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Demographics and outcomes of eccrine porocarcinoma: Results from the National Cancer Database

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Eccrine porocarcinoma (EP) is a rare type of skin malignancy arising from eccrine sweat glands. Owing to limited case-load, larger database studies are needed for the epidemiologic characterization and assessment of treatment outcomes of EP. The National Cancer Database (NCDB) was queried for all confirmed EP cases of the skin diagnosed between 2004 and 2015. Frequency functions, Kaplan-Meier and Cox regression models were used to analyze demographics, treatment, and survival. We identified 568 cases. The most common primary site was the lower limb and hip (33.1%) followed by the head and neck (28.9%). The average age of diagnosis was 62.1. Men and women were affected equally ($P > .05$). The most common treatment was surgery alone (91.8%). 61.9% of patients were diagnosed with AJCC stage I and 30.0% with stage II disease. 1.1% of patients had metastases at diagnosis. The sites of metastases were bone (50%), lung (40%), and brain (10%). The median tumor size was 25.5 millimeters (largest diameter). The 5-year and 10-year overall survivals (OS) were 68.7% and 53.3%, respectively. On univariate analysis, younger age, surgery, radiation, black race, and lower stages were associated with increased OS ($P < .05$). Trunk lesions were associated with decreased OS (5-year 58.5% and 10-year 41.2%). On multivariate analysis, older age (HR 1.067; $P < .05$) and tumors greater than 60 millimeters (largest diameter) (HR 2.8; $P < .05$) were associated with decreased OS. This study represents the largest cohort of eccrine porocarcinoma to our knowledge. More evidence is needed to create treatment guidelines for this rare condition.

Commercial disclosure: None identified.

18132

Hydroxychloroquine for refractory oral lichen planus

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Background: Atrophic and erosive oral lichen planus (AEOLP) is a painful, chronic inflammatory disease without a cure that requires long-term treatment. Many patients fail to improve on topical steroid therapy alone and data on the use of systemic therapies are limited. Several medications, mostly immunosuppressive, have been reported in small numbers of patients, but pose potentially serious side effects and little information on long term outcomes is available. Owing to its relatively favorable safety profile, we evaluated the efficacy and safety of hydroxychloroquine (HCQ) in patients with AEOLP refractory to topical steroids.

Methods: A retrospective chart review was performed on patients seen at the Oregon Health & Science University Dermatology Department, identifying those with biopsy-proven disease who failed to improve with topical steroids and started HCQ as their next therapy.

Results: Of 59 patients screened, 30 met inclusion/exclusion criteria. At 3 months, 6 months, and long term (average 27 months) time points, 75%, 85%, and 87% of patients, respectively, showed at least mild to moderate improvement. No patient worsened and there were no serious adverse effects.

Conclusions: This is the largest number of AEOLP patients treated with HCQ reported to date. The majority improved with sustained benefit over an average of 27 months, without serious side effects. Its favorable safety profile makes HCQ a desirable therapy for chronic diseases and it is a reasonable option for systemic therapy in AEOLP patients not responding to topical steroids.

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18148

Myelin basic protein autoantibodies as a biomarker in morphea

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The immune underpinnings of morphea are poorly described. Although numerous reports document autoantibodies associated with morphea, few have been replicated in large, controlled studies or are of clinical or mechanistic importance. We sought to characterize immunologic alterations and identify autoantibodies associated with morphea. We found increased frequency of CD3−CD138+ plasma cells and a decrease in CD19+CD138+ plasmablasts in morphea patients compared with controls, suggesting an alteration in the maturation, activity level, and function of antibody-secreting B cells of morphea patients. We then utilized an autoantigen microarray to assess potential autoantibodies associated with morphea, which identified increased frequency of myelin basic protein (MBP) autoantibodies in morphea patients (27.1% vs 5.7% in controls). Epitope mapping revealed target epitopes for MBP autoantibodies in morphea were distinct from epitopes in multiple sclerosis patients. Immunohistochemistry of biopsies revealed increased inflammation surrounding peripheral cutaneous nerves in morphea samples. When analyzing clinical severity, morphea patients with MBP autoantibodies were more likely to have increased pain symptoms (13.2% vs 0%, $P < .05$), higher disease damage (20 vs 11.5, $P < .05$), and more limitations in normal activities due to their skin lesions (73.7% vs 41.7%, $P < .05$) as measured by validated clinical scores and quality of life measures. Taken together, these results identify MBP autoantibodies as a potential driver of morphea pathogenesis and as a possible biomarker for morphea disease severity.

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18178

Clinical characteristics and treatment of lupus erythematosus tumidus

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Lupus erythematosus tumidus (LET) is a rare photosensitive dermatosis. LET was considered a subtype of chronic cutaneous lupus erythematosus (CLE); however, LET's clinical course and favorable prognosis led to its reclassification into another category called intermittent CLE. Although known for over 100 years, LET's association with systemic lupus erythematosus (SLE), auto-antibody profile, and disease prognosis is not well characterized. The purpose of this study was to describe the demographics, clinical characteristics, auto-antibody profile, comorbidities, and treatment of LET. A retrospective review was conducted in subjects with histologically diagnosed LET from July 2012 to July 2018 at Wake Forest Department of Dermatology. Inclusion criteria included men or women aged 18-75 with biopsy-proven LET. Biopsy-proven LET was defined as superficial and deep lymphocytic infiltrate with abundant mucin deposit in the reticular dermis and absent or focal dermoepidermal junction alterations. Charts were evaluated for demographics, clinical characteristics, diagnoses, auto-antibodies, treatment, and recurrence. We observed that hydroxychloroquine as first-line systemic treatment was worthwhile, with 60% of cases reaching clearance with this intervention alone. Moreover, we found that recalcitrant disease can be managed with other immunomodulators including quinacrine, methotrexate, or thalidomide with varying degrees of efficacy.

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