. Brown TJ, Tischen JA (1999) Primary carcinosarcoma of the skin: report of a case and review of the literature. *Dermatol Surg* 25:498–500
5. McCluggage WG (2002) Malignant biphasic uterine tumours: carcinosarcomes or metaplastic carcinomas? *J Clin Pathol* 55:321–325
6. Syme-Grant J et al (2006) Are primary cutaneous carcinosarcomas underdiagnosed? Five cases and a review of the literature. *J Plast Reconstr Aesthet Surg* 59:1402–1408
7. Rose et al (2008) Basal cell carcinoma with a sarcomatous component (carcinosarcoma): a series of 5 cases and a review of the literature. *J Am Acad Dermatol* 59:627–32