A rapidly-growing friable nodule on the cheek

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Abstract

Atypical fibroxanthoma and pleomorphic dermal sarcoma are on a spectrum of cutaneous tumors that present as ulcerated lesions in older adults. We present an 84-year-old man with pleomorphic dermal sarcoma, initially presenting as a bleeding lesion of the cheek that progressed to an eroded nodule.

Keywords: atypical fibroxanthoma, dermal sarcoma, neoplasm, pleomorphic, spindle cell, undifferentiated

Introduction

Atypical fibroxanthoma (AFX) and pleomorphic dermal sarcoma (PDS) represent a continuum of cutaneous tumors that range from confinement within the dermis to subcutaneous spread [1]. They typically present as ulcerated tumors located on chronically sun-exposed skin and are diagnosed later in life [2]. Because they can present similarly to other skin neoplasms, these tumors are often considered a diagnosis of exclusion.

Case Synopsis

An 84-year-old man with a history of squamous cell carcinoma presented to his primary care physician for evaluation of a bleeding skin lesion on his left cheek, present for four months (**Figure 1**). When he was seen in dermatology clinic one month later, physical examination showed a markedly larger 2cm eroded nodule (**Figure 2**). A shave biopsy was done to evaluate for malignancy. Histology sections showed a tumor of atypical spindle cells in the dermis, transected at the base (**Figure 3**). Tumor cells were positive for CD10 and SMA (**Figure 4A**); they were negative for S100, desmin, Cam5.2 (**Figure 4B**), AE1/AE3 (**Figure 4C**), and CD34. This histological and immunohistochemical characterization was consistent with pleomorphic dermal sarcoma (PDS).

Mohs micrographic surgery was performed, and intraoperative frozen sections revealed 6.0mm invasion depth with subcutaneous fat invasion and tumor necrosis. There was no lymphovascular invasion and no perineural invasion; margins were clear.

Case Discussion

Pleomorphic dermal sarcoma is a tumor typically arising in sun-exposed areas of elderly patients [2]. Histologically, PDS is characterized by spindle cells mingled with atypical histiocytic cells. Mitotic figures, prominent eosinophilic nucleoli, and a

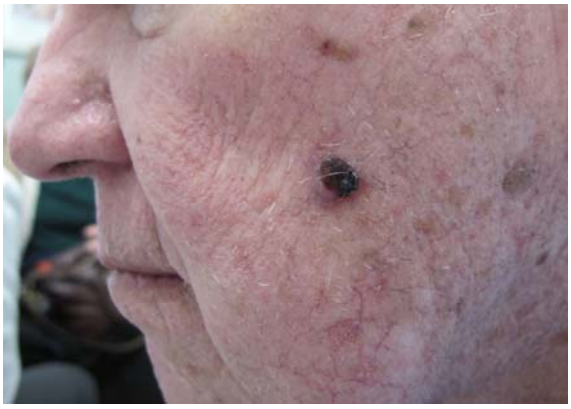

Figure 1. *Burgundy-crusted papule on the left cheek.*

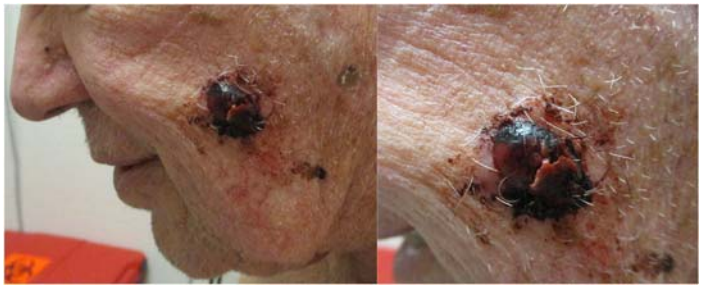

Figure 2. *One month later, the lesion was bleeding and markedly increased in size.*

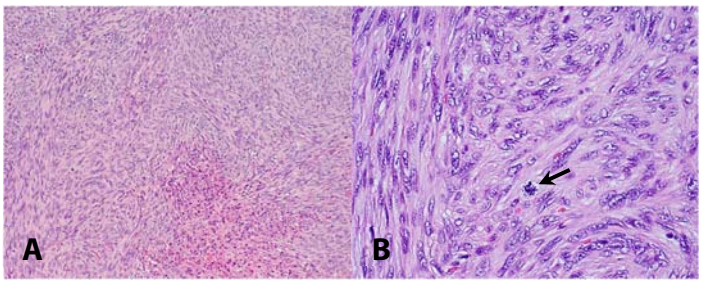

Figure 3: *H&E histopathology of the tumor showing **A)** atypical spindle cell proliferation in the dermis with focal tumor necrosis, 100×; and **B)** atypical mitotic figure depicted at the arrow, 400×.*

biphasic tumor cell population are common [3]. It is considered a diagnosis of exclusion, as other neoplasms such as squamous cell carcinoma and angiosarcoma should be ruled out first [2]. Pleomorphic dermal sarcoma exists on a clinicopathologic spectrum with AFX, with PDS having a more aggressive clinical and histological phenotype [3]. Both pathologies commonly present as ulcerated tumors of the skin, most commonly on the scalp [2].

Pleomorphic dermal sarcoma and AFX are typically differentiated solely by the increased propensity of pleomorphic dermal sarcoma to metastasize and recur when compared to atypical fibroxanthoma [2]. Despite this distinction, AFX and PDS rarely metastasize and have a low recurrence rate (5.6%). Recurrence is more likely for tumors infiltrating the subcutis, especially when excision margins are involved [2]. Thus, the difference in recurrence between AFX and PDS is believed to relate to differences in spindle morphology, with PDS morphology leading to increased subcutaneous invasion and more aggressive behavior [2].

The primary treatment goal for AFX is tumor removal with clear surgical margins. Alhough there is a paucity of data for the use of Mohs micrographic surgery to treat AFX, staged excision with margin control is believed to reduce the likelihood of recurrence. Wide local excision has led to varying rates of recurrence and is another first-line treatment [1].

Conclusion

Atypical fibroxanthoma and PDS are a spectrum of cutaneous tumors characterized by proliferation in the dermis, and occasionally into the subcutaneous tissues. We present this exemplary case of PDS to highlight its clinical appearance, tendency to progress rapidly, and recent literature on the subject.

Potential conflicts of interest

The authors declare no conflicts of interest.

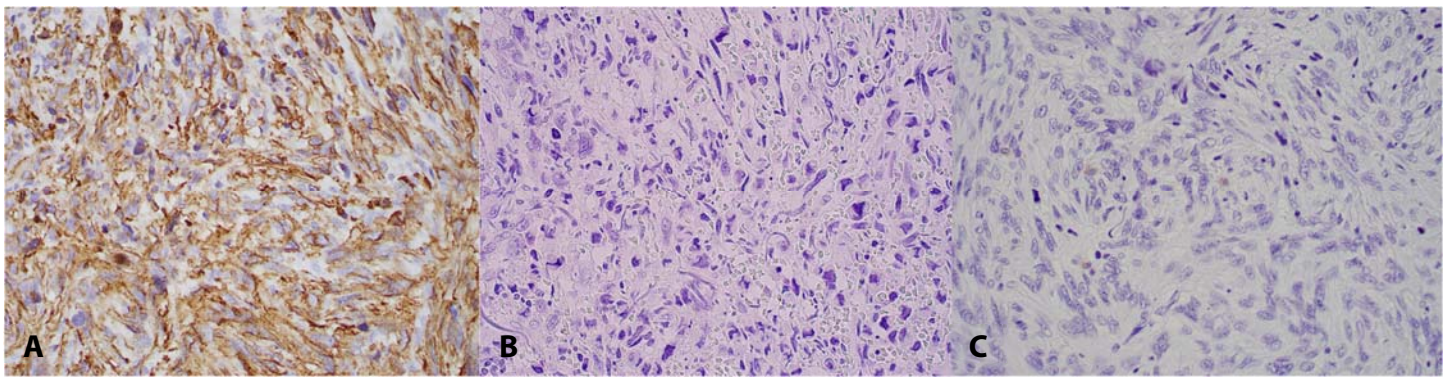

Figure 4. *Immunohistochemistry of the tumor. The tumor cells are positive for **A)** SMA; and negative for **B)** Cam5.2, and **C)** AE1/AE3. **A-C)** 400×.*

References

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