

## SHORT COMMUNICATION

**A New Skin-Colored Nodule in a Patient with Cushing's Disease**

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A 26-year-old woman with newly diagnosed Eosinophilic Granulomatosis with Polyangiitis and a recent history of steroid-induced Cushing's Disease, as evidenced by an 18 kilogram weight gain, moon facies, scattered ecchymoses, and new-onset stage I hypertension, presented with a newly found growth on the back. She denied trauma, itching, pain, or discharge. There was no personal or family history of melanoma or non-melanoma skin cancer. Physical examination revealed a single 6mm pink nodule of the left paraspinal mid-back (Figure 1) with no other cutaneous findings. Excisional biopsy specimens were obtained (Figure 1).

Initial review of the histology showed fascicles of epithelioid and fusiform cells with abundant cytoplasm in a background mucinous stroma. Immunohistologic examination showed EMA positivity around the theques as well as positivity for S-100, SOX-10, MiTF, HMB-45 and NKI-C3. Stains for AE1/AE3 cytokeratin, p40, Chromogranin A, GFAP, CD68, Smooth Muscle Actin and CD57 were negative. After excision of the initial nodule, a second similar nodule appeared eight months later, which was excised. The patient was subsequently tapered off of corticosteroid therapy and has had no further recurrence for over two years.

This lesion's positive immunoreactivity with melanoma markers along with its low Ki-67 proliferation profile and positive EMA reactivity surrounding tumor theques indicate both melanocytic and nerve sheath differentiation<sup>1</sup>(Figure 2). These two distinct staining patterns combined with the histologic findings lead to a final diagnosis of melanocytic schwannoma, first described by Carney in 1990<sup>2,3</sup>. The terminology "melanotic schwannoma" and "melanocytic schwannoma" can be found variably throughout the literature. However, some authors suggest the utilization of melanotic schwannoma for schwannomas that produce melanin pigment, and melanocytic schwannoma for those that are amelanotic, but that demonstrate both schwannian and melanocytic lineage<sup>2</sup>. In this case, the tumor did not produce melanin pigment and did show evidence of schwannian and melanocytic differentiation. As such, the term melanocytic schwannoma will be used to describe this case. Melanocytic schwannoma is an extremely rare tumor and dermal presentations are even more rare. To date, less than 20 primary cutaneous or subcutaneous tumors have been reported in the literature<sup>1</sup>. These tumors may arrive sporadically or in the setting of the Carney complex<sup>4</sup>, which involves mutations in the PRKAR1A gene and associated endocrinopathies, cutaneous and cardiac myxomas, and additional cutaneous findings

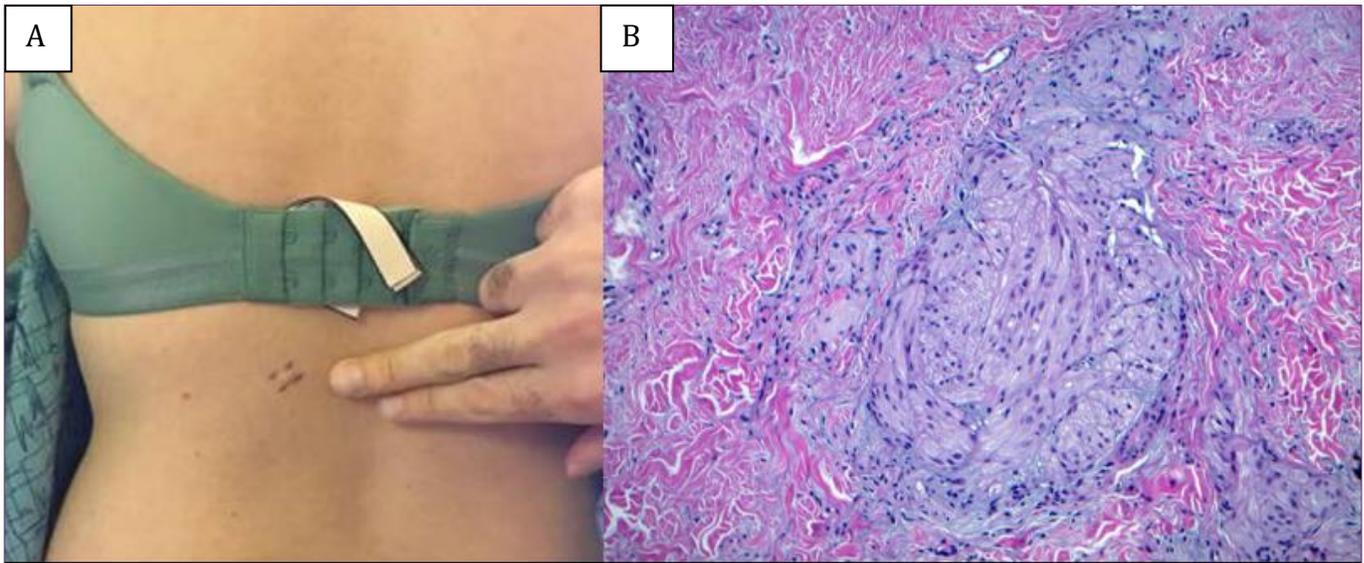


Figure 1. A) Melanocytic schwannoma clinical photo B) H&E stain at 10X power

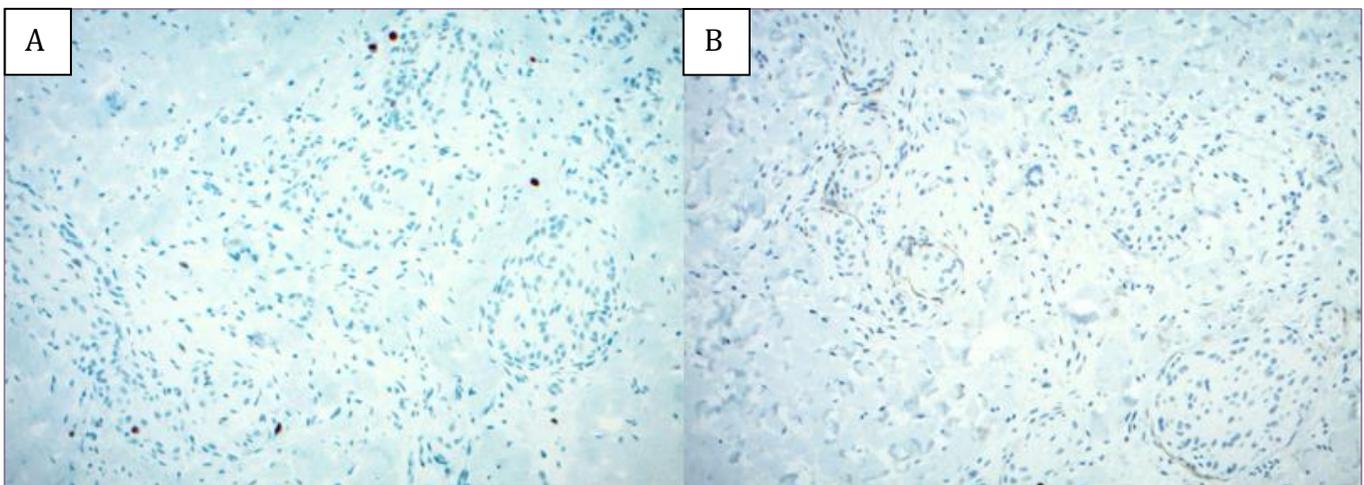


Figure 2. A) Ki-67 staining at 10X B). EMA staining at 10X

such as lentigines and blue nevi<sup>3</sup>. In Carney's identification of seventeen patients diagnosed with melanotic schwannomas, nine patients also had a diagnosis of Cushing's<sup>3</sup>. Our patient exhibited no additional findings of the Carney complex, however in light of this observation, it is possible that there is a correlation between her iatrogenic Cushing's through high dose exogenous corticosteroid immunosuppressive therapy and her development of a cutaneous melanocytic

schwannoma; however, this remains to be clarified in further studies. Melanocytic schwannomas are rare tumors, and their clinical courses are uncertain. Additionally, they may mimic other cutaneous lesions and be difficult to distinguish on pathology. Therefore, complete excision is recommended, along with possible genetic analysis for diagnostic confirmation<sup>4,5</sup>.

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