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Abstract

Advanced basal cell carcinomas appear preferentially on the scalp of patients with Basal Cell Carcinoma Nevus Syndrome

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Basal Cell Carcinoma Nevus Syndrome is a rare condition in which patients need to meet certain major or minor diagnostic criteria or test positive for chromosome 9 or *PTCH1* mutations to be diagnosed. The results of an internet based survey of self-identified adults with BCCNS which was launched through SurveyMonkey with access provided by the Basal Cell Carcinoma Nevus Syndrome Life Support Network (www.BCCNS.org). Of 45 respondents, 11 individuals reported having locally advanced BCCs and 6 individuals reported having both locally advanced and metastatic BCCs. Of the 11 individuals with locally advanced BCCs, 8 reported having lesions on the scalp. Of the 6 patients with locally advanced BCCs, 3 reported having abnormally thick hair distribution. There were 6 individuals with both locally advanced and metastatic BCCs and 5 reported having BCCs on their scalp. Thick hair distribution was reported by 3 of 4 patients with locally advanced and metastatic BCCs. Overall, it was determined that patients with BCCNS develop aggressive BCCs on their scalp despite thick hair distribution. Patients with BCCNS have an overactive hedgehog signaling pathway. This pathway is also known to play a role in follicle cycling and is believed to lead to increased rates of malignancy.[1,2] Despite the presence of thick hair, patients with BCCNS should have their scalp skin closely examined to identify and treat BCCs at early stages of development.