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Primary Cutaneous Carcinosarcoma: A Rare Case Report with Review of Literature

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Abstract: ***Primary cutaneous carcinosarcoma is a rare and aggressive biphasic malignant neoplasm that exhibits both epithelial and mesenchymal components. This malignancy is more commonly described arising from organs such as the uterus, breast, bladder, and lung, and is rarely seen on the skin. Here we report a rare case of primary cutaneous carcinosarcoma presenting on the skin of forearm of a 62-year-old male with intermingled chondrosarcoma and poorly differentiated carcinoma. Relevant literature is reviewed.***

Keywords: carcinosarcoma, skin, primary

1. Introduction

Carcinosarcomas are rare malignant biphasic tumours that contain intermingled carcinomatous and sarcomatous elements. Primary cutaneous carcinosarcomas (PCCS) are exceedingly rare.[1,2]

Dawson[3]described the first cutaneous carcinosarcoma in 1972. Since then approximately 120 cases have been reported in the literature[4] Cutaneous carcinosarcoma typically arises on sun-damaged skin as a nodular lesion and often showing ulceration. The most commonly affected sites include the face and scalp.[5] In a series of cases reported by Kwak et al [4] most common site of cutaneous carcinosarcoma was the head and neck, followed by the lower extremities, and penis.They are also reported in upper extremities.[6] We report one such rare occurence of cutaneous carcinosarcoma in the forearm of a 62 year old male.

2. Case Report

A 62 year old patient presented with a swelling in the left forearm just below the elbow joint. Patient noticed a painless swelling in the left forearm since 5 months which rapidly increased to the present size. There was no history of trauma. But patient complained of rapid loss of weight since 5 months.On examination, there was a spherical mass measuring 4x4 cms in size, hard in consistency and mobile . Overlying skin was stretched, shiny and thin capillaries were visible on the surface. MRI revealed well defined a well defined exophytic round to oval mass with rim enhancement in the posterior compartment of forearm (Figure 1) displacing surrounding muscles. There was no infiltration in to muscles or underlying radia/ulna or nearby elbow joint. The mass was clinically and radiologically diagnosed as Malignant mesenchymal tumor. The mass was subsequently excised and sent to us for histopathological examination. On gross examination the mass was well circumscribed, solid with grey white cut surface showing many mucoid/ cystic areas. Overlying skin appeared normal. On microscopic examination the tumor predominantly comprised of high grade chondrosarcoma showing hypercellularity, pleomorphic chondrocytes with irregular hyperchromatic nuclei, tumor giant cells, frequent mitotic figures embedded in a chondroid stroma. There was a minor component of poorly differentiated carcinoma comprising of nests,cords and isolated tumor cells diffusely infiltrating the fibrous stroma. These tumor cells were showing rounded vesicular nuclei,prominent nucleoli and scanty eosinophilic cytoplasm with frequent mitotic figures.(Figure2). Immunohistochemically the carcinomatous component was positive for AE1/AE3(Figure 3).However tumor cells were negative for EMA, p63 and Cytokeratin 18. The case was diagnosed as Primary cutaneous carcinosarcoma.

3. Discussion

Primary cutaneous carcinosarcoma is an aggressive biphasic tumor with overt carcinomatous and sarcomatous features. Müller et al[7]proposed 3 diagnostic criteria for primary cutaneous CS:

(1) a bimodal neoplasm consisting of both malignant epithelial and mesenchymal components confirmed by histological examination and IHC (2) exclusion of distant metastasis and true collision neoplasm defined as coexistence of 2 different tumors in the same specimen, and (3) a solid coherent neoplasm with exclusion of reactive stromal changes.

The epithelial component is most commonly a basal-cell carcinoma or squamous-cell carcinoma, but it can also be associated with adnexal-derived tumors including spiradenocarcinoma, porocarcinoma, proliferating trichilemmal cystic carcinoma, and metrical carcinoma. The mesenchymal component may be of osseous, cartilaginous or, more rarely, skeletal or smooth-muscle lineage. In most of the reported cases, the epithelial component has shown positivity for cytokeratin markers including CK 5/6, MNF116, 34βE12, and AE1/AE3.[7-10] In PCCS with basaloid epithelial component (basal cell carcinosarcomas), markers such as BerEP4 and Bcl-2 have also shown positivity.[9] The mesenchymal part can be composed of osteosarcoma, neurogenic sarcoma, rhabdomyosarcoma, chondrosarcoma, and malignant fibrous histiocytoma.[7-9] The mesenchymal component has been reported to be positive for vimentin, SMA, desmin, and neuron-specific enolase according to mesenchymal differentiation.[7-9] It has been reported that p53 is coexpressed by both epithelial and mesenchymal components.[9] Recently, p63 was proposed to be a useful adjunct tool for the diagnosis of PCCS.[7,9] Epithelial component in the present case was positive for AE1/AE3 and and mesenchymal component showed clear cut chondrosarcoma.

As differential diagnoses, spindle cell neoplasm (including spindle cell squamous cell carcinoma [SpSCC], spindle cell

melanoma [SCM], and atypical fibroxanthoma) and biphasic neoplasms (including biphasic synovial sarcoma and malignant mixed tumor) should be considered. The 3 diagnostic criteria mentioned above and results of IHC are helpful for diagnosis of PCCS.

Several hypotheses have been proposed for the histogenesis of carcinosarcoma, based largely on the pathology of the disease[10,11]; first, the collision tumor hypothesis, which proposes the collision of separate neoplasms at the same site resulting in a single neoplasm, based on the fact that skin cancers and malignant fibrous histiocytomas are commonly present in patients with actinically damaged skin; second, the composition hypothesis, which proposes that the mesenchymal component stands for a pseudosarcomatous reaction to the epithelial malignancy; third, the combination hypothesis, which proposes that both epithelial and mesenchymal elements of the tumor originate from a common pluripotential stem cell that undergoes divergent differentiation; and fourth, the conversion/divergence hypothesis, which argues that the sarcomatous component of the tumor represents metaplastic sarcomatous transformation of the epithelial component.[10-13]Recent immunohistochemical, molecular genetics, and ultrastructural studies suggest and favor the concept of monoclonality in carcinosarcoma from various sites, including uterus, gastrointestinal tract, lung, breast, and bladder.[10,11,14]In addition, identical p53 and Kirsten rat sarcoma 2 viral oncogene homolog mutations have been demonstrated in both epithelial and mesenchymal elements of carcinosarcoma, suggesting an early alteration in the histogenesis of the tumor with degeneration of the epithelial element into the sarcomatous element[12]

Treatment of PCC is predominantly surgical with wide local excision or, as in the case of our patient, wide excision of the mass was done. defect created was repaired with split skin graft taken from lateral aspect of left arm. Adjuvant radiotherapy is not currently recommended.[15] Regular clinical follow-up is paramount. Cutaneous carcinosarcomas typically have a better prognosis than carcinosarcomas arising in visceral organs, but nonetheless these tumors can be aggressive. Prognosis seems to be most closely linked to the origin of the epithelial component. One meta-analysis found that PCCs containing a basal- or squamous-cell carcinoma had a fiveyear survival rate of 70%.[16] Conversely, PCCs with an epithelial element of adnexal origin have a poorer prognosis, with a 25% five-year disease-free survival rate.5 Other poor prognostic factors include age younger than 65, tumor size greater than 2 cm, a recent growth pattern, longer duration of existing skin tumor, and metastasis to lymph nodes.[16,17]Even after surgical excision, 7% to 19% of PCCs recur.[18,19] Diagnosis and treatment is necessary, with locoregional and distant metastases documented in 19% and 26% of cases, respectively.[18] PCC can also be fatal, with one report documenting PCC with cerebral metastases resulting in death.[20]

References

[1] Berg JO, Tei TM. Primary cutaneous carcinosarcoma: Review and presentation of a case with emphasis on clinical characteristics and management.European Journal of Plastic Surgery 2005;28:99-104.
[2] El Harroudi T, Ech-Charif S, Amrani M, Jalil A. Primary carcinosarcoma of the skin. J Hand Microsurg 2010;2:79-81.
[3] Dawson EK. Carcino-sarcoma of the skin. J R Coll Surg Edinb 1972;17:243-6.
[4] Kwak HB , Park J , Kim HU , Nam KH, Yun SK. Clinicopathologic Analysis of Cutaneous Carcinosarcoma Cases. J Korean Med Sci. 2019;34:1-10.
[5] Calonje E, Brenn T, Lazar A, Mckee PH. Tumors of the surface epithelium, In: Calonje E, Brenn T, Lazar A, Mckee PH, editors. McKee's Pathology of the Skin. 4th ed. Oxford: Elsevier Saunders; 2012, 1139-40.
[6] Bellew S, Del Rosso JQ, Mobini N. Primary carcinosarcoma of the ear. J Clin Anesthet Dermatol. 2009;2:33-35.
[7] Müller CS, Pfohler C, Schiekofer C, Korner R, Vogt T. Primary cutaneous carcinosarcomas: a morphological histogenetic concept revisited. Am J Dermatopathol. 2014;36:328-339.
[8] Clark JJ., Bowen AR., Bowen GM. Cutaneous carcinosarcoma: a series of six cases and a review of the literature. J Cutan Pathol 2017;44:34-44.
[9] Zbacnik A P, Rawal A, Lee B, Werling R, Knapp D, Mesa H. Cutaneous basal cell carcinosarcoma: case report and literature review. J Cutan Pathol 2015;42:903-910.
[10] Bigby SM, Charlton A, Miller MV, Zwi LJ, Oliver GF. Biphasic sarcomatoid basal cell carcinoma (carcinosarcoma): four cases with immunohistochemistry and review of the literature. J Cutan Pathol 2005;32:141-147.
[11] Zidar N, Gale N. Carcinosarcoma and spindle cell carcinoma--monoclonal neoplasms undergoing epithelial-mesenchymal transition. Virchows Arch 2015;466(3):357-8.
[12] Loh TL, Tomlinson J, Chin R, Eslick GD. Cutaneous carcinosarcoma with metastasis to the parotid gland. Case Rep Otolaryngol 2014;2014:173235.
[13] Dadzie OE, Mahalingam M. Pearls and Pitfalls in Neoplastic Dermatopathology. Cambridge: Cambridge Medicine; 2016.
[14] Thompson L, Chang B, Barsky SH. Monoclonal origins of malignant mixed tumors (carcinosarcomas). Evidence for a divergent histogenesis. Am J Surg Pathol 1996;20(3):277-85.
[15] Syme-Grant J, Syme-Grant NJ, Motta L, et al. Are primary cutaneous carcinosarcomas underdiagnosed? Five cases and a review of the literature. J Plas Reconstr Aesthet Surg 2006;59:1402-1408.
[16] Tran TA, Muller S, Chaudahri PJ, et al. Cutaneous carcinosarcoma. Adnexal vs epidermal types define high and low-risk tumors. Results of a meta-analysis. J Cutan Pathol. 2005;32:2-11.
[17] Hong SH, Hong S-J, Lee Y, Kang E-Y. Primary cutaneous carcinosarcoma of the shoulder: case report and literature review. Dermatol Surg 2013;39:338-40.
[18] Kwan JM, Satter EK. Carcinosarcoma: a primary cutaneous tumor with biphasic differentiation. Cutis 2013;92:247-249.
[19] Brasanac D, Boricic I, Todorovic V, Tomanovic N. Primary cutaneous carcinosarcoma: case report with expanded immunohistochemical analysis. Int J Dermatol 2008;47:496-501.

[20] Chittari K, Birnie AJ, Kulkarni KR, et al. Sarcomatoid carcinoma of the hand: a clinical case with an aggressive and uncommon presentation. Clin Exp Dermatol 2012;37:505-8.

Legends

Figure 1: MRI revealing a well defined exophytic round to oval mass with rim enhancement in the posterior compartment of forearm

Figure 2: Photomicrograph revealing both chondrosarcoma and poorly differentiated carcinoma components(H&E, 40x)

Figure 3: AE1/AE3 positivity in carcinomatous cells(IHC, 40x)

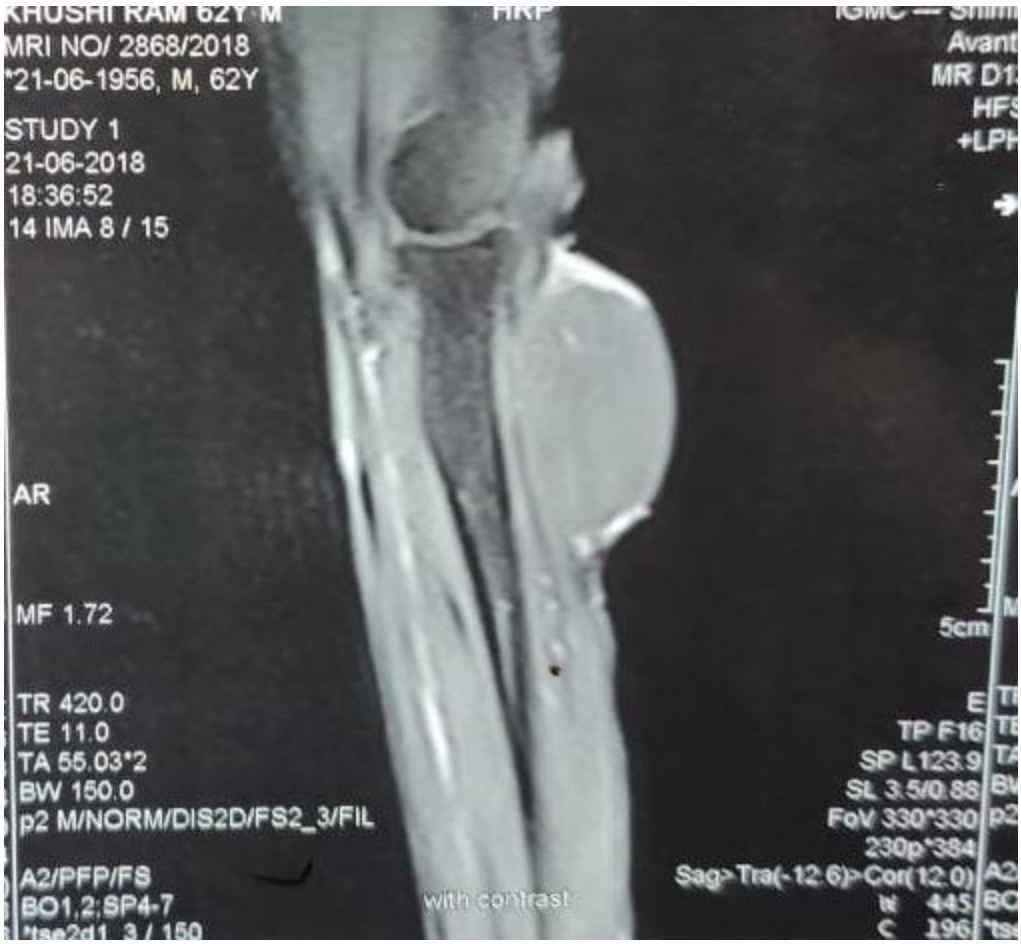

Figure 1

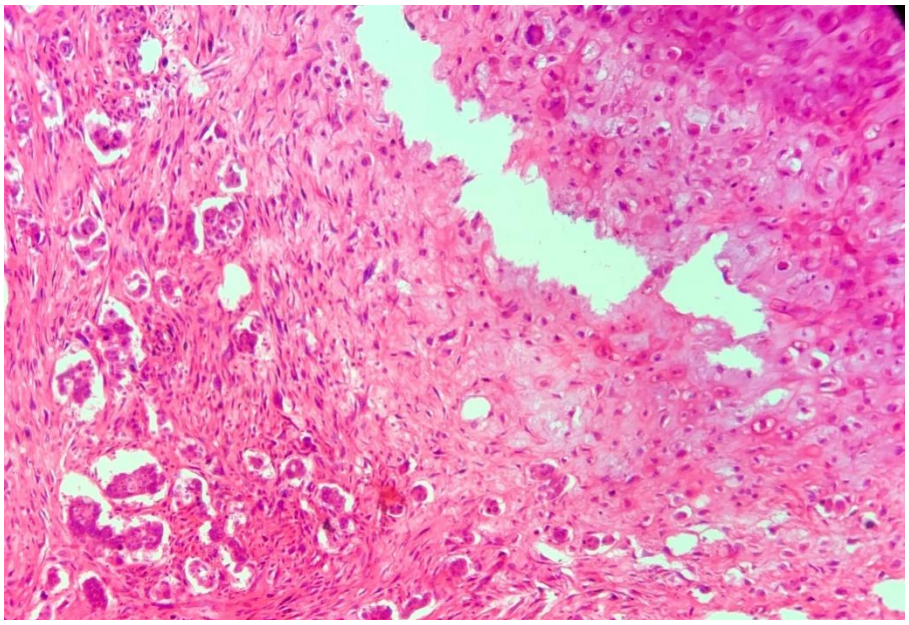

Figure 2

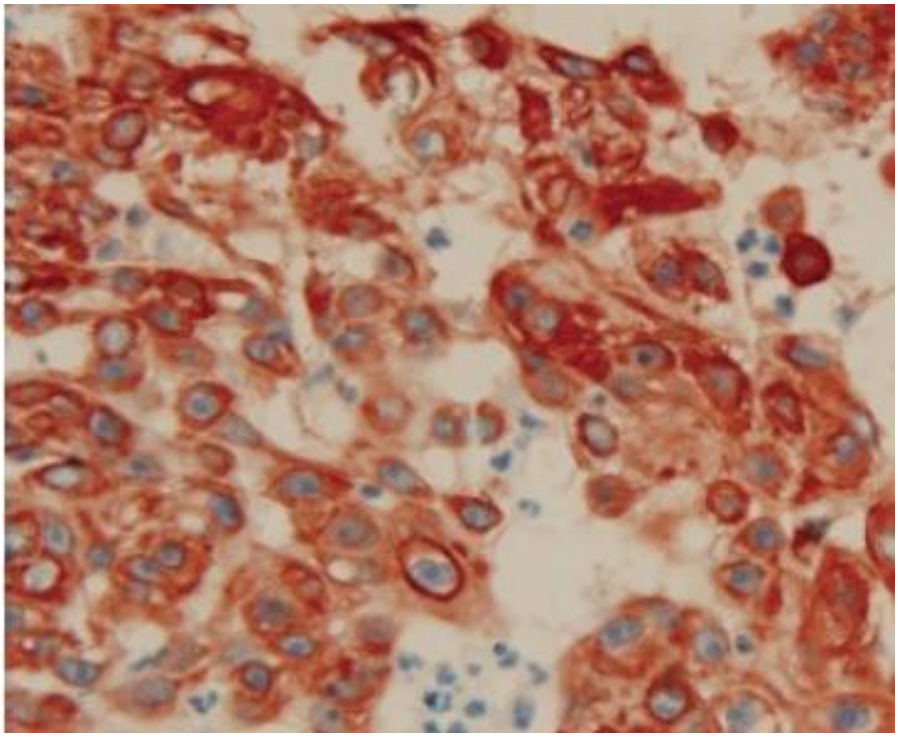

Figure 3