GENETICS AND GENODERMATOSES

PREVALENCE STUDY OF SKIN CANCERS IN A RETROSPECTIV COHORT OF XERODERMA PIGMENTOSUM (XP) IN BLACK PATIENTS

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Introduction: XP is a rare genetic disease associated with early onset of numerous skin cancers.

Objective: Our main objective was to determine prevalence of skin cancers in a cohort of XP-C black Mahori patients.

Materials and methods: A monocentric cohort consisting of all XP patients followed at Mayotte Island until May 2017 by dermatologists from Saint Denis University Hospital (Reunion island) during dermatological missions.

Results: Eighteen patients with median age of 15.3 years (61% male) belonged to 14 families. All carried the same mutation (XP-C). Median age at clinical diagnosis was 1.8 years. A total of 144 skin cancers (94 squamous cell carcinomas (SCC), 30 basal cell carcinomas (BCC), 14 melanomas, 5 sarcomas and 1 sarcomatoid carcinoma) were observed among 11/18 patients (61%). Eleveen patients (61%) had at least 1 SCC, 6 (33%) at least 1 BCC, 6 (33%) had at least 1 melanoma and 4 (22%) had at least 1 sarcoma. Cancers occurred in light-exposed areas for 95.5%. Median age at first cancer was 5.4 years, and 6.4 years at second cancer. SCC and sarcomas occurred earlier than CBC and melanomas ($p < 0.0001$). All patients had mild to severe poikiloderma and suffered from photophobia. Half had palmo-plantar pigmented lesions, one-third oral mucosal involvement, 78% ocular or palpebral lesions, 17% nail and hair involvement. Median Dermatology Life Quality Index (DLQI) was 4/30. The severity of poikiloderma was significantly correlated with occurrence of skin cancers.

Conclusion: Our cohort showed high prevalence of skin cancer in XP-C black patients exposed to UV radiation in a tropical area. Most common tumors are SCC. Prevalence of melanoma is high, with major risk compared to same skin phototype. We found high prevalence of sarcomas. First cancer occurs early (5.4 years) compared to data from

literature. SCC and sarcomas occurred significantly faster than melanomas and BCC.