Pleomorphic dermal sarcoma on a leg with extensive sun damage

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Case description

• A 76 year old Caucasian man presents with fungating basal cell carcinomas (BCC’s) on the lower limbs, and a suspicious nodule on the left shin (Figure 1)
• He reports the nodule had developed rapidly over the last month, but denied itch or tenderness
• Past medical history was significant for multiple BCC’s following decades of unprotected exposure to the Australian sun in his work as a farmer

Histopathology

• The nodule was excised and an exophytic, ulcerated dermal spindle-cell tumour was appreciated (Figure 2), 60x55x27 mm
• The tumour expanded to the deep reticular dermis with a rounded edge and focal infiltrative advancements
• Pleomorphic spindle cells with highly atypical nuclei were arranged in a fascicular/storiform architecture
• The tumour stained diffusely strong for CD10, but negative for S100/MART1/Desmin/Cytokeratins 5 and 6

Discussion

• Due to the rapid growth, large size, and prominent ulceration, the tumour was diagnosed as a pleomorphic dermal sarcoma (PDS)
• PDS are rare cutaneous neoplasms of fibrohistiocytic mesenchymal origin almost predominantly found on the head/neck of older men, sites most affected by chronic UV-induced DNA damage¹
• PDS resemble atypical fibroxanthoma (AFX), but tend to be larger, more locally invasive, and more likely to recur and metastasize (28 and 10% rates, respectively)²
• PDS and AFX are diagnoses of exclusion, after appropriate stains to rule out spindle cell melanoma, SCC, leiomyosarcoma
• PDS have not been reported in lower limbs, however the history of chronic Australian sun exposure and adjacent advanced BCC’s (a surrogate marker for extensive actinic damage) explains the atypical location in this case

Management

• Excision is gold standard for PDS, including Moh’s surgery³ - no recurrence has been noted 1 year post excision in our case
• The BCC’s responded well to vismodegib, an oral Hedgehog inhibitor, however was accompanied by severe muscle cramps

References

1. Soleymani et al. (2019); Dermatology Clinics
2. Mentzel et al. (2017); Surgical Pathology Clinics
3. Rosenfeld et al. (2019); International Journal of Dermatology

Figure 1: erythematous nodule with overlying scale on the left anterior shin (arrow), note adjacent ulcerated BCC inferomedially

Figure 2: (a) Ulcerated dermal spindle cell tumour; (b) highly atypical spindle cells, 5+ mitoses (arrow); and (c) strong diffuse staining for CD10