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INTERESTING MEDICAL IMAGE

Primary Cutaneous Carcinosarcoma

A cutaneous neoplasm with an exceptional presentation

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ساركومة سرطانية جلدية أولية

ورم جلدي في تَجَلِّي استثنائي

وريكاردو رويز-فيلا فيردي و هوزي انيريوس فيرناندس

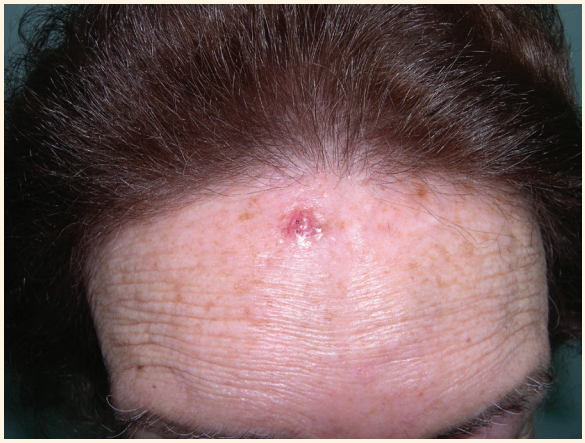

Figure 1: Photograph of an excrescent ulcerated tumour on the upper forehead of an 85-year-old woman.

An 85-year-old woman with a history of hypertension and diabetes mellitus was referred to the Dermatology Outpatient Clinic of the Complejo Hospitalario de Granada, Granada, Spain, in 2016 with an excrescent tumour on her upper forehead measuring 8 mm in diameter that had been present for the past two years [Figure 1]. She had been exposed to significant ultraviolet (UV) radiation and undergone excision of two basal cell carcinomas (BCCs) 10 years prior. Upon physical examination, the lymph nodes were not palpable and there was no evidence of hepatosplenomegaly.

A blood cell count and general biochemistry tests showed no abnormalities. The tumour was removed via complete conventional surgical excision with a 1 cm margin. Upon histological examination, the neoplasia consisted of double cellular components. The first comprised typical BCC cells, while the other was composed of short spindle cells with moderate pleomorphism and pronounced mitotic activity corresponding to epithelial and sarcomatous components, respectively [Figure 2]. An immunohistochemical analysis revealed the BCC component to be cytokeratin (CK) AE1/AE3-positive and vimentin-negative, while the sarcomatous component was CK-negative and vimentin-positive [Figure 3]. At a follow-up appointment one year later, there was no evidence of recurrence of the lesion.

Comment

Visceral carcinosarcomas have been described in numerous locations, including the adrenal glands, breast tissue, colon, endometrium, lungs and urogenital tract.[1] First described in 1953, cutaneous carcinosarcomas (CCSs) are uncommon cutaneous neoplasms involving biphasic malignant epithelial and sarcomatous components.[2] The epithelial component is represented by a BCC, squamous cell carcinoma or adnexal carcinoma of the skin such as a porocarcinoma, trichilemmal cystic carcinoma or spiradenocarcinoma, whereas the

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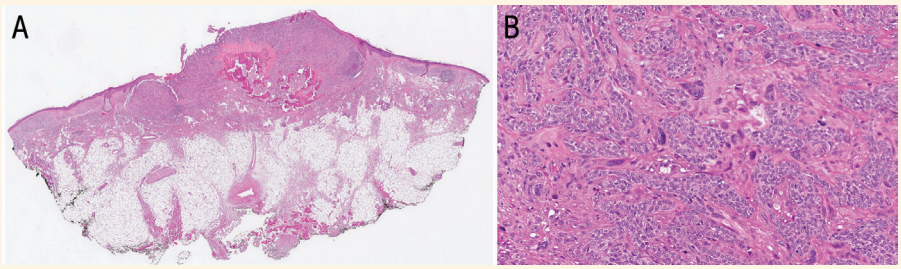

Figure 2: Haematoxylin and eosin stains at (**A**) x2 magnification showing the panoramic pathological view of the lesion and (**B**) x10 magnification showing a biphasic tumour with an epithelial component (atypical basaloid proliferation) and a sarcomatous component (pleomorphic spindle cells). No vascular or perineural invasion was detected.

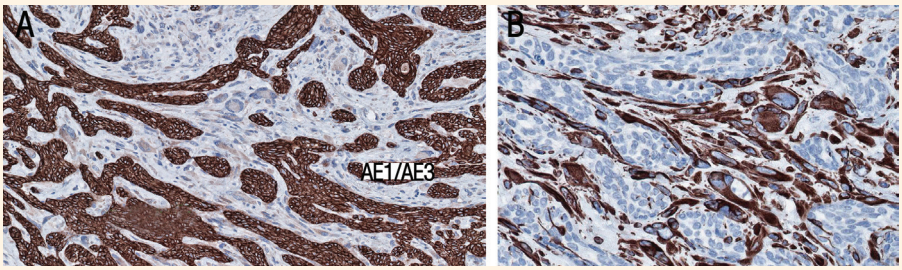

Figure 3: Immunohistochemistry stains at x20 magnification showing (**A**) cytokeratin AE1/AE3-positivity in the epithelial component and (**B**) vimentin-positivity in the sarcomatous component of the lesion.

sarcomatous component is formed of an osteosarcoma, chondrosarcoma, fibrosarcoma or malignant fibrous histiocytoma.[3]

The diagnostic criteria for a CCS include an absence of transitional areas in the malignant components and lack of cytokeratin expression in the sarcomatous components.[3] However, Müller's criteria should be additionally considered to more thoroughly confirm the diagnosis.[4] According to Müller *et al.*, a CCS is a malignant entity composed of two well-defined cell populations as evidenced by conventional histology (i.e. haematoxylin and eosin stains) and an adequate immunohistochemical panel. Metastases from distant sites and sarcomatous changes in the *stroma* of the neoplasm should also be excluded.[4]

There are at least four aetiopathogenic theories to explain the genesis of CCS. Of these, the monoclonal theory is particularly important as it justifies the rapid growth of the lesion by implicating the metaplastic transformation of the sarcomatous component.[5] Other theories to explain the pathogenesis of this variety of tumour are polyclonal-, collision- and composition-based.[4] From a clinical perspective, CCSs typically occur in damaged and sun-exposed skin. In the current case, the presence of p53 mutations in the two components of the neoplasm confirms the role of UV radiation as a triggering factor.[5]

In CCS cases, Mohs micrographic surgery is the treatment of choice and has been reported to result in a cure rate of ≥98%; in contrast, a recurrence rate of 33% has been reported without this surgery.[4] As such, close observation of the patient is required if Mohs surgery is not performed. There is no evidence to support the use of adjuvant radiotherapy. The prevalence of metastasis in CCS with a basal cell epithelial component has been reported in 2% of cases.[4,5]

References

1. Rose RF, Merchant W, Stables GI, Lyon CL, Platt A. Basal cell carcinoma with a sarcomatous component (carcinosarcoma): A series of 5 cases and a review of the literature. J Am Acad Dermatol 2008; 59:627–32. doi: 10.1016/j.jaad.2008.05.035.
2. Ormea F. [Carcinosarcomas and cutaneous pseudosarcomas]. Minerva Dermatol 1953; 28:153–9.
3. Tran TA, Muller S, Chaudahri PJ, Carlson JA. Cutaneous carcinosarcoma: Adnexal vs. epidermal types define high- and low-risk tumors - Results of a meta-analysis. J Cutan Pathol 2005; 32:2–11. doi: 10.1111/j.0303-6987.2005.00260.x.
4. Müller CS, Pföhler C, Schiekofer C, Kömer R, Vogt T. Primary cutaneous carcinosarcomas: A morphological histogenetic con cept revisited. Am J Dermatopathol 2014; 36:328–39. doi: 10.1097/DAD.0b013e318297cc34.
5. Clark JJ, Bowen AR, Bowen GM, Hyngstrom JR, Hadley ML, Duffy K, et al. Cutaneous carcinosarcoma: A series of six cases and a review of the literature. J Cutan Pathol 2017; 44:34–44. doi: 10.1111/cup.12843.